Case Report

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Management of ischemic priapism in a 14 year old patient

14 yaşındaki hastada iskemik priapizmin yönetimi

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Abstract

Priapism can occur in all age groups, including neonatales, children and adolescents. Pediatric priapism treatment is similar to adult priapism treatment, but there are no guidelines for the management of priapism, which is not common in children. We present a pediatric low-flow priapism case who was unresponsive to conservative methods and underwent T-shunt operation. In the present case, we wanted to emphasize when distal shunt surgery, which is rarely performed in pediatric patients, is necessary and how it is managed.

Keywords: Pediatric ischemic priapism, T-shunt surgery, pediatric priapism case report.

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Öz

Priapizm yenidoğan, çocuk ve ergenler de dahil olmak üzere tüm yaş gruplarında ortaya çıkabilir. Pediatrik priapizm tedavisi erişkin priapizm tedavisine benzer ancak çocuklarda yaygın olmayan priapizmin tedavisine yönelik bir kılavuz bulunmamaktadır. Konservatif yöntemlere yanıt vermeyen ve T-şant ameliyatı geçiren pediatrik düşük akımlı priapizm olgusunu sunmayı amaçladık. Bu olguda çocuk hastalarda nadiren uygulanan distal şant ameliyatının ne zaman gerekli olduğunu ve nasıl yönetildiğini vurgulamak istedik.

Anahtar kelimeler: Pediatrik iskemik priapizm, T-şant ameliyatı, pediatric priapizm olgu sunumu.

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Introduction

Priapism is defined as having a full or partial penile erection for more than 4 hours without sexual stimulation [1]. Priapism can occur in all age groups, including neonatales, but it is rarely observed in children and adolescents. Its incidence in all ages is estimated to be 0.3–5.3/100,000 per year, with the 5th decade being the most common [2-4]. Data on the prevalence of the disease in children are insufficient [5].

Hematological diseases, infections, drugs, trauma, and iatrogenic causes are the most common causes of priapism in children [6]. Most cases of low-flow pediatric priapism occur in boys with sickle cell anemia, leukemia, and other hematological disorders [7]. Low-flow priapism, which can cause temporary or irreversible erectile dysfunction, penile deformity, and psychological sequelae, is a urological emergency [8]. High-flow non-ischemic priapism is most commonly observed as a complication of penile trauma, with up to 62% of these cases resolving spontaneously [9].

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Pediatric priapism treatment is similar to adult priapism treatment. First, conservative methods such as exercise, cold application, urination, mechanical compression, and masturbation are applied. When those treatment fails, corporal aspiration, blood transfusion, and shunt surgeries are applied [8]. There are no guidelines for the management of priapism, which is not common in children. Therefore, the treatments applied can help to guide disease management.

Case presentation

A 14-year-old patient presented to the emergency department with a painful erection that had been going on for a week. It was discovered that the erection developed spontaneously, and the patient had never experienced such a situation before. The patient did not have dysuria, hematuria, abdominal pain, fever, vomiting, diarrhea, or constipation. Moreover, there was no history of trauma. The patient was on a usual, normal diet, and his urine output was normal. There was no hematological disease, malignancy, or drug history. The body mass index was 38.06. Genital examination revealed that the penis was painful and rigid, the corpus cavernosum was hard, and the glans penis and corpus spongiosum were soft. No signs of trauma or ecchymosis were observed. Although the patient had a long-term erection, no signs of penile necrosis were observed. The patient's complete blood count and bleeding parameters were normal. Penile Doppler ultrasonography revealed that no significant arterial flow was observed in both cavernous bodies, indicating ischemic priapism.

As the first-line therapy, corporal aspiration accompanied with analgesia was applied. Corporal aspiration was performed with a 20-gauge butterfly needle from the corpus cavernosum at the 3–9 o'clock position, close to the root of the penis. Aspiration was not successful, and appropriate blood gas could not be obtained. A cavernosal saline infusion and a 1/1,000,000-mg epinephrine infusion were administered four times at 10-minute intervals. Penile rigidity partially decreased after the infusion. A cold application was applied after compression dressing with Coban. The penis became rigid again in the follow-up the next day (Figure 1), and the patient was scheduled for distal shunt surgery.

