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CASE REPORT

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A case of non-ischemic priapism in a healthy 7-year-old boy

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Abstract

Priapism is a urologic emergency more commonly encountered in the adult population. Causes include medication adverse effect, chronic disease, malignancy, dialysis, spinal cord lesions, trauma, infection/inflammation, neuropathy, and idiopathic. Evaluation and treatment focus on identifying and eliminating etiology along with supportive measures. If medical management is not effective, surgery is usually performed to preserve function.

KEYWORDS
constipation, erection, non-ischemic priapism, pediatric priapism, penile pain, urologic emergency

1 | INTRODUCTION

Priapism is characterized as a persistent erection of the penis or clitoris lasting at least 4 hours in duration. The incidence in the United States is 0.73 per 100,000 males, with bimodal peaks occurring at 5 to 10 years of age in children and 20 to 50 years of age in adults.[1,2] The incidence in children is unknown, although it has been well established that sickle cell anemia (SCA) is the most common cause, whereas other etiologies include leukemia, trauma, medications, and ingestions. Much like the approach in adults, identifying ischemic etiologies is essential.[3] Ischemic priapism occurs when relaxation and paralysis of the cavernosal smooth muscle is impaired, resulting in compartment syndrome of the penis. As a result of increasing hypoxia and tissue acidosis,[4] injury begins within 4 hours. Without treatment, significant destruction occurs at 12 hours and irreversible damage after 24 hours, making this a urologic emergency.[5] Initial management may include trials of exercise, urination, cold packs or baths in non–sickle cell patients, and oral fluids.[6] Further treatments include pain control, oxygen, and repeated irrigation and aspiration of phenylephrine into the cavernosum after a penile dorsal nerve block. Ultimately, stent placement may be necessary to bypass the occlusion. Although ischemic priapism requires emergent management, up to 62% of non-ischemic priapism cases resolve spontaneously.[7] These cases are generally caused by traumatic injury, urologic procedures, medications, and obstructive processes. SCA can also cause non-ischemic and stuttering priapism depending on the location of sickling, inflammation, and vasoconstriction. While considered less emergent, stuttering or intermittent priapism can be extremely bothersome, embarassing, and painful for patients of all ages.

2 | CASE REPORT

A 7-year-old boy presented to a community hospital emergency department (ED) complaining of penile pain. His mother reported that the pain started the night before while he was fully erect. She noted no skin changes or rash and normal testicles. When this continued into the next morning, she called the pediatrician, who directed them to the ED. He denied dysuria, hematuria, trauma, abdominal pain, fever, vomiting, diarrhea, and constipation. He had been eating, drinking, and voiding his usual amounts. His 14-point review of systems was negative except for penile pain and sustained erection. The patient's medical history was only significant for attention-deficit hyperactivity disorder without kidney, urologic, neurologic, or hematologic disease. There was

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no significant surgical or family history, and the patient had no known allergies or exposures.

Laboratory tests were performed after oxygen was started via a non-rebreather mask and a 20 mL/kg normal saline fluid bolus was administered. A complete blood count revealed a normal WBC count and differential with no signs of inflammation or malignancy. His hemoglobin, hematocrit, and platelets were within normal limits. The patient provided a urine sample without difficulty. Urinalysis revealed a mildly elevated specific gravity at 1.027, a normal pH of 7.5. It was negative for nitrites, leukocyte esterase, glucose, ketones, and bilirubin. There were 10 mg/dL protein, 1 WBC, and 2 RBC per high-power field. Pediatric urology was consulted by phone, and the patient was transferred to our pediatric ED.

Upon arrival, the patient appeared comfortable without acute distress. He rated his pain a 0 on a scale of 1 to 10. His blood pressure was 104/70 while lying supine. His pulse was 82 beats per minute, respirations were 22 breaths per minute without retractions, and oxygen saturations were 100% on room air. His temperature was 36.8°C taken orally, and his weight was 26.2 kg, which was at the 75th percentile for age. He had a normal head, ears, eyes, nose, throat, skin, cardiopulmonary, musculoskeletal, and neurologic examination. There was no notable lymphadenopathy in the neck, axilla, or groin. His abdomen was soft and non-tender in all quadrants without distension or guarding. There were no masses, organomegaly, or hernias. His scrotum was normal for the stated age. His testes were normal in size and shape, without tenderness. Cremasteric reflex was present bilaterally. His circumcised penis was fully erect without tenderness. There was good capillary refill in the glans and no notable lesions. There was mild erythema indiscriminately on the shaft.

