Joint Paediatric and Urology Clinical Guidelines for the management of Children with Priapism and Sickle Cell Disease

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<tbody>
<tr>
<td>Authors:</td>
<td>Dr David Rees, Dr Susan Height, Dr Moira Dick</td>
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<tr>
<td>Responsible committee or Director:</td>
<td>Child Health</td>
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For Clinical Guidelines Groups’ use only

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<tr>
<th>Ratified by:</th>
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<tr>
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JOINT PAEDIATRIC AND UROLOGY CLINICAL GUIDELINES FOR THE MANAGEMENT OF CHILDREN WITH PRIAPISM AND SICKLE CELL DISEASE

This guideline is relevant to all children in King’s College Hospital with Sickle Cell Disease and priapism. The guideline applies to all patients with sickle cell disease who are currently under the care of the Paediatric Haematology team. It is mainly aimed at being a tool for the medical team managing these patients, but any member of the multidisciplinary team may find it useful.

DEFINITIONS

Priapism is a sustained, painful, and unwanted erection. It is classified into a) prolonged if it lasts more than three hours or b) stuttering if it lasts for more than a few minutes but less than three hours; stuttering episodes may recur and/or develop into more prolonged episodes. Priapism is common in sickle cell disease and as many as 90 percent of males with SCD will have experienced one or more episodes of priapism by the age of 20 years. Priapism in SCD is due to vaso-occlusion, which causes obstruction of the venous drainage of the penis. Prolonged priapism is an emergency that requires urologic intervention.

PSYCHOSOCIAL AND COUNSELING ASPECTS OF PRIAPISM

In early childhood, males need to know that priapism is one aspect of SCD and that they should tell their parents or other appropriate adult if it occurs. Parents and children should be given the information leaflet on Priapism in the first year of life and again age 11-12 years old. If untreated, priapism can result in impotence. It can be triggered by a number of factors such as full bladder and sexual activity. Recurrence can be prevented by the use of medication.

EVALUATION AND TREATMENT

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<th>PROLONGED PRIAPISM IS AN EMERGENCY AND REQUIRES URGENT ASSESSMENT AND TREATMENT.</th>
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<td>• Document the time of onset of the episode.</td>
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<td>• Precipitating factors, such as trauma, infections, or the use of drugs (e.g., alcohol, psychotropic agents, sildenafil, testosterone, cocaine).</td>
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<td>• A careful physical examination should reveal a hard penis with a soft glans.</td>
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The aim of therapy is to relieve pain, abort the erection and preserve future erectile function.
EPISODES LASTING LESS THAN 2 HOURS

- Patients should be advised to drink extra fluids, use oral analgesics, and attempt to urinate as soon as priapism begins. Walking and warm baths may also help to avert early priapism.

EPISODES LASTING MORE THAN TWO HOURS

- Patient should be referred directly to the Paediatric Urology Team (02071884610) at Evelina Children’s Hospital urgently.

- While waiting to transfer to Evelina Hospital, the patient should be given intravenous fluids and adequate analgesia. The patient should be kept nil-by-mouth.

- Blood transfusion will not normally be necessary before any surgical procedure, but may be appropriate in patients with a history of chest crises, or post-anaesthetic problems.

- The initial treatment is usually penile aspiration and irrigation with an α adreno-receptor agonist. If this does not relieve the priapism, shunting procedures may be necessary. This plan is determined by the Urologist.

- If priapism recurs and further surgery required, an exchange blood transfusion may be necessary before the second anaesthetic, if this has not taken place already.

- Complications of priapism and treatment include bleeding from the holes placed in the penis as part of the aspiration or shunting procedures, infections, skin necrosis, damage or strictures of the urethra, fistulae, and impotence. If impotence persists for 12 months, the patient may be referred to the andrology team at UCH for consideration for implantation of a semi rigid penile prosthesis. Patients should be given a follow-up appointment with Kalpana Patil or Arash Taghizadeh Arash in the paediatric urology clinic at Evelina Hospital, following discharge.

FUTURE PREVENTION

Etilefrine is first-line treatment and usually effective.

<2 years: 1mg-2.5mg 3 times a day
2-6 years: 2.5mg-5mg 3 times a day
>6 years: 5mg-10mg 3 times a day
If further episodes occur despite etilefrine, other options include:

Leuprolein acetate (3.75mg every four weeks), a gonadotropin-releasing hormone analogue that suppresses the hypothalamic-testicular axis and the production of testosterone, used with some degree of success.

Oral stilbestrol in doses of 5 mg daily for 3 to 4 days could abort episodes of priapism and smaller doses can be used to prevent recurrence.

PATIENT PATHWAY FOR PRIAPISM LASTING MORE THAN 2 HOURS

- Contact Paediatric Urology team at Evelina Hospital. Paediatric urology registrar can be contacted via the Evelina Hospital switchboard Monday-Friday, 9am – 5pm. Outside of these hours; the on-call paediatric urology consultant can be contacted through Evelina Sky level 02071884611.

- The patient will be admitted under Paediatric Urology team, and will also be assessed by the paediatric registrar at Evelina Hospital (02071887188 Bleep 0339), who will contact Dr Baba Inusa (consultant paediatrician) as appropriate (02071887774) through St Thomas’s switchboard) (or Dr Sue Height or Dr David Rees through King’s College Hospital switchboard if he is not available).

- The patient should be started on IV fluids and analgesia with the aim of urgent surgical intervention if the priapism has persisted for more than 3 hours. Surgical Intervention should not be delayed by either medical treatment or the provision of blood products, except in case of repeated, major surgical procedure.

Dr David Rees
Dr Moira Dick
Dr Sue Height

December 2009