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Adult Guideline: Management of Priapism in adults with Sickle Cell Disease

NOTE
This guideline is not intended to be construed or to serve as a standard of care. Standards of care are determined on the basis of all clinical data available for an individual case and are subject to change as scientific knowledge and technology advance and patterns of care evolve. Adherence to guideline recommendations will not ensure a successful outcome in every case, nor should they be construed as including all proper methods of care or excluding other acceptable methods of care aimed at the same results. The ultimate judgement must be made by the appropriate healthcare professional(s) responsible for clinical decisions regarding a particular clinical procedure or treatment plan. This judgement should only be arrived at following discussion of the options with the patient, covering the diagnostic and treatment choices available. It is advised, however, that significant departures from the national guideline or any local guidelines derived from it should be fully documented in the patient's case notes at the time the relevant decision is taken.

Introduction

Priapism is defined as a pathologically prolonged erection (>4 hours) in the absence of sexual stimulation. Priapism has been divided into three main categories: ischemic, non-ischemic and stuttering priapism. Stuttering priapism, also termed intermittent or recurrent priapism, is characterized by recurring episodes of ischemic priapism, though relatively rare, affects a high prevalence of men with sickle-cell disease (SCD). Chronic stuttering episodes may last a few minutes but by definition less than four hours and can herald a fulminant episode. Fulminant episodes last more than four hours and are a medical emergency; without prompt treatment erectile dysfunction and penile scarring may occur. ***The longer the duration, the greater the chance of a poor outcome.***

Patients with sickling disorders are at particular risk of priapism with an annual incidence in adults of 29-42%. Children may also be affected albeit less commonly. Up to 90% of patients will have had an episode by the age of 20. Priapism may develop with any sickling disorder but the risk appears highest in HbSS.

95% of cases in sickle patients are ischaemic. Due to dysregulation of penile circulation, blood becomes trapped at increased pressure within the corpus cavernosum. This creates a hypoxic, hypercapneoic, acidotic environment analogous to a compartment syndrome. If left untreated permanent tissue damage will occur. Pathophysiology is poorly understood but abnormal blood rheology and alterations in NO metabolism may contribute to the high incidence in sickle patients.

Patient education

Priapism is often under reported. Male sickle patients should be educated about the dangers of priapism at their first visit and given written information. **They should be instructed to present to hospital immediately if the priapism does not resolve within two hours.**

The following supportive measures can be started whilst the patient is at home **but patients must be aware not to delay seeking help:**

- Plentiful oral hydration
- Pain relief (as per patient's usual for crisis pain)
- Advise the patient to empty their bladder
- Encourage ejaculation
- Encourage gentle exercise if able ('steal syndrome' may help resolve stuttering episodes)
- A warm shower or bath may be helpful but evidence is lacking
- **Ice/cold packs must not be applied**

Principles of management

Patients with SCD should be managed jointly with the haematology team.

Inform Urology Registrar on-call immediately if a patient presents with suspected priapism. Do not delay whilst initial supportive measures are being carried out. Prompt surgical intervention may be required.

Diagnosis

Focused history

- **Duration/time of onset must be documented**
- Pain severity (fulminant priapism usually very painful)
- Medication:
  - intracavernosal drugs, sildenafil, antihypertensives, antipsychotics and anticoagulants are associated with development of priapism
  - analgesia taken prior to arrival
- Recreational drugs: alcohol, marijuana, cocaine
- Prior episodes
- Pre-existing erectile dysfunction
- History of genital trauma
- Crisis pain elsewhere

Examination

- Brief general examination
- Full set of observations
- External genitalia
  - Degree of tumescence
  - Abdominal examination: organomegaly, masses etc

Initial management

- Opiate analgesia as required (sedation may be needed if severe)
- IV fluids
- Supplemental oxygen should be considered in all patients (mandatory if saturations <94% on air)
- Catheterise only if bladder palpable or unable to void
- Encourage gentle exercise if patient systemically well