Erect (Figure 1) and supine (Figure 2) radiographs of the abdomen revealed a non-obstructive bowel gas pattern with diffuse dense stool throughout the colon and rectum. Urology recommended aggressive treatment, and a sodium phosphate, mineral oil, and glycerin enema was administered. The patient had 3 large bowel movements, and his priapism completely resolved. When specifically questioned, he could not recall his last bowel movement but denied hard stools. His mother denied any history of constipation. The urologist arrived soon after resolution and reassured the family. The patient was discharged home with instructions for an aggressive bowel cleanout using polyethylene glycol 3350 followed by a daily regimen for 1 week until follow-up with the pediatrician. The urologist contact information was also provided.

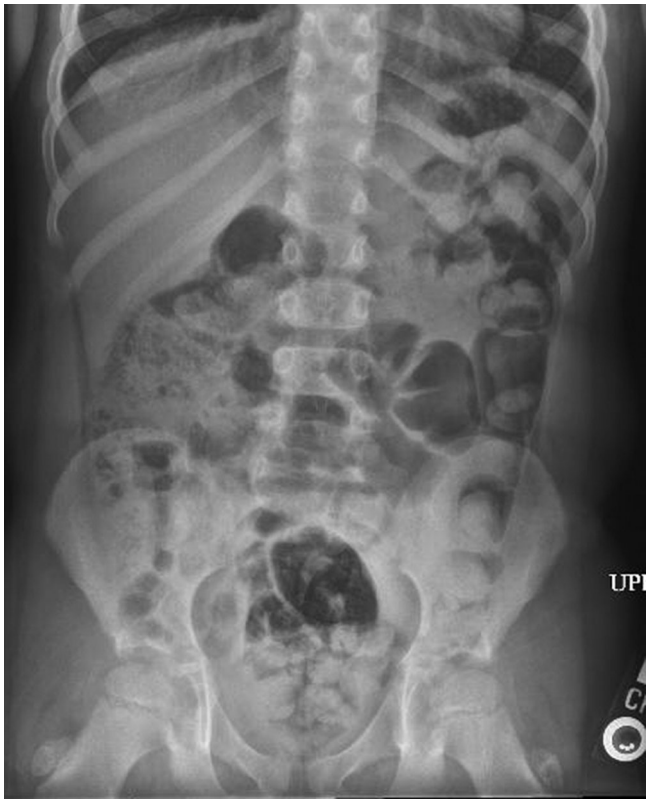

FIGURE 1 Upright abdominal radiograph demonstrating formed stool throughout the colon and rectum

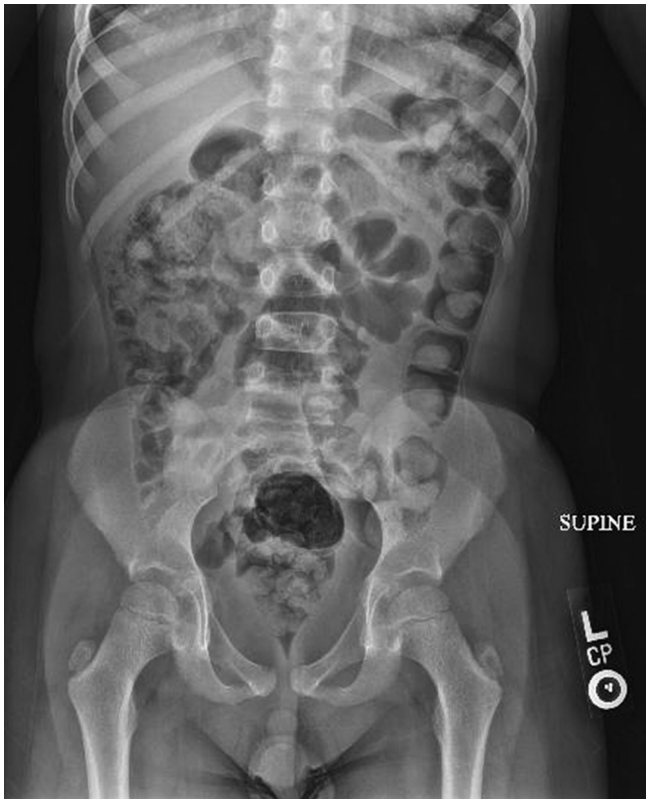

FIGURE 2 Supine abdominal radiograph demonstrating formed stool throughout the colon and rectum

3 | DISCUSSION

An excessive stool burden causes priapism by physically obstructing the pelvic blood vessels. The penis consists of 3 cylindrical compartments of spongy soft tissue called the corpus cavernosum and corpus spongiosum, supplied by a trabeculated system of arteries and veins. The network allows blood to flow in and out of the compartments to promptly create and resolve an erection. This is mediated by autonomic and motor neurons via the cavernous and pudendal nerves,

respectively. To create an erection, the internal pudendal artery expands to increase blood flow into the corpus cavernosum and corpus spongiosum. The complex venous system narrows when the arteries relax to maintain the erection until the task is completed.

