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.
Priapism: Guidelines for Management in Children with Sickle Cell Anemia

Child presents in ED with Priapism

Gather history and complete physical exam.

Consult urology immediately if insertion with sedation was considered, keep NPO.

ED Management
- Complete labs:
  - If dehydrated, consider 10 mL/kg bolus over 1 hour.
  - If does not require to be NPO for aspiration, encourage oral fluids. Refer to Fluid and Electrolyte Guidelines:
    - Administer 1st dose of oral pseudoephedrine. Refer to e-formulary;
    - Ice or cold packs should NOT be used
    - Sitting in warm bath can be helpful; and
    - Administer O2 saturation ≥95%.

Inform Haematology fellow

Does child require admission for pain control? Criteria: severe or prolonged events ≥12 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 baths can be helpful. Ice or cold packs should NOT be used
- Encourage oral fluids. Refer to Fluid and Electrolyte Guidelines:
  - Administer O2 saturation ≥95%.
- Administer O2 concentration ≥35%.

If does not require to be NPO for aspiration:
- Administer Intravenous fluids at maintenance rate with D5W
- Blood work: CBC, reticulocyte count, blood typing and extended cross matching, Na, K, creatinine, and glucose. Ensure that Sickle Cell Disease is confirmed. Patient must be subject to Acute Painful Episodes VOC guideline
- Discharge planning process document:
  - Urology follow up arranged.
  - Other follow ups arranged (including blood culture follow up).

Child discharged from inpatient unit with appropriate follow up if:
- Refer to discharge planning process document;
- Child has stable vital signs i.e. stable urine output and source;
- Tolerating fluids and medications by mouth;
- Child’s pain is controlled by oral analgesics;
- 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 continuous pseudoephedrine (refer to e-formulary for dosing);
- Urology follow up arranged; and
- Other follow-ups arranged (including blood culture follow up).
References


Reviewers

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Attachments:

- priapism_pathway_final.pdf
- Revision History.docx
- SC_clinic follow up.pdf