Effective Date **6/1/2023**
Next Review Date… **6/1/2024**
Coverage Policy Number **IP0120**

Crizanlizumab-tmca

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Related Coverage Resources

INSTRUCTIONS FOR USE
The following Coverage Policy applies to health benefit plans administered by Cigna Companies. Certain Cigna Companies and/or lines of business only provide utilization review services to clients and do not make coverage determinations. References to standard benefit plan language and coverage determinations do not apply to those clients. Coverage Policies are intended to provide guidance in interpreting certain standard benefit plans administered by Cigna Companies. Please note, the terms of a customer's particular benefit plan document [Group Service Agreement, Evidence of Coverage, Certificate of Coverage, Summary Plan Description (SPD) or similar plan document] may differ significantly from the standard benefit plans upon which these Coverage Policies are based. For example, a customer's benefit plan document may contain a specific exclusion related to a topic addressed in a Coverage Policy. In the event of a conflict, a customer's benefit plan document always supersedes the information in the Coverage Policies. In the absence of a controlling federal or state coverage mandate, benefits are ultimately determined by the terms of the applicable benefit plan document. Coverage determinations in each specific instance require consideration of 1) the terms of the applicable benefit plan document in effect on the date of service; 2) any applicable laws/regulations; 3) any relevant collateral source materials including Coverage Policies and; 4) the specific facts of the particular situation. Coverage Policies relate exclusively to the administration of health benefit plans. Coverage Policies are not recommendations for treatment and should never be used as treatment guidelines. In certain markets, delegated vendor guidelines may be used to support medical necessity and other coverage determinations.

Overview

This policy supports medical necessity review for crizanlizumab-tmca (**Adakveo®**).

Receipt of sample product does not satisfy any criteria requirements for coverage.

Medical Necessity Criteria

Crizanlizumab-tmca (Adakveo) is considered medically necessary when the following are met:

Sickle Cell Disease. Individual meets **ALL** of the following criteria:
A. Age 16 years or older
B. Has had at least one vaso-occlusive crisis (VOC) in the previous 12 month period (before initiating Adakveo therapy)
C. Will be used concurrently with hydroxyurea, unless has had failure, contraindication, or intolerance to hydroxyurea

D. Medication is prescribed by, or in consultation with, a hematologist or a physician who specializes in sickle cell disease

Dosing. 5 mg/kg administered by intravenous infusion at Week 0, Week 2, and then once every 4 weeks thereafter

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When coverage is available and medically necessary, the dosage, frequency, duration of therapy, and site of care should be reasonable, clinically appropriate, and supported by evidence-based literature and adjusted based upon severity, alternative available treatments, and previous response to therapy.

Reauthorization Criteria

Continuation of crizanlizumab-tmca (Adakveo) is considered medically necessary for the treatment of sickle cell disease when the above medical necessity criteria are met AND there is documentation of beneficial response.

Authorization Duration

Initial approval duration: up to 12 months.
Reauthorization approval duration: up to 12 months

Conditions Not Covered

Any other use is considered experimental, investigational or unproven including the following (this list may not be all inclusive):

Concomitant Oxbryta™ Therapy. The efficacy of Adakveo was established in one Phase II, randomized, double-blind, placebo-controlled, multicenter 12-month study called SUSTAIN (published) [n = 198]. The study included two different doses of Adakveo: 2.5 mg/kg IV (which is not the FDA-approved dose) [n = 66] and 5 mg/kg IV (n = 67). Patients received Adakveo or placebo with or without hydroxyurea. Adakveo is an effective add-on therapy for those who continue to experience sickle cell-related pain while on hydroxyurea. In addition, Adakveo as monotherapy represents a treatment option for patients who cannot tolerate or cannot take hydroxyurea. The place in therapy for Adakveo will be further defined with its placement in guidelines, use in clinical practice, and additional efficacy data.

Coding Information

Note: 1) This list of codes may not be all-inclusive.
2) Deleted codes and codes which are not effective at the time the service is rendered may not be eligible for reimbursement.

Considered Medically Necessary when criteria in the applicable policy statements listed above are met:

HCPCS Codes	Description
J0791	Injection, crizanlizumab-tmca, 5 mg

Background

OVERVIEW
Adakveo, a monoclonal antibody, is indicated to **reduce the frequency of vasoocclusive crises** in patients 16 years and older with **sickle cell disease**.[1]

Adakveo is given by intravenous infusion over a period of 30 minutes at Week 0, Week 2, and every 4 weeks thereafter; the dose is 5 mg/kg.

Guidelines
The American Society of Hematology guidelines for sickle cell disease: management of acute and chronic pain associated with sickle cell disease (2020) does not address the use of Adakveo.[2] The National Institutes of Health – National Heart, Lung, and Blood Institute issued the Evidence-Based Management of Sickle Cell Disease, Expert Panel Report in 2014.[3] These guidelines were published prior to the approval of Adakveo. Hydroxyurea has been shown to reduce the frequency of painful episodes, the incidence of acute coronary syndrome events, and the need for transfusions and hospitalizations.

References

1. Adakveo® intravenous infusion [prescribing information]. East Hanover, NJ: Novartis; September 2022.
2. Brandow AM, Carroll CP, Creary S, et al. American Society of Hematology 2020 guidelines for sickle cell disease: management of acute and chronic pain. *Blood Adv*. 2020;4:2656-2701.
3. The National Institutes of Health – National Heart, Lung, and Blood Institute Evidence-Based Management of Sickle Cell Disease, Expert Panel Report 2014. Available at: https://www.nhlbi.nih.gov/sites/default/files/media/docs/sickle-cell-disease-report%20020816_0.pdf. Accessed on November 28, 2022.