Crizanlizumab-tmca (Adakveo®)

NDC CODE(S) 00078-0883-XX ADAKVEO 100MG/10ML Solution (NOVARTIS)

DESCRIPTION

Crizanlizumab-tmca is a humanized IgG2 kappa monoclonal antibody that binds to P-selectin and blocks interactions with its ligands including P-selectin glycoprotein ligand 1. Binding P-selectin on the surface of the activated endothelium and platelets blocks interactions between endothelial cells, platelets, red blood cells, and leukocytes.

POLICY

- Crizanlizumab-tmca to reduce the frequency of vasoocclusive crises in individuals with sickle cell disease is considered medically necessary if the medical appropriateness criteria are met. . (See Medical Appropriateness below.)
- Crizanlizumab-tmca for the treatment of other conditions/diseases is considered investigational.

MEDICAL APPROPRIATENESS

INITIAL APPROVAL CRITERIA

Universal Criteria
- Therapy will not be used in conjunction with voxelotor (Oxbryta) or L-glutamine (Endari); **AND**

Sickle Cell Disease
- Patient is at least 16 years of age; **AND**
- Patient has a confirmed diagnosis of sickle-cell disease, of any genotype (e.g., HbSS, HbSC, HbS/beta0-thalassemia, HbS/beta+-thalassemia, and others) as determined by one of the following:
  - Identification of significant quantities of HbS with or without an additional abnormal β-globin chain variant by hemoglobin assay; **OR**
  - Identification of biallelic HBB pathogenic variants where at least one allele is the p.Glu6Val pathogenic variant on molecular genetic testing; **AND**
- Patient had an insufficient response to a minimum 3-month trial of hydroxyurea (unless contraindicated or intolerant); **AND**
- Patient experienced one or more vaso-occlusive crises (VOC)* in the previous year despite adherence to hydroxyurea therapy

*VOC is defined as an event prompting either a visit or outreach to the provider which results in a diagnosis of VOC being made necessitating subsequent interventions such as narcotic pain management, non-steroidal anti-inflammatory therapy, hydration, etc.

RENEWAL CRITERIA

- Patient continues to meet the universal and other indication-specific relevant criteria identified in initial approval criteria; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include the following: severe infusion related reactions (e.g., ever, chills, nausea, vomiting, fatigue, dizziness, pruritus, urticaria, sweating, shortness of breath or wheezing), etc.; **AND**

- Disease response compared to pretreatment baseline as evidenced by a decrease in the frequency of vaso-occlusive crises (VOC) necessitating treatment, reduction in number or duration of hospitalizations, and/or reduction in severity of VOC

DOSAGE/ADMINISTRATION

INDICATION	DOSE
Sickle-cell Disease	Administer Adakveo 5 mg/kg by intravenous infusion over a period of 30 minutes at Week 0, Week 2, and every 4 weeks thereafter.

LENGTH OF AUTHORIZATION

Coverage will be provided for 6 months initially and may be renewed annually thereafter.

DOSAGE LIMITS

Max Units (per dose and over time) [HCPCS Unit]:

- 120 billable units at weeks 0 and 2 and every 4 weeks thereafter

APPLICABLE TENNESSEE STATE MANDATE REQUIREMENTS

BlueCross BlueShield of Tennessee's Medical Policy complies with Tennessee Code Annotated Section 56-7-2352 regarding coverage of off-label indications of Food and Drug Administration (FDA) approved drugs when the off-label use is recognized in one of the statutorily recognized standard reference compendia or in the published peer-reviewed medical literature.

IMPORTANT REMINDER

We develop Medical Policies to provide guidance to Members and Providers. This Medical Policy relates only to the services or supplies described in it. The existence of a Medical Policy is not an authorization, certification, explanation of benefits or a contract for the service (or supply) that is referenced in the Medical Policy. For a determination of the benefits that a Member is entitled to receive under his or her health plan, the Member's health plan must be reviewed. If there is a conflict between the Medical Policy and a health plan, the express terms of the health plan will govern.

ADDITIONAL INFORMATION

For appropriate chemotherapy regimens, dosage information, contraindications, precautions, warnings, and monitoring information, please refer to one of the standard reference compendia (e.g., the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) published by the National Comprehensive Cancer Network®, Drugdex Evaluations of Micromedex Solutions at Truven Health, or The American Hospital Formulary Service Drug Information).

SOURCES

1. Adakveo [package insert]. East Hanover, NJ; Novartis Pharmaceuticals, Inc., November 2019. Accessed November 2020.
2. Bender MA. Sickle Cell Disease. 2003 Sep 15 [Updated 2017 Aug 17]. In: Adam MP, Ardinger HH, Pagon RA, et al., editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2019. Available from: https://www.ncbi.nlm.nih.gov/books/NBK1377/.

3. Ataga KI, Kutlar A, Kanter J, et al. Crizanlizumab for the Prevention of Pain Crises in Sickle Cell Disease. N Engl J Med. 2017 Feb 2;376(5):429-439. doi: 10.1056/NEJMoa1611770. Epub 2016 Dec 3.
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. Lexicomp Online. (2020, February). AHFS DI. Crizanlizumab-tmca (Retrieved January 20, 2021 from Lexicomp Online with AHFS.
6. MICROMEDEX Healthcare Series. Drugdex Evaluations. (2019, December). Crizanlizumab-tmca. Retrieved January 20, 2021 from MICROMEDEX Healthcare Series.

EFFECTIVE DATE 6/2/2021

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