Scope: Hospital-wide Patient Care
Document Type: Clinical Practice Guideline
Approved on: 2025-08-11
Next Review Date: 2027-08-11

Priapism: Guidelines for Management in Children with Sickle Cell Anemia

Version: 3

1.0 Background

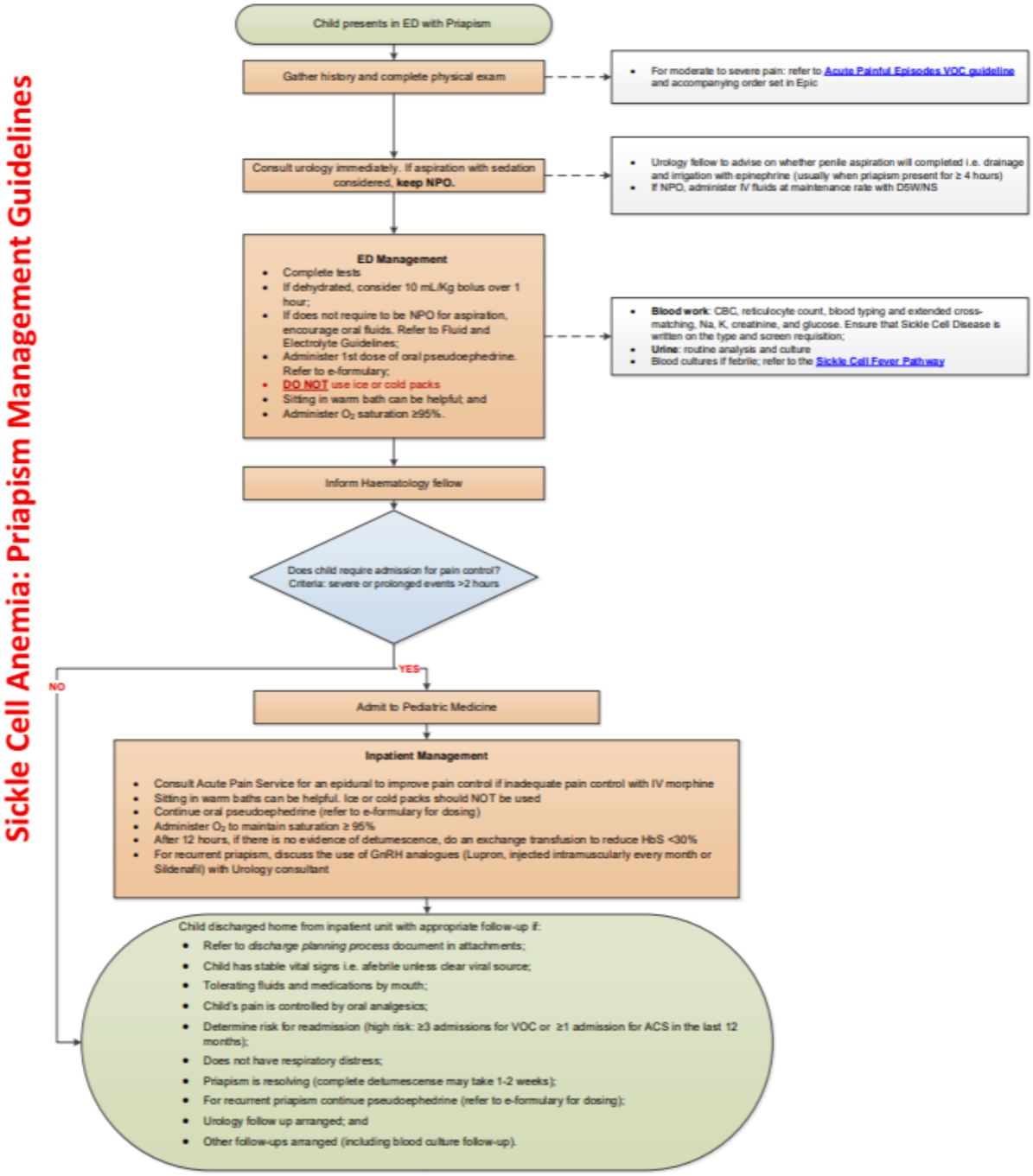
Priapism, a painful, prolonged (>30 minutes) erection of the penis, has been found to occur at least once in 75% of males with sickle cell anaemia before the age of 21 years. There are two bimodal peaks (ages 5–13 and 21–29y). In a majority of cases, priapism occurs during early-morning sleep and awakens the patient. Sexual activity is a precipitating event in approximately 20% of cases; however, in 40% of cases no provoking event is identified. Priapism occurs in two forms: stuttering, which lasts 2h or less, and severe, which lasts more than 2h and may result in impotence.

The pathophysiology of priapism in sickle cell disease remains elusive. It is thought that the relative stasis of blood within the corpora during normal erection decreases the oxygen tension and pH, both circumstances conducive to sickling. Sickling of RBCs and sludging of blood within the corpora leads to further hypoxia and acidosis, which in turn promotes further sickling.

Eventually an inflammatory response is elicited, resulting in fibrosis; it is this that ultimately is responsible for impotence. Priapism is more common in patients with SS disease but can also occur in those with SC disease or Sß thalassemia.

The best treatment for priapism in patients with sickle cell disease is not known. Analgesia and hydration are of benefit, but the role of transfusion (if any) or surgery is not clear. Positive prognostic predictors in priapism include being prepubertal, early presentation, and early treatment. In managing priapism, one should distinguish stuttering priapism from the severe form. In stuttering priapism, no specific intervention is required for a single episode: Simple treatments such as hydration, warm baths, and analgesics are usually sufficient to end it. Severe or prolonged events (>2h) are to be considered emergencies requiring prompt medical intervention.

2.0 Clinical Practice Guideline



PRINTABLE VERSION

3.0 References

1. Burnett et al. Randomized controlled trial of sildenafil for preventing recurrent ischemic priapism in sickle cell disease. *Am J Med*. 2014 Jul;127(7):664-8.
2. Hamre MR, Harmon EP, Kirkpatrick DV, et al. Priapism as a complication of sickle cell disease. *J Urol*. 1991;145:1-5.
3. Powars DR, Johnson CS. Priapism. *Hematol Oncol Clin North Am*. 1996;10:1363–72.
4. Sharpstein JR, Powars D, Johnson C, et al. Multisystem damage associated with tricorporal priapism in sickle cell disease. *Am J Med*. 1993;94:289–95 [a prospective 25-year cohort study of 461 men with sickle cell, of whom 38 developed clinically significant priapism].

Attachments:

Priapism Care Pathway FINAL.pdf

Revision History.docx

SC_Clinic Follow Up Revised FINAL.pdf