VENTURA COUNTY HEALTH CARE PLAN

Utilization Review Policy

POLICY: Sickle Cell Disease – Adakveo® (crizanlizumab-tmca injection, for intravenous use)

DATE REVIEWED: 11/20/2019

OVERVIEW

Adakveo, a monoclonal antibody, is indicated to reduce the frequency of vasocclusive crises in adults and pediatric patients aged 16 years and older with sickle cell disease.[1]

Disease Overview

Sickle cell disease, a multisystem disorder, is the most common condition caused by a single gene mutation.[2] In the US, population estimates suggest that a total of 100,000 persons have the disease. Approximately 300,000 babies are born with sickle cell anemia each year and it is estimated that the number could be as high as 400,000 by 2050.

Sickle cell disease is characterized by the presence of abnormal erythrocytes damaged by the sickle hemoglobin gene – this variant of the normal adult hemoglobin can be inherited from both parents or from one parent along with another variant, such as hemoglobin C or with β-thalassemia.[2] Complications of sickle cell disease include vaso-occlusion (which can result in pain and organ failure), hemolytic anemia, and large-vessel vasculopathy (cerebrovascular disease, pulmonary hypertension, ischemic organ damage, hyposplenism, renal failure, bone disease, liver failure).

Guidelines

Adakveo has not been added to guidelines. The National Institutes of Health – National Heart, Lung, and Blood Institute issued the Evidence-Based Management of Sickle cell disease, Expert Panel Report in 2014.[3] There are two effective disease-modifying therapies for sickle cell disease: hydroxyurea and chronic blood transfusions. Hydroxyurea has been shown to reduce: the frequency of painful episodes, incidence of acute coronary syndrome events, and the need for transfusions and hospitalizations.

POLICY STATEMENT

Prior authorization is recommended for medical benefit coverage of Adakveo. Approval is recommended for those who meet the Criteria and Dosing for the listed indication(s). Requests for doses outside of the established dosing documented in this policy will be considered on a case-by-case basis by a clinician (i.e., Medical Director or Pharmacist). All approvals are provided for the duration noted below.

Because of the specialized skills required for evaluation and diagnosis of patients treated with Adakveo as well as the monitoring required for adverse events and long-term efficacy, approval requires Adakveo to be prescribed by, or in consultation with, a physician who specializes in the condition being treated.

RECOMMENDED AUTHORIZATION CRITERIA

Coverage of Adakveo is recommended in those who meet the following criteria:

FDA-Approved Indications

1. **Sickle Cell Disease.** Approve for 1 year if the patient meets the following criteria (A and B):
   - **A)** The patient is ≥ 16 years of age; AND
   - **B)** Adakveo is prescribed by, or in consultation with, a physician who specializes in the treatment of sickle cell disease (e.g., a hematologist).

   Dosing. Approve up to 5 mg/kg by intravenous infusion at Weeks 0 and 2, and then not more frequently than once every 4 weeks.

CONDITIONS NOT RECOMMENDED FOR APPROVAL

Adakveo has not been shown to be effective, or there are limited or preliminary data or potential safety concerns that are not supportive of general approval for the following conditions. Rationale for non-coverage for these specific conditions is provided below. (Note: This is not an exhaustive list of Conditions Not Recommended for Approval.)

1. Coverage is not recommended for circumstances not listed in the Recommended Authorization Criteria. Criteria will be updated as new published data are available.

REFERENCES

1. Adakveo® injection for intravenous use [prescribing information]. East Hanover, NJ: Novartis; November 2019.
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 2014. Available at: http://www.nhlbi.nih.gov/guidelines. Accessed on November 18, 2019.

HISTORY

Type of Revision	Summary of Changes	Date Reviewed
New Policy	--	11/20/2019
Update	Changed policy name from "Hematology – Adakveo" to "Sickle Cell Disease – Adakveo"	12/04/2019