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

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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. 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. 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, injury begins within 4 hours. Without treatment, significant destruction occurs at 12 hours and irreversible damage after 24 hours, making this a urologic emergency. Initial management may include trials of exercise, urination, cold packs or baths in non–sickle cell patients, and oral fluids. Further treatments include pain control, oxygen, and repeated irrigation and aspiration of phenylephrine into the cavernous 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. 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, embarrassing, 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. 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. The patient’s medical history was only significant for attention-deficit hyperactivity disorder without kidney, urologic, neurologic, or hematologic disease. There was 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 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 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 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 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.

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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.

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.
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.

ACKNOWLEDGMENTS
We thank Kaitlin Severini for editing.

CONFLICT OF INTEREST
The authors declare no conflict of interest.

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REFERENCES
1. Roghmann F, Becker A, Sammon JD, et al. Incidence of priapism in emergency departments in the United States. J Urol 2013;190:1275.
2. Cherian J, Rao AR, Thwaini A, et al. Medical and surgical management of priapism. Postgrad Med J 2006;82:89.
3. Donaldson JF, Rees RW, Steinbrecher HA. Priapism in children: a comprehensive review and clinical guideline. J Ped Urol. 2014;2014:11–24.
4. Pryor J, Akkus E, Alter G, et al. Priapism. J Sex Med. 2004;1:116.
5. Spycher MA, Hauri D. The ultrastructure of the erectile tissue in priapism. J Urol 1986;135:142.
6. Maples BL, Hagemann TM. Treatment of priapism in pediatric patients with sickle cell disease. Am J Health Syst Pharm. 2004;61(4):355–363.
7. Montague DK, Jarow J, Broderick GA, et al. American Urological Association guideline on the management of priapism. *J Urol* 2003;170:1318.

8. Clement P, Giuliano F. Neurology of Sexual and Bladder Disorders. Handbook of Clinical Neurology; 2015:19–37.

9. Hinman F, Baumann FW. Vesical and ureteral damage from voiding dysfunction in boys without neurologic or obstructive disease. *J Urol*. 2017;197(2S):S127–S131. https://doi.org/10.1016/j.juro.2016.10.098. Epub 2016 Dec 21.

10. Bauer SB. The Hinman syndrome. *J Urol*. 2017;197(2S):S132–S133. https://doi.org/10.1016/j.juro.2016.11.026. Epub 2016 Dec 20.

11. Mannemuddhu SS, Bayne CE, Tufan Pekkucuksen N, Shoemaker LR. Attention to detail: not simple bed wetting-Hinman-Allen syndrome. *J Nephrol*. 2021;34(1):247–250. https://doi.org/10.1007/s40620-020-00810-4. Epub 2020 Jul 27.

12. MacGeorge CA, Simpson KN, Basco WT, Jr., Bundy DG. Constipation-related emergency department use, and associated office visits and payments among commercially insured children. *Acad Pediatr*. 2018;18(8):952–956. https://doi.org/10.1016/j.acap.2018.04.004. Epub 2018 Apr 16.

13. Mutyala R, Sanders K, Bates MD. Assessment and management of pediatric constipation for the primary care clinician. *Curr Probl Pediatr Adolesc Health Care*. 2020;50(5):100802. https://doi.org/10.1016/j.cpped.2020.100802. Epub 2020 Jun 10.

14. Tabbers MM, DiLorenzo C, Berger MY, et al; European Society for Pediatric Gastroenterology, Hepatology, and Nutrition; North American Society for Pediatric Gastroenterology, Evaluation and treatment of functional constipation in infants and children: evidence-based recommendations from ESPGHAN and NASPGHAN. *J Pediatr Gastroenterol Nutr*. 2014;58(2):258–274. https://doi.org/10.1097/MPG.0000000000000266.

**How to cite this article:** Mercurio D, O’Donnell J. A case of non-ischemic priapism in a healthy 7-year-old boy. JACEP Open. 2022;3:e12785. https://doi.org/10.1002/emp2.12785