HAP CareSource™

PHARMACY POLICY STATEMENT
HAP CareSource™ Marketplace

DRUG NAME	Adakveo (crizanlizumab-tmca)
BENEFIT TYPE	Medical
STATUS	Prior Authorization Required

Adakveo, approved by the FDA in 2019, is a selectin blocker indicated to reduce the frequency of vaso-occlusive crises (VOCs) in adults and pediatric patients aged 16 years and older with sickle cell disease. It may be given with or without 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.
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 (crizanlizumab-tmca) will be considered for coverage when the following criteria are met:

Sickle Cell Disease (SCD)

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; AND
3. Member has a documented diagnosis of sickle cell disease with at least **TWO** vaso-occlusive pain crises in the past 12 months; AND
4. Member has tried hydroxyurea for at least 6 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.

HAP 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.
01/23/2024	Updated references; increased hydroxyurea trial length from 3 months to 6 months; removed prescriber specialty of a physician who has experience in treating sickle cell disease.

References:

1. Adakveo [Package Insert]. East Hanover, NJ: Novartis; 2022.
2. Ataga KI, Kutlar A, Kanter J, et al. Crizanlizumab for the Prevention of Pain Crises in Sickle Cell Disease. *N Engl J Med.* 2017;376(5):429-439. doi:10.1056/NEJMoa1611770
3. Yawn BP, Buchanan GR, Afenyi-Annan AN, et al. Management of sickle cell disease: summary of the 2014 evidence-based report by expert panel members [published correction appears in JAMA. 2014 Nov 12;312(18):1932] [published correction appears in JAMA. 2015 Feb 17;313(7):729]. *JAMA.* 2014;312(10):1033-1048. doi:10.1001/jama.2014.10517
4. Bradt P, Spackman E, Synnott PG, Chapman R, Beinfeld M, Rind DM, Pearson SD. 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/.
5. 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.

Effective date: 01/01/2025
Revised date: 01/23/2024