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# Crizanlizumab-tmca

## (Adakveo®) J0791 (5mg)

### Covered with prior authorization

Requests for Adakveo® (crizanlizumab-tmca) may be approved if the following criteria are met:

- Sickle Cell Disease and individual meets **ALL** of the following criteria:
  - 16 years of age or older
  - Documented diagnosis of Sickle Cell Disease (SCD)
  - Individual has had at least one vaso-occlusive crisis (VOC) in the past 12 months
  - Will be used concurrently with hydroxyurea unless individual has a contraindication per FDA label, significant intolerance (e.g., unacceptable toxicity), or is otherwise not a candidate for hydroxyurea (e.g., individual who is planning to become pregnant, is currently pregnant, or has an immunosuppressive condition, such as cancer)
  - Medication is being prescribed by, or in consultation with, a hematologist or a physician who specializes in sickle cell disease

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.

Requests for Crizanlizumab-tmca may **not** be approved if the above criteria are not met and for all other indications not included above.

**FDA Approved Dosing and Availability**
Adakveo® (crizanlizumab) 10 mg/ ml vial is dosed at 5 mg/ kg every 4 weeks. Initiation of therapy for Adakveo® includes loading dosing so may approve 5mg/kg at week 0 and week 2.

**Initial and reauthorization is up to 12 months.**

**Annual reauthorizations will require medical chart documentation that the patient has been seen within the past 12 months and that markers of disease are improved by therapy.**

**Deleted codes and codes which are not effective at the time the service is rendered may not be eligible for reimbursement.**

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**Exclusion criteria:**

- 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.
- Doses, durations, or dosing intervals that exceed FDA maximum limits for any FDA-approved indication or are not supported by industry-accepted practice guidelines or peer-reviewed literature for the relevant off-label use.
- Individuals with significant known risk factors unless the record provides an assessment of clinical benefit that outweighs the risk.

**U.S. Food and Drug Administration:**
This section is to be used for informational purposes. FDA approval alone is not a basis for coverage. Adakveo® (crizanlizumab-tmca) is a monoclonal antibody that binds to and inhibits Pselectin, an adhesion protein found on the surface platelets and endothelial cells. In those with sickle cell disease, P-selectin promotes blood vessels ``sticking" with sickle cells, which causes inflammation and pain crises also called vaso-occlusive crises (VOCs). VOCs are unpredictable, acute episodes of severe pain that can lead to serious life-threatening complications and death in people with sickle cell disease. Adakveo® is administered as a once monthly infusion based on the individual's weight. Adakveo® may be used with or without hydroxyurea.

**Key References Accessed 8/2022:**
1. Adakveo® intravenous infusion [prescribing information]. East Hanover, NJ: Novartis; July 2021.
2. Piel FB, Steinberg MH. Sickle cell disease. N Engl J Med. 2017;376:1561-1573.
3. The National Institutes of Health – National Heart, Lung, and Blood Institute Evidence-Based Management of Sickle Cell Disease, Expert Panel Report. Available at: https://www.nhlbi.nih.gov/sites/default/files/media/docs/sickle-cell-disease-report%20020816_0.pdf.
4. 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.
5. Clinical Pharmacology [database online]. Tampa, FL: Gold Standard, Inc.: 2022. URL: http://www.clinicalpharmacology.com. Updated periodically.
6. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2022; Updated periodically

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| Date | Summary of Changes |
|---|---|
| August 2022 | Criteria for use summary developed by the Ascension Medical Specialty Prior Authorization Team. |
| September 2022 | Criteria for use summary approved by the Ascension Ambulatory Care Expert Review Panel. |
| October 2022 | Criteria for use summary approved by the Ascension Therapeutic Affinity Group. |

If you have questions, call 833-980-2352 to speak to a member of the Ascension Rx prior authorization team, or email your questions to smarthealthspecialty@ascension.org.