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 Sβ 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

Child presents in ED with Priapism

- Gather history and complete physical exam.

Consult urology immediately if aspiration with sedation considered; keep NPO.

ED Management
- Complete tests:
  - CBC, reticulocyte count, blood typing and extended cross matching; No, K, creatinine, and glucose. Ensure that Sickle Cell Disease is written on the type and screen requisition.
  - Urine: routine analysis and culture.
  - Blood cultures if febrile.
- Blood work: CBC, reticulocyte count, blood typing and extended cross matching; No, K, creatinine, and glucose. Ensure that Sickle Cell Disease is written on the type and screen requisition.
- Electrolyte Guidelines:
  - Administer 1st dose of oral pseudoephedrine; refer to a-formulary; DO NOT use.
  - Sitting in warm bath can be helpful; and
  - Administration O2 saturation ≥95%.

Inform Haematology fellow.

Does child require admission for pain control?
- Child, severe or prolonged event ≥2 hours.

Admit to Pediatric Medicine

Inpatient Management
- Consult Acute Pain Service for an epidural to improve pain control; if inadequate pain control with IV morphine.
- Sitting in warm bath can be helpful; use or cool packs should NOT be used.
- Administer O2 to maintain saturation ≥95%.
- Red blood cell transfusion: if there is no evidence of detumescence; refer to e-formulary for dosing.
- If dehydrated, consider 10 mL/kg bolus over 1 hour.
- After 12 hours, if there is no evidence of detumescence, do an exchange transfusion to reduce HbS ≥30%.
- Refer to Urology fellow to advise on whether penile aspiration will be completed; i.e. drainage and irrigation with epinephrine (usually when priapism present for ≥4 hours).
- If NPO, administer IV fluid at maintenance rate with GS/NS.
- Blood work: CBC, reticulocyte count, blood typing and extended cross matching; No, K, creatinine, and glucose. Ensure that Sickle Cell Disease is written on the type and screen requisition.
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NO

Admit to Pediatric Medicine

- Refer to discharge planning process document.
- Child has stable vital signs i.e. stable unless clear oral source.
- Tolerating fluids and medications by mouth;
- Child's pain is controlled by oral analgesia;
- Determine risk for readmission (high risk: ≥3 admissions for VOC or ≥1 admission for ACS in the last 12 months);
- Does not have respiratory distress;
- Priapism is resolving (complete detumescence may take 1-2 weeks);
- For recurrent priapism continue pseudoephedrine (refer to a-formulary for dosing);
- Urology follow up arranged; and
- Other follow-ups arranged (including blood culture follow-up).

YES

Admit to Pediatric Medicine

- For moderate to severe pain: refer to Acute Pain Episode VOC guideline and accompanying order set in Epic.
- Urology fellow to advise on whether penile aspiration will be completed; i.e. drainage and irrigation with epinephrine (usually when priapism present for ≥4 hours).
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Inpatient Management
- Consult Acute Pain Service for an epidural to improve pain control; if inadequate pain control with IV morphine.
- Sitting in warm bath can be helpful; use or cool packs should NOT be used.
- Administer O2 to maintain saturation ≥95%.
- Red blood cell transfusion: if there is no evidence of detumescence; refer to e-formulary for dosing.
- If dehydrated, consider 10 mL/kg bolus over 1 hour.
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- Refer to Urology fellow to advise on whether penile aspiration will be completed; i.e. drainage and irrigation with epinephrine (usually when priapism present for ≥4 hours).
- If NPO, administer IV fluid at maintenance rate with GS/NS.
- Blood work: CBC, reticulocyte count, blood typing and extended cross matching; No, K, creatinine, and glucose. Ensure that Sickle Cell Disease is written on the type and screen requisition.
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3.0 References


**Attachments:**

- [Priapism Care Pathway 2021 FINAL.pdf](#)
- [Revision History.docx](#)
- [SC_Clinic Follow Up Revised 2021_FINAL.pdf](#)