PRIOR AUTHORIZATION POLICY

POLICY: Sickle Cell Disease – Endari Prior Authorization Policy
- Endari™ (L-glutamine oral powder – Emmaus Medical)

REVIEW DATE: 01/03/2024

OVERVIEW

Endari is indicated to **reduce the acute complications of sickle cell disease** in patients ≥ 5 years of age.[1]

L-glutamine is an essential amino acid and serves as a precursor of nucleic acids and nucleotides including the pyridine nucleotides (nicotinamide adenine dinucleotide and reduced nicotinamide adenine dinucleotide).[1,2] These pyridine nucleotides play key roles in the regulation and prevention of oxidative damage in red blood cells and studies have shown that oxidative phenomena may play a significant role in the pathophysiology of sickle cell disease.

Guidelines

The American Society of Hematology guidelines for sickle cell disease: management of acute and chronic pain associated with sickle cell disease (2020) does address Endari's place in therapy.[2] 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] The use of L-glutamine products in sickle cell disease is not mentioned (guidelines were published before the approval of Endari). Hydroxyurea has been shown to reduce the frequency of painful episodes, the incidence of acute coronary syndrome events, and the need for transfusions and hospitalizations.

POLICY STATEMENT

Prior Authorization is recommended for prescription benefit coverage of Endari. All approvals are provided for the duration noted below. Because of the specialized skills required for evaluation and diagnosis of patients treated with Endari as well as the monitoring required for adverse events and long-term efficacy, approval requires Endari to be prescribed by or in consultation with a physician who specializes in the condition being treated.

Documentation: Documentation is required for use of Endari as noted in the criteria as **[documentation required]**. Documentation may include, but is not limited to, chart notes, prescription claims records, prescription receipts, and/or other information.

Automation: None.

RECOMMENDED AUTHORIZATION CRITERIA

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

FDA-Approved Indication

1. **Sickle Cell Disease [documentation required].** Approve for 1 year if the patient meets the following criteria (A and B):
   A) Patient is ≥ 5 years of age; AND
   B) The medication is prescribed by or in consultation with a physician who specializes in sickle cell disease (e.g., a hematologist).

CONDITIONS NOT RECOMMENDED FOR APPROVAL

Coverage of Endari is not recommended in the following situations:

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. Endari™ oral powder [prescribing information]. Torrance CA: Emmaus Medical; October 2020.
2. Brandow AM, Carroll CP, Creary S, et al. American Society of Hematology 2020 guidelines for sickle cell disease: management of acute and chronic pain. *Blood Adv*. 2020;4:2656-2701.
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: https://www.nhlbi.nih.gov/sites/default/files/media/docs/sickle-cell-disease-report%20020816_0.pdf. Accessed on December 5, 2023.