CareSource Rxinnovations

PHARMACY POLICY STATEMENT
Arkansas PASSE

DRUG NAME	Adakveo (crizanlizumab-tmca)
BILLING CODE	J0791 (1 unit = 5 mg)
BENEFIT TYPE	Medical
SITE OF SERVICE ALLOWED	Outpatient Hospital/Office/Infusion Site
STATUS	Prior Authorization Required

Adakveo was approved by the FDA in 2019. It is indicated to reduce the frequency of vaso-occlusive crises (VOCs) in adults and pediatric patients aged 16 years and older with sickle cell disease. Adakveo may be given with or without hydroxyurea.
Sickle cell disease is caused by an inherited mutation in the beta globin gene, resulting in abnormal hemoglobin called sickle hemoglobin (HbS). Red blood cells become rigid, undergo premature hemolysis leading to anemia, and become unable to transport oxygen to critical organs. Patients experience severe pain from vaso-occlusive crises. First line therapy for sickle cell disease is hydroxyurea.
Adakveo binds to P-selectin and blocks interactions with its ligands, including P-selectin glycoprotein ligand 1 (PSGL-1). It can also dissociate preformed Pselectin/PSGL-1 complex. Binding P-selectin on the surface of the activated endothelium and platelets blocks interactions between endothelial cells, platelets, red blood cells, and leukocytes.

Adakveo (crizanlizumab-tmca) will be considered for coverage when the following criteria are met:

Sickle Cell Disease

For **initial** authorization:
1. Member must be 16 years of age or older; AND
2. Medication must be prescribed by or in consultation with a hematologist or a physician who has experience in treating sickle cell disease; AND
3. Member has a documented diagnosis of sickle cell disease with at least 2 vaso-occlusive pain crises in the past 12 months; AND
4. Member has tried hydroxyurea for at least 3 months and it was ineffective or not tolerated; AND
5. Medication will not be used concurrently with Oxbryta (voxelotor) therapy.
6. **Dosage allowed/Quantity limit:** 5 mg/kg intravenously at week 0, week 2, and every 4 weeks thereafter.

If all the above requirements are met, the medication will be approved for 6 months.

For **reauthorization**:
1. Chart notes have been provided to show that the member has experienced a reduction in frequency of vaso-occlusive crises since starting treatment.

If all the above requirements are met, the medication will be approved for an additional 12 months.

CareSource considers Adakveo (crizanlizumab-tmca) not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.

DATE	ACTION/DESCRIPTION
04/17/2020	New policy for Adakveo created.
06/18/2020	New J Code added
08/21/2020	Removed Endari from trial requirement.
02/21/2022	Transferred to new template. Removed “all initial criteria” from reauth. Added diagnosis of sickle cell to pain crisis criteria. Modified wording of hydroxyurea trial to match Oxbryta policy.

References:

1. Adakveo [Package Insert]. East Hanover, NJ: Novartis; July 2021.
2. Ataga KI, Kutlar A, Kanter J, et al. Crizanlizumab for the prevention of pain crises in sickle cell disease. *N Engl Med*. 2017;376(5):429-439.
3. Evidence-Based Management of Sickle Cell Disease. US Department of Health and Human Services. 2014.
4. Niihara Y, Miller ST, Kanter J, et al. A phase 3 trial of L-glutamine in sickle cell disease. *N Engl Med*. 2018;379:226-235.
5. Reprixys Pharmaceutical Corporation. Study to Assess Safety and Impact of SelG1 With or Without Hydroxyurea Therapy in Sickle Cell Disease Patients With Pain Crises (SUSTAIN). NLM Identifier: NCT01895361.
6. Kutlar A, Kanter J, Liles DK, et al. Effect of crizanlizumab on pain crises in subgroups of patients with sickle cell disease: A SUSTAIN study analysis. Am. 2019;94(1):55-61.
7. Bradt P, Spackman E, Synnott PG, Chapman R, Beinfeld M, Rind DM, Pearson SD.
8. Crizanlizumab, Voxelotor, and L-Glutamine for Sickle Cell Disease: Effectiveness and Value. Institute for Clinical and Economic Review, January 23, 2020. https://icer-review.org/material/sickle-cell-disease-draft-evidence-report/.
9. Voskaridou E, Christoulas D, Bilalis A, et al. The effect of prolonged administration of hydroxyurea on morbidity and mortality in adult patients with sickle cell syndromes: results of a 17-year, single-center trial (LaSHS). *Blood*. 2010;115(12):2354-2363.
10. Rodgers GP, George A. Hydroxyurea use in sickle cell disease. In: UpToDate, Post TW (Ed), UpToDate, Waltham, MA. (Accessed on August 21, 2020).

Effective date: 07/01/2022
Revised date: 02/21/2022