Drainage of blood from the penis is intricate. When the cavernous and/or pudendal nerves signal drainage, the venous system expands from the previously narrowed state. There are 3 venous drainage systems from the penis, all of which drain into various venous plexuses or the internal pudendal veins. In young boys, the proximity of this complex drainage system to the sigmoid colon and rectum is only a few millimeters. A large stool burden can physically obstruct any part of the system, creating low-flow, non-ischemic priapism.

As we know, most boys with a large stool burden do not experience sustained erections. However, constipation and elimination dysfunction are exceedingly common in the United States. Although constipation refers to infrequent, hard, and sometimes painful stools, elimination dysfunction can include difficulty releasing both stool and urine and often occurs after a stressful potty-training event or painful diaper rash. In such cases, constipation may occur as a result of withholding even though the stool itself is not dry and hard. A thorough history can often help make the diagnosis. This includes a complete dietary history and voiding regimen as well as eliciting characteristics from patients about their stool, such as size, quantity, and consistency. If history is not helpful or difficult to collect, consider a radiograph of the abdomen to assess stool burden or administration of an enema or oral magnesium citrate in the ED and monitor for response. Most patients will have at least a moderate reduction in symptoms after even partial evacuation of their bowels. They may experience stuttering priapism, or intermittent reoccurrence shortly after, as more stool from above fills the lower colon and rectum. Therefore, we suggest an aggressive approach with bowel cleanout either inpatient or outpatient, depending on the patient and family capabilities.

Finally, if constipation is unlikely to be the cause, other etiologies include undiagnosed sickle cell disease, envenomation (snakes, scorpions, or spiders), poisoning (psychoactive medications, sildenafil, or cocaine), cavernositis, recent epidural anesthesia, leukemia, hypercoagulable state with thrombus, or medullary tumors.[8] Laboratory assessment should be considered to evaluate for these conditions. Also, consider ultrasound to assess for a pelvic mass. In fact, ultrasonography with Doppler can evaluate blood flow in the pelvic vessels and has been shown to be 100% sensitive and specific with a diagnosis of priapism as well as helping to detect fistulae formation and monitoring response to therapeutic interventions.[3] Because trauma is also on the differential, consider sexual abuse, foreign body, or exploratory behaviors when indicated. Any child, boy or girl, with bowel or urinary obstructive symptoms should also warrant consideration of concomitant obstructive uropathy, bowel injury with ischemia, or pyelonephritis. Severe constipation with obstructive uropathy has been reported with acute renal failure and even severe resultant bowel dysfunction. This is referred to as Hinman syndrome.[9–11] Prompt urology consultation to help guide evaluation is almost always recommended given the rarity of priapism in children and the potential need for urgent interventions.

Constipation and elimination dysfunction in the United States are common problems. Pediatric patients brought to the ED for abdominal pain are often accompanied by parents worried for something far more intimidating, such as appendicitis. MacGeorge et al, at the Medical University of South Carolina, demonstrated that from 2012 to 2013, in their population of 17 million commercially insured children aged 0 to 17 years, 2.6% were diagnosed with constipation, and 14.5% of them were treated in an ED. Of those 65,163 patients, 45% had no visits to their primary care doctors 30 days before or after the visit.[12] Unfortunately, this tells us that many children with constipation will see only an emergency provider for their burdensome, painful, and sometimes chronic symptoms. In addition, Mutyala et al estimated that 40% to 50% of patients experience at least 1 relapse of constipation within 5 years, suggesting that many will return for similar symptoms.[13]

There are numerous methods to treat constipation acutely and chronically. In general, the North American Society for Pediatric Gastroenterology, Hepatology & Nutrition recommends lifestyle and dietary modifications.[14] Some children report that they are frankly too busy to poop, whereas others are embarrassed to go in school or scared of experiencing pain. Another common cause is chronic dehydration leading to dry, inspissated stools. Asking simple guided questions to understand the reasons for infrequent or withholding stools can guide interventions effectively. Recommend a healthy diet with adequate fiber and fluids as well as exercise to stimulate movement in a sluggish gut. Timed toileting can help younger children. This means the child sits on the toilet, with a book or tablet, 3 times daily for at least 10 minutes. This will help train the body to evacuate often. Finally, medical interventions usually include osmotic laxatives only. If dietary changes, behavior modifications, and osmotic agents are not enough, some providers will try a stimulant agent. When interventions include more than daily stimulants such as polyethylene glycol, referral to a pediatric gastroenterologist should be considered.

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CONFLICT OF INTEREST

The authors declare no conflict of interest.

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