

## RX.PA.003.MPC ADAKVEO (CRIZANLIZUMAB-TMCA)

### Description

Adakveo® (Crizanlizumab-tmca) is a selectin blocker

### FDA Approved Indication(s)

To reduce the frequency of vasoocclusive crises (VOC) in adults and pediatric patients aged 16 years and older with sickle cell disease (SCD).

### Policy/Criteria

*Provider must submit documentation (such as office chart notes, lab results or other clinical information) supporting that member has met all approval criteria.*

#### A. Initial Approval Criteria

##### A. Sickle Cell Disease (must meet all):

1. Diagnosis of SCD with one of the following genotypes:
  - a. Homozygous hemoglobin S (HbSS);
  - b. Hemoglobin S $\beta^0$ -thalassemia;
  - c. Hemoglobin S $\beta^+$ -thalassemia;
  - d. Hemoglobin SC (HbSC);
2. Age  $\geq$  16 years;
3. Prescribed by or in consultation with a hematologist, or other specialist with training in management of sickle cell disease;
4. Hb level  $\geq$  4 g/dL;
5. Member meets one of the following (a or b):
  - a. Member has experienced at least 2 VOC within the past 6 months while on hydroxyurea at up to maximally indicated doses
  - b. Member has intolerance or contraindication to hydroxyurea and has experienced at least 2 VOC within the past 12 months
6. Failure of L- glutamine at up to maximally tolerated doses, unless contraindicated or clinically significant adverse effects are experienced;
7. Documentation of baseline incidence of VOC over the last twelve months;
8. Adakveo is prescribed concurrently with hydroxyurea, unless contraindicated or clinically significant adverse effects are experienced;
9. Adakveo is not prescribed concurrently with Oxbritya®;
10. Member is not concurrently being treated with chronic prophylactic blood transfusion therapy
11. Dose does not exceed 5 mg/kg doses on Day 1 and Day 15, followed by

**B. Must be prescribed at a dose within the manufacturer’s dosing guidelines (based on diagnosis, weight, etc) listed in the FDA approved labeling.**

**C. Adakveo will be considered investigational or experimental for any other use and will not be covered.**

**D. Continued Therapy**

All prior authorization renewals are reviewed on an annual basis to determine the Medical Necessity for continuation of therapy. Authorization may be extended at 1-year intervals based upon all of the following:

- MPC Renewal:
  - Chart documentation confirming positive response to therapy as evidenced by a documented improvement in the incidence of VOC from baseline;
  - Documentation that Adakveo continues to be prescribed concurrently with Hydroxyurea, unless contraindicated or clinically significant adverse effects are experienced;
  - Prescriber attests that Adakveo is not prescribed concurrently with Oxbryta
  - Prescribed by or in consultation with a hematologist, or other specialist with training in management of sickle cell disease
- Renewal from Previous Insurer:
  - Members who have prior approval (from insurer other than MPC), or have been receiving medication samples, should be considered under criterion A (Initial Authorization Criteria).
  - Provider has a documented clinical response of the member’s improvement in the incidence of VOC from baseline

**E. Limitations**

Length of Authorization (if above criteria met)	
Initial Authorization	Up to 6 months
Reauthorization	Up to 1 year

**F. Dosage and Administration**

Indication	Dosing Regimen	Maximum Dose
SCD	Administer 5 mg/kg by intravenous infusion over a period of 30 minutes on Week 0, Week 2, and every 4 weeks thereafter.	5 mg/kg

**G. Product Availability**

Single-dose vial for injection: 100 mg/10 mL (10 mg/mL)

**H. References**

1. Adakveo Prescribing Information. East Hanover, NJ: Novartis Pharmaceuticals Corporation; November 2019. Available at [https://www.pharma.us.novartis.com/sites/www.pharma.us.novartis.com/files/exjad\\_e.pdf](https://www.pharma.us.novartis.com/sites/www.pharma.us.novartis.com/files/exjad_e.pdf). Accessed December 2, 2019.
2. 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 J Hematol. 2019;94:55-61.
3. Ataga K, Kutlar A, Kanter J, et al. Crizanlizumab for the Prevention of Pain Crises in SickleCell Disease. N Engl J Med. 2017 Feb 2;376(5):429-439.
4. 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. JAMA. 2014 Sep 10;312(10):1033-48.
5. Micromedex® Healthcare Series [Internet database]. Greenwood Village, CO: ThomsonHealthcare. Updated periodically. Accessed December 4, 2019.

**HCPSC Code(s) and ICD-10-CM Diagnosis Codes that Support Coverage**

**Criteria** The following is a list of diagnosis codes that support coverage for the applicable covered procedure code(s).

HCPSC Code	Description
J0791	Injection, crizanlizumab-tmca, 5mg
ICD-10-CM Code	Description
D57.0	Hb-SS disease with crisis
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
D57.09	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
D57.21	Sickle-cell/Hb-C with crisis
D57.211	Sickle-cell/Hb-C with acute chest syndrome
D57.212	Sickle-cell/Hb-C with splenic sequestration
D57.213	Sickle-cell/Hb-C with cerebral vascular involvement
D57.218	Sickle-cell/Hb-C with other specified complication
D57.219	Sickle-cell/Hb-C with unspecified complication
D57.4	Sickle-cell thalassemia
D57.41	Sickle-cell thalassemia, unspecified with 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 vascularinvolvement
D57.418	Sickle-cell thalassemia, unspecified with other specifiedcomplication
D57.419	Sickle-cell thalassemia, unspecified with crisis
D57.42	Sickle-cell thalassemia beta zero without crisis
D57.43	Sickle-cell thalassemia beta zero 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 vascularinvolvement
D57.438	Sickle-cell thalassemia beta zero with other specified complication
D57.439	Sickle-cell thalassemia beta zero with unspecified complication
D57.44	Sickle-cell thalassemia beta plus without crisis

D57.45	Sickle-cell thalassemia beta plus with crisis
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 vascularinvolvement
D57.458	Sickle-cell thalassemia beta plus with other specified complication
D57.459	Sickle-cell thalassemia beta plus with unspecified complication
D57.80	Other sickle-cell disorders without crisis
D57.81	Other sickle-cell disorders with 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

ICD-10-CM Code	Description
D57.818	Other sickle-cell disorders with other specified complication
D57.819	Other sickle-cell disorders with unspecified complication

**REVIEW HISTORY**

DESCRIPTION OF REVIEW / REVISION	DATE APPROVED
Annual review Change in Non-MPC renewal to renewal from previous	02/2024

ADAKVEO (CRIZANLIZUMAB-TMCA)

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REVISION DATE: 02/2024

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<i>insurer</i>	
<i>Annual review</i>	<i>02/2023</i>
<i>Update of reauthorization criteria for MPC vs Non-MPC renewals. Update to PA durations table.</i>	<i>09/2022</i>
<i>Annual review</i>	<i>02/2022</i>
<i>Addition of dosing requirements and off-label restrictions</i>	<i>12/2021</i>
<i>P&amp;T Review</i>	<i>02/2021</i>
<i>New Policy</i>	<i>01/2021</i>