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Effective Date: 06/06/2019

Crizanlizumab™ (SEG101)

FDA approval: Anticipated 2020

HCPCS: N/A

Benefit: Medical

Policy:

Requests must be supported by submission of chart notes and patient specific documentation.

- A. Coverage of the requested drug is provided when all the following are met:
 - a. Diagnosis of sickle cell disease
 - b. Prescribed by or in consultation with a hematologist
 - c. FDA approved age
 - d. Patient has experienced 2 or more sickle cell-related crises in the past 12 months
 - e. If using hydroxyurea concurrently, patient must have been using the drug for at least 6 months and on a stable dose for the past 3 months
 - f. Must not be using long-term red blood cell transfusion therapy
- B. Quantity Limitations, Authorization Period and Renewal Criteria
 - a. Quantity Limit: FDA approved dosing
 - b. Initial Authorization Period: 6 months
 - c. Renewal Criteria: Patient has experienced a decrease in the number of sickle cell-related crises
 - d. Renewal Authorization Period: 1 year

***Note: Coverage may differ for Medicare Part B members based on any applicable criteria outlined in Local Coverage Determinations (LCD) or National Coverage Determinations (NCD) as determined by Center for Medicare and Medicaid Services (CMS). See the CMS website at <http://www.cms.hhs.gov/>. Determination of coverage of Part B drugs is based on medically accepted indications which have supported citations included or approved for inclusion determined by CMS approved compendia.

Therapeutic considerations:

- A. **FDA approved indication / Diagnosis**
 - a. The prevention of vaso-occlusive crises (VOC) in sickle cell disease (SCD) patients age 16 years and over

**Please refer to most recent prescribing information.*

https://www.accessdata.fda.gov/drugsatfda_docs/nda/2017/208587Orig1s000MedR.pdf

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B. Background Information

- a. Sickle cell disease is the most common inherited blood disorder in the United States affecting approximately 100,000 children and adults
- b. In the United States, SCD occurs in 1 in 365 African Americans and 1 in 16,300 Hispanics
- c. The disease involves multicellular adhesion between red blood cells, white blood cells, platelets, and endothelial cells resulting in vaso-occlusive crises
- d. VOC's are the hallmark of SCD and experienced by approximately 70% of patients
- e. They are recurring, unpredictable, painful events that decrease organ function with complications including stroke, pneumonia, vision loss, pulmonary hypertension, leg ulcers, and sepsis
- f. VOC's are often treated as an emergency requiring acute care and are the number one reason patients with SCD visit the emergency room or are hospitalized
- g. Hospital readmission rates are common with a >40% rehospitalization rate within 30 days
- h. Treatment guidelines recommend the use of opioids for the treatment of acute pain to due VOC
- i. To prevent VOC's, hydroxyurea is recommended for the following:
 - i. Those with 3 or more pain crises within the past year
 - ii. Patients experiencing pain that interferes with their daily activities or quality of life
 - iii. Patients with a history of recurrent or severe acute chest syndrome
 - iv. Patients with symptomatic chronic anemia
 - v. Children greater than 9 months of age regardless of symptoms
- j. Endari, another oral option for the prevention of VOC's, is not discussed in the current treatment guidelines

C. Efficacy

**Please refer to most recent prescribing information.*

D. Medication Safety Considerations

Black Box Warning: No

**Please refer to most recent prescribing information.*

E. Dosing and administration

- a. Dosing:
 - i. 5 mg/kg at day 1 and 15 then every 4 weeks thereafter

**Please refer to most recent prescribing information.*

F. How supplied

- a. Unknown at this time

References:

1. Ataga KI, Kutlar A, Kanter J, et al. Crizanlizumab for the prevention of pain crises in sickle cell disease. *New Engl J Med.* 2017; 376(5): 429 – 439.
2. Centers for Disease Control and Prevention. Sickle cell disease: data and statistics. Updated August 9, 2017. Available at: <https://www.cdc.gov/ncbddd/sicklecell/data.html>. Accessed on: May 7, 2019.
3. Ogedegbe HO. Sickle cell disease: an overview. *Laboratory Medicine.* 2002; 33(7): 515 – 543.
4. Yale SH, Nagib N, & Guthrie T. Approach to the vaso-occlusion crisis in adults with sickle cell disease. *Am Fam Physician.* 2000; 61(5): 1349 – 1356.
5. Centers for Disease Control and Prevntion. Sickle cell disease: complications and treatments. Updated August 9, 2017. Available at: <https://www.cdc.gov/ncbddd/sicklecell/treatments.html> Accessed on: May 7, 2019.

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6. Kanter J & Kruse-Jarres R. Management of sickle cell disease from childhood through adulthood. Blood Rev. 2013; 27(6): 279 – 287.
7. Brousseau DC, Panepinto JA, Nimmer M, et al. The number of people with sickle-cell disease in the United States: national and state estimates. Am J Hematol. 2010; 85(1): 77 – 78.
8. National Heart, Lung, and Blood Institute. Evidence-based management of sickle cell disease: expert panel report, 2014. Available at: <https://www.nhlbi.nih.gov/sites/default/files/publications/56-364NFULL.pdf>. Accessed on: May 7, 2019.

Policy History												
#	Date	Change Description										
1.0	Effective Date: 06/06/2019	Preliminary drug review <table><tr><th>Line of Business</th><th>PA Required (Yes/No)</th></tr><tr><td>BCBS</td><td>No</td></tr><tr><td>BCN</td><td>No</td></tr><tr><td>MAPPO</td><td>No</td></tr><tr><td>BCNA</td><td>No</td></tr></table>	Line of Business	PA Required (Yes/No)	BCBS	No	BCN	No	MAPPO	No	BCNA	No
Line of Business	PA Required (Yes/No)											
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BCNA	No											

* The prescribing information for a drug is subject to change. To ensure you are reading the most current information it is advised that you reference the most updated prescribing information by visiting the drug or manufacturer website or <http://dailymed.nlm.nih.gov/dailymed/index.cfm>.