Case Reports/Case Series

Management of priapism in a child with sickle cell anemia; successful outcome using epidural analgesia

[Prise en charge du priapisme chez un enfant souffrant d’anémie falciforme ; résultat favorable avec l’analgésie péridurale]

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Purpose: To describe the successful management of priapism secondary to sickle cell anemia in a child using neuraxial analgesia provided via an epidural catheter.

Clinical features: A seven-year-old male presented with chest crisis and priapism which, following hemoglobin electrophoresis led to a new diagnosis of sickle cell anemia. Epidural management was attempted as an alternative to surgery after failure of more conventional medical and surgical methods to treat the priapism. The patient’s clinical condition improved with this intervention and together with further conservative therapy resulted in complete resolution of the priapism.

Conclusions: Priapism is a well described complication of sickle cell anemia that is painful and difficult to manage. Surgical intervention is the last therapeutic resort and often results in significant long-term morbidity. This case highlights how select cases of priapism can be successfully managed with epidural neuraxial blockade which not only provides superior analgesia for the often painful conservative treatments, but may also per se impart a direct and salutary therapeutic benefit.

PRIAPISM is a well described complication of sickle cell anemia. We describe the therapeutic management of a seven-year-old male with newly diagnosed sickle cell anemia, confirmed by hemoglobin electrophoresis, whose initial presentation was chest crisis and priapism. After failure of conventional medical and surgical management, neuraxial analgesia was provided via an epidural catheter, as an alternative to surgery. The management of priapism with neuraxial anesthesia is uncommon and there are only two case reports of unintended, suc-
cessful resolution of priapism following epidural placement for either operative management or analgesia.\textsuperscript{1,2} There are also case reports, though likewise rare, of persistent erections during and after neuraxial anesthesia and this complication may have discouraged its use as a therapeutic modality for priapism.\textsuperscript{3,4}

Verbally expressed consent for publication, according to our institutional criteria, was obtained from the mother of the child whose case we report here.

Case report
A seven-year-old, 17-kg male presented to a referring hospital with pleuritic chest pain and dyspnea at rest. A chest radiograph demonstrated an infiltrate on the left lung and was diagnosed as community acquired pneumonia. The patient was discharged home on a course of oral antibiotics but returned the next day with worsening dyspnea, chest pain and a persistent erection. A diagnosis of sickle cell anemia was confirmed by hemoglobin electrophoresis demonstrating a hemoglobin of 68 g·dL\textsuperscript{-1} with a hemoglobin S fraction of 83.2\%. The patient was transferred to the Hospital for Sick Children, Toronto and over the next 24 hr was treated with intravenous hydration, oxygen therapy, intravenous antibiotic therapy and morphine by intravenous infusion. Exchange transfusion was performed and the patient was subsequently referred to the Department of Urology for further management of his priapism. Though his respiratory function improved and his hemoglobin rose to 97 g·dL\textsuperscript{-1} with a hemoglobin S fraction of 16\%, the traditional approaches of cavernosal phlebotomy and local irrigation with phenylephrine failed, despite multiple attempts. Approximately 36 hr after the onset of priapism, the on-call urologist contacted the on-call anesthesiologist to request provision of anesthesia for surgical intervention. A trial of epidural analgesia was suggested and was agreed upon by the urologists. Consultation with the specialist inpatient (acute) pain service was not sought.

On presentation to the operating room the patient’s vital signs were within normal limits, and oxygen saturation of 96\% was observed on supplementary oxygen. Asked retrospectively, on a four-point verbal rating scale (none, mild, moderate, severe), the pain had been either moderate or severe for much of the previous 36 hr. There was no evidence of respiratory distress. Physical examination of the genitalia revealed a tender erection. He was somewhat drowsy on iv morphine infusing at 40 µg·kg\textsuperscript{-1}·hr\textsuperscript{-1}. After parental consent and transfer to the procedure room, the patient was sedated with propofol 40 mg and sedation was maintained with propofol by infusion at a rate of 200 µg·kg\textsuperscript{-1}·min\textsuperscript{-1}. A lumbar epidural was sited using a midline approach and the epidural space identified by loss of resistance to saline at the L3/4 interspace with an 18G Tuohy needle. A test dose of 2 mL of 0.25 % bupivacaine with 1:200,000 mg epinephrine was administered without complication. A further 10 mL of 0.25 % bupivacaine with 1:200,000 mg epinephrine was then given via the epidural catheter and the patient transferred to the postanesthetic care unit.

Fifteen minutes after the procedure, the patient reported no pain, and there was no penile tenderness on examination. The morphine infusion was continued for the chest crisis and an infusion of bupivacaine 0.125 % without opioid was commenced via the epidural catheter at a rate of 4 mL·hr\textsuperscript{-1} (i.e., 0.25 mL·kg\textsuperscript{-1}·hr\textsuperscript{-1}). The patient was transferred to the ward where the Urology services noticed partial detumescence without further intervention and felt that a surgical shunt was no longer warranted. The priapism gradually resolved over the next 16 hr. There was no worsening or recurrence of the priapism. Phlebotomy and intracavernosal phenylephrine injection was performed on one more occasion after placement of the catheter as part of the routine conservative therapy that had been instituted. The patient experienced no pain or discomfort during this procedure. The epidural catheter was removed 48 hr after placement. The patient remained pain free throughout, and was discharged home the following day.

Discussion
Priapism is a persistent erection of the penile corpora cavernosa lasting greater than six hours.\textsuperscript{5} Prolonged episodes of priapism can result in corporal fibrosis and subsequent erectile dysfunction, hence proper diagnosis and treatment is considered a medical emergency. Etiology is commonly idiopathic but precipitating factors include alcohol ingestion, pharmacological ingestion/toxicity, hematological disease and trauma.\textsuperscript{6,7} Sickle cell anemia is an autosomal recessive genetic disease that is caused by the production of defective hemoglobin (hemoglobin S) that “sickles” for example during hypoxemia, hypotension, acidemia, episodes of pain and stress resulting in end-organ ischemic damage. Priapism is a well described complication of sickle cell anemia with an incidence of 30–45\% and bimodal peaks of distribution at 5–13 yr and 21–29 yr of age.\textsuperscript{8,9} Traditional management of sickle cell associated priapism comprises therapies focusing on the causative pathophysiology (i.e., supplemental oxygen therapy, intravenous hydration, analgesia and exchange transfusion). As recommended by the American Urological Association guidelines these interventions should be