

## PRIAPISM

### A COMPLICATION OF SICKLE CELL DISEASE

**P**riapism<sup>1</sup>, sustained and painful erection, is a vaso-occlusive complication of sickle cell disease. The prevalence of priapism in young males with sickle cell anemia or sickle beta-thalassemia may be as high as 89%<sup>2</sup>. Potentially serious consequences of prolonged and untreated priapism are penile fibrosis and impotence. Two clinical patterns have been described: Severe episodes that last more than 2 to 3 hrs. requiring medical intervention, and shorter episodes called stuttering spells lasting from a few minutes to 2 hrs. which resolve spontaneously.

Mantadakis et al<sup>3</sup> recently reported on a procedure of penile aspiration and epinephrine irrigation to treat priapism in 15 patients with sickle cell anemia ranging in age from 3.9 to 18.3 yrs. After conscious sedation of the patient and infiltration of lidocaine, a needle is inserted into the corpus cavernosum and blood aspirated into a syringe. A 1:1million solution of epinephrine is then introduced to irrigate the corpora cavernosa. Additional blood is aspirated until detumescence occurs.

After needle withdrawal, firm pressure is applied for 5 min. to prevent hematoma formation.

The 15 patients were treated on 39 occasions (ten patients once, one patient twice, two patients 3 times, one patient 6 times and one patient 15 times) and detumescence was successful on 37 of the 39 occasions. No serious side effects were observed.

Anticipatory guidance regarding the signs and symptoms of priapism should begin during the early school age years.

Early recognition and initiation of treatment can prevent serious consequences of priapism.

### A Recent Case at Strong

**A**n eight-year-old with hemoglobin SC disease was referred to emergency secondary to symptoms of priapism. He reported having a painful erect penis, but upon exam the penis was flaccid. He was discharged with instructions to the family to increase oral hydration, utilize a warm bath/shower and initiate oral pain medication should the priapism return.

Several days later, his mother called to report that her son was experiencing another episode of priapism. Despite following the home management instructions, he was in extreme pain, without any resolution in the erection. Initial treatment in the Pediatric Sickle Cell Clinic consisted of IV hydration and IV pain control.

Pediatric urology was consulted when detumescence was not achieved after 4 hours of treatment. Dr. Robert Merovach performed a penile aspiration and epinephrine irrigation procedure in the Pediatric Treatment Center. Approximately 50% detumescence was achieved. The patient was admitted to the hospital for continued IV hydration and pain management. After an eight-day hospitalization, which was complicated by some generalized vaso-occlusive crisis pain, total detumescence was achieved.

This recent experience with a young patient with priapism underscores the importance of early anticipatory guidance with regard to priapism, as well as early initiation of treatment. As described by Mantadakis et al, early activation of penile aspiration and epinephrine irrigation, usually within 4 hours of the onset of symptoms, can produce detumescence in the outpatient setting, without need for further hospitalization.

<sup>1</sup> Powars, D., et. al., *Hematol/Onc clinics of N. Amer.*: 10(6), 1363-1372, 1996 Dec.

<sup>2</sup> Mantadakis E., et al, *J Pediatr Hematol Oncol.* 1999;21:518-522

<sup>3</sup> Mantadakis E., et. al., *Blood*: 95(1): 78-82, 2000 Jan. 1

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