BlueCross BlueShield Federal Employee Program

# SIKLOS (hydroxyurea)

## RATIONALE FOR INCLUSION IN PA PROGRAM

### Background

Sickle cell anemia is a genetically inherited condition which causes red blood cells to become sickle shaped. These “sickled” red blood cells can cause many complications including infections, hypertension, renal disease, stroke, retinopathy, and other chronic conditions. Painful crises are an acute condition caused by chronic sickle cell disease that causes much distress to people with sickle cell anemia. Hydroxyurea has been used to prevent these acute painful crises from occurring, as well as to improve survival and reduce other complications. Siklos is a new formulation of hydroxyurea specifically designed for use in sickle cell patients to decrease the need for blood transfusions as well as decrease the frequency of painful crises (1-2).

### Regulatory Status

FDA-approved indication: Siklos is indicated to reduce the frequency of painful crises and to reduce the need for blood transfusions in adult and pediatric patients, 2 years of age and older, with sickle cell anemia with recurrent moderate to severe painful crises (2).

Siklos carries boxed warnings for myelosuppression and malignancies. Due to the risk of myelosuppression, blood counts should be monitored every 2 weeks throughout the duration of therapy. Hydroxyurea is a human carcinogen and secondary leukemia has been reported in patients receiving long-term. Skin cancer has also been reported in patients receiving long-term hydroxyurea. Advise protection from sun exposure and monitor for the development of secondary malignancies (2).

Initiation of therapy is started at 20 mg/kg/day and is based on the patient’s actual or ideal body weight, whatever is less. Dosing adjustments are made based on blood counts, and therefore it is imperative to monitor patients’ CBCs during therapy (2).

Siklos can cause fetal harm when administered to pregnant women. Verify the pregnancy status of females of reproductive potential prior to initiating Siklos therapy. Females and males with partners of reproductive potential should be advised to use effective contraception during and after treatment with Siklos for at least 6 months after therapy (2).

Safety and effectiveness in pediatric patients 2 years of age and older have been established (2).

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## Summary

Painful crises are an acute condition caused by chronic sickle cell disease that causes much distress to people with sickle cell anemia. Hydroxyurea has been used to prevent these acute painful crises from occurring, as well as to improve survival and reduce other complications. Siklos is indicated to reduce the frequency of painful crises and to reduce the need for blood transfusions patients with sickle cell anemia with recurrent moderate to severe painful crises (1-2).

Prior authorization is required to ensure the safe, clinically appropriate, and cost-effective use of Siklos while maintaining optimal therapeutic outcomes.


## References


1. National Heart, Lung, and Blood Institute (NHLBI): Evidence-Based Management of Sickle Cell Disease. Expert Panel Report, 2014. Published by U.S. Department of Health and Human Services, National Institutes of Health. https://www.nhlbi.nih.gov/sites/default/files/media/docs/sickle-cell-disease-report%20020816_0.pdf
2. Siklos [package insert]. Bryn Mawr, PA: Medunik USA Inc.; November 2023.