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 NDPO.
- ED Management:
  - Complete tests.
  - If dehydrated, consider 10 mL/kg bolus over 1 hour.
  - If does not require to be NDPO for aspiration, encourage oral fluids. Refer to Fluid and Electrolyte Guidelines.
  - Administer 1st dose of oral pseudoephedrine. Refer to e-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?
- CBC, serum and or-prolonged event >3 hours

Admit to Pediatric Medicine

Inpatient Management:
- Consult Acute Pain Service for an epidural to improve pain control if IV morphine.
- Sitting in warm bath can be helpful if or cold/packs should NOT be used.
- Continue oral pseudoephedrine (refer to e-formulary for dosing).
- Administer O₂ to maintain saturation ≥95%
- After 12 hours, if there is no evidence of detumescence, do an exchange transfusion to reduce HbS <30%
- For recurrent priapism consider use of GnRH analogues (Lupron, injected intramuscularly every month or Sildenafil) with Urology consultant.

Child discharged home from inpatient unit with appropriate follow-up if:
- 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 e-formulary for dosing);
- Urology follow up arranged; and
- Other follow-ups arranged (including blood culture follow-up).

3.0 References


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Attachments:

*Priapism Care Pathway 2021 FINAL.pdf*

*Revision History.docx*

*SC_Clinic Follow Up Revised 2021_FINAL.pdf*