

**Pharmacy Prior Authorization
Clinical Guidelines - Endari**

Authorization Guidelines:

May be authorized when all the following criteria are met:

- Diagnosis is for Sickle Cell Disease
- Request is to reduce the acute complications experienced from Sickle Cell Disease
- Member is 5 years of age or older
- There was a previous trial and failure, intolerance, or a contraindication to hydroxyurea
- Endari will be used concurrently with hydroxyurea
- All other indications are considered experimental/investigational and not medically necessary

Initial Approval:

12 months

Renewal Approval:

12 months

Requires:

Member experienced a reduction in acute complications of sickle cell disease (For example, reduction in number of sickle cell crises, acute chest syndrome episodes, fever, occurrences of priapism, splenic sequestration)

References:

1. National Institutes of Health (NIH): National Heart, Lung, and Blood Institute (NHLBI). Evidence-Based Management of Sickle Cell Disease: Expert Panel Report, 2014. https://www.nhlbi.nih.gov/sites/default/files/media/docs/sickle-cell-disease-report%20020816_0.pdf. Accessed May 3, 2021.
2. Vichinsky, E.P. (2020). Disease-modifying therapies for prevention of vaso-occlusive pain in sickle cell disease. In M. R. DeBaun (Ed.), *UpToDate*. Retrieved May 3, 2021 from: <https://www.uptodate.com/contents/disease-modifying-therapies-for-prevention-of-vaso-occlusive-pain-in-sickle-cell-disease>.
3. Endari [package insert]. Torrance, CA: Emmaus Medical, Inc; April 2020.
4. Niihara Y, Miller ST, et al. A phase 3 trial of l-glutamine in sickle cell disease. *N Engl J Med*. 2018;379(3):226-235