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 16years and older with sickle cell disease (SCD).

Policy/Criteria
Provider must submit documentation (such as office chart notes, lab results or other clinicalinformation) 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 ≥ 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 ≥ 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 Oxbryta®;
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 5 mg/kg every 4 weeks.

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
- Non- MPC Renewal:
  - Members who have previously been taking Adakveo and are requesting a non-MPC renewal should be considered under criterion A (Initial Authorization Criteria).
  - Member has not been receiving medication samples for Adakveo; AND
  - 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/exjade.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 Sep10;312(10):1033-48.
5. Micromedex® Healthcare Series [Internet database]. Greenwood Village, CO:

ThomsonHealthcare. Updated periodically. Accessed December 4, 2019.

HCPCS 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 coveredprocedure code(s).

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