Initial investigations

- FBC, reticulocyte count, film
- Haemoglobin electrophoresis and sickle solubility test (if not known to unit)
- Group and Save (in case needs exchange transfusion, blood loss usually minimal with shunt procedures)
- Electrolytes, CRP
- Consider toxicology screen

Further management

If the priapism has been present for ***less than two hours*** a trial of an oral alpha or beta adrenergic agonist ***may be appropriate*:**

- Etilifrine 50mg po stat, Ephedrine 15-30mg po stat or Terbutaline 5mg po stat depending on local policy and availability.
- If used monitor for hypertension and/or tachycardia.
- Contraindicated in patients with uncontrolled hypertension or cardiac disease.
- If possible, patient should be made aware that these are *unlicensed* indications.
- If priapism settles with the above, consider overnight admission to urology for monitoring.

If detumescence does not occur within 30 minutes of oral therapy, or duration of priapism greater than four hours, inform on-call Urologist immediately, as penile aspiration and/or irrigation of the corpus cavernosum will be required.

- Penile aspiration:
  - **This should be performed by a Urologist but if this is not possible in an emergency situation Medical staff may aspirate if competent to do so**
  - Clean the penis with antiseptic solution as per local policy.
  - Using a 19G butterfly needle and a heparinised 20ml syringe, blood should be aspirated from the corpus cavernosum with a lateral approach (10 or 2 o'clock position), taking care to avoid the dorsal vein (superior aspect) and the urethra (inferior aspect). This will often provide immediate relief.
  - **Aspirated blood must be sent for blood gas analysis to confirm ischaemic priapism (pO2 <30mmHg and pCO2 >60mmHg, pH <7.25 expected in ischaemic priapism)**
  - 50-500mls should be aspirated until detumescence is achieved and the cannula then flushed with saline.
  - Leave cannula in situ for thirty minutes before removing.

- If still no resolution, intracavernosal irrigation with normal saline can be performed
- If no resolution, intracavernosal injection of adrenoreceptor agonist can be performed in combination with aspiration and irrigation. (ECG and continuous BP monitoring is required).
- Intracavernous injection of phenylephrine is recommended and given in 200 µg doses every 10 minutes. Maximum dosage is 1 mg within 1 hour. This may cause transient hypertension. The lower doses are recommended in children and patients with severe cardiovascular disease.
- If this fails, the patient may require a penile shunt procedure.
- Late presentations with a duration of >72 hours where the risk of total loss of sexual function and penile scarring is high may benefit from an immediate penile implant procedure.

Should the patient require surgery, the need for a top-up or partial exchange transfusion should be considered as per the peri-operative guideline, but beware the association of sickle cell disease with priapism, exchange transfusion and neurological events (ASPEN syndrome). *Care should be taken to avoid an excessively high post-transfusion haemoglobin.*

THE DECISION TO TRANSFUSE SHOULD BE MADE BY A CONSULTANT HAEMATOLOGIST.

THERE IS LIMITED EVIDENCE FOR THE USE OF TRANSFUSION AS AN AID TO DETUMESCENCE.

Secondary Prevention

Advice

- Avoid alcohol at night
- Awareness of the link between illicit drugs and priapism (cocaine, marijuana)
- Maintain good fluid intake
- Empty bladder before bed
- Inform doctor about stuttering episodes

Outpatient management of stuttering priapism

Management should be in conjunction with a Sexual Health specialist with an interest in priapism.

Psychosocial aspects should also be considered.

Pharmacological therapy should be reserved for patients with recurrent episodes or who have previously suffered a fulminant priapism.

Options include:

- Regular oral alpha adrenergic agonist e.g. Etilephrine 50 -100 mg daily- monitor BP every 2 weeks and discontinue if > 150/90
- Hormonal therapies- side effects include loss of libido and impotence
- Trial of PDE5 inhibitor e.g. Sildenafil. (Treatment should be initiated only when the penis is in its flaccid state)
- Hydroxycarbamide or regular transfusion programme if previous severe episodes - discuss at SPAH MCN MDT prior to commencement

References

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