During the operation, a No.11 scalpel was used to perform a T-shunt technique from the glans on the right lateral side of the urethral meatus. After the incision, milking was performed. Because sufficient detumescence was provided in the penis, no intervention in the left cavernosal body was needed (Figure 2).

Blood gas was taken perioperatively after milking, and partial oxygen pressure was 44 mmHg, partial carbon dioxide pressure was 42 mmHg, and pH was 7.29. Then, a 6-fr feeding tube was placed and secured in the right corpus cavernosum (Figure 3).

As the penile rigidity did not recur, the feeding tube was removed on the 2nd postoperative day and the patient was discharged. The patient was consulted to pediatric hematology, and the hemoglobin electrophoresis result was normal. No hematological diseases, such as sickle cell anemia or hemolytic anemia, were detected. Three months after the procedure, penile erections were normal and there was no recurrence of priapism.

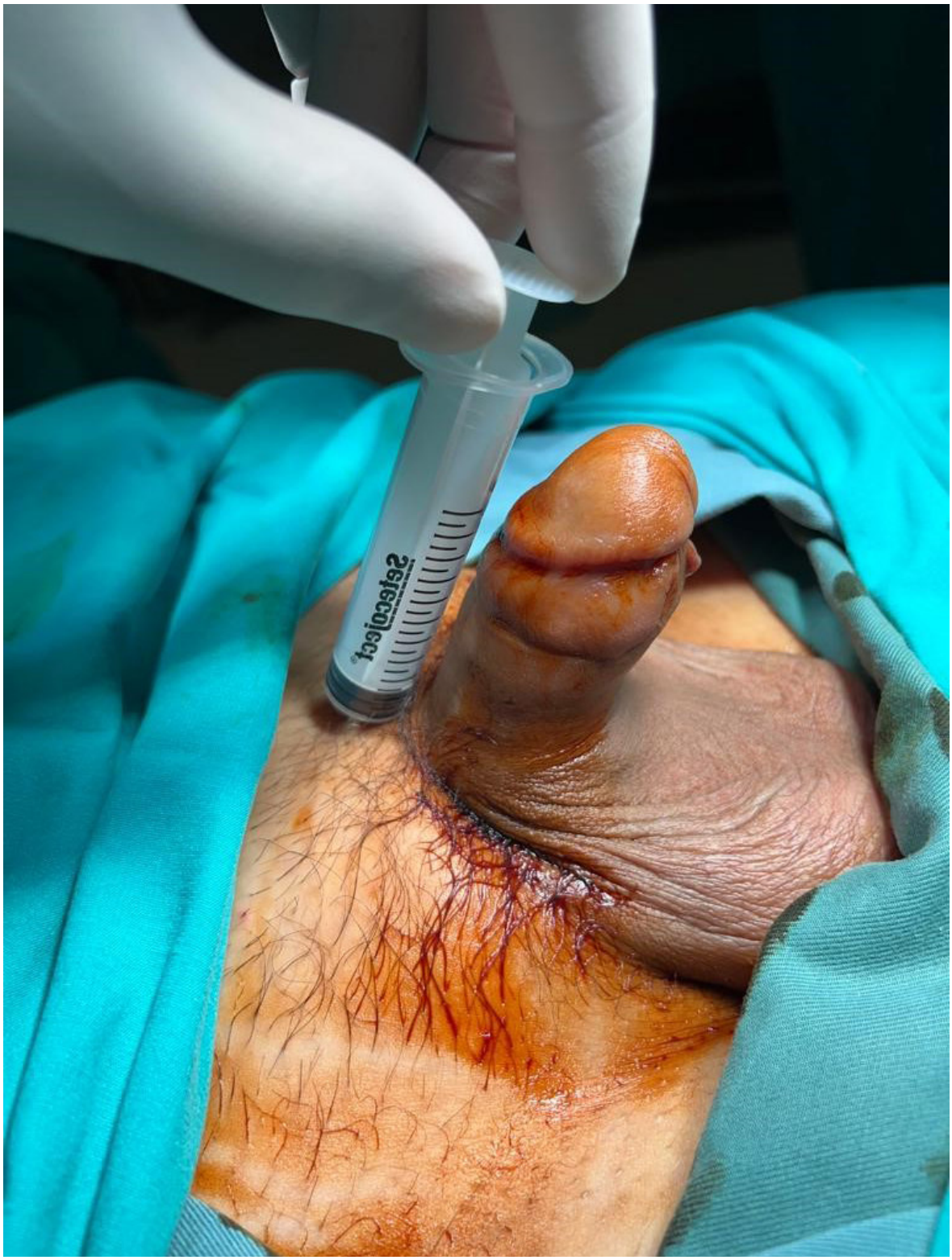

Figure 1. Rigid penis prior to distal shunt surgery

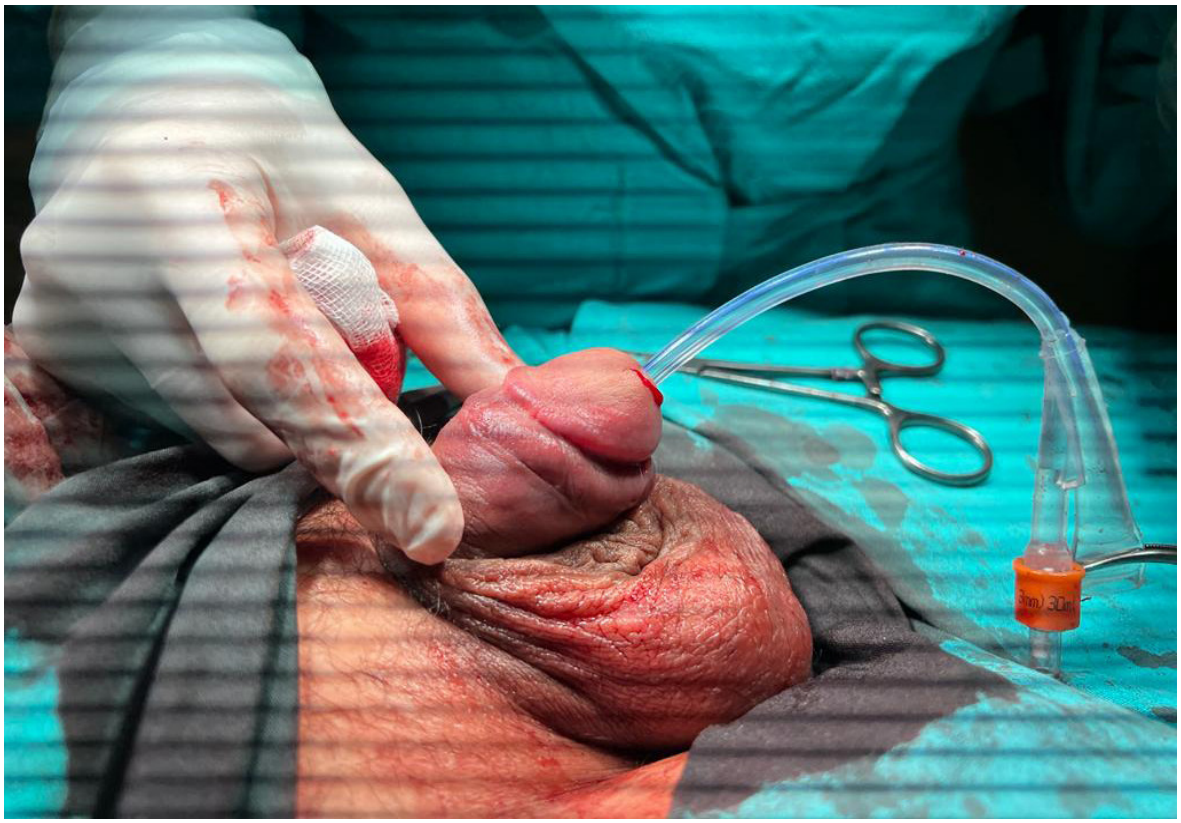

Figure 2. Detumescent penis after distal shunt surgery and milking

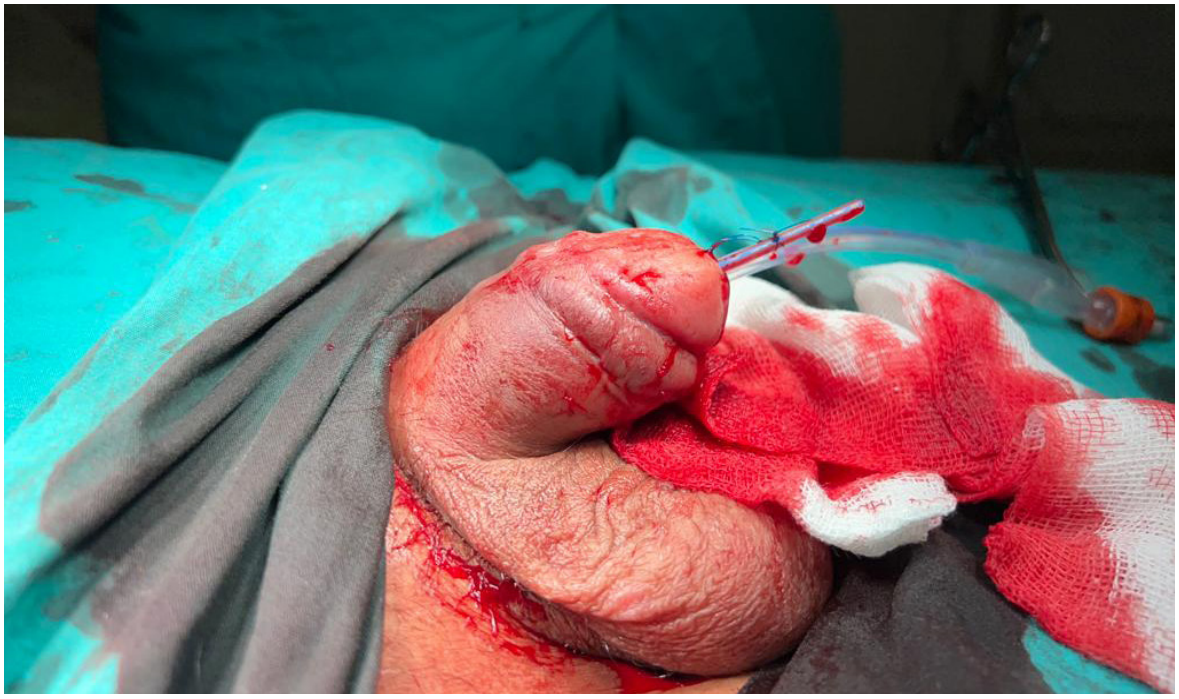

Figure 3. A feeding tube was placed in the penis after detumescence

Discussion

Priapism should be treated promptly because it can cause serious, irreversible complications after a prolonged erection. However, in different ethnic, religious, and social environments, the disease may be diagnosed late owing to family members' feelings of shame and stigma.

Various treatment modalities, such as mechanical (continuous perineal compression and cold application), pharmacological (intracavernous, venous, or oral drug therapy), radiological (selective transcatheter embolization therapy), and surgical (arterial ligation or arteriovenous shunts), are used in the treatment of priapism. The use of noninvasive, conservative methods with high success rates is reduces the need for surgical intervention [10].

Especially before invasive methods are applied, it should be determined whether the priapism is low or high flow. Intracavernosal blood gas analysis and penile Doppler ultrasonography are widely used to distinguish between low- and high-flow priapism. High-flow priapism, which rarely causes pain, is usually caused by excessive arterial flow due to perineal or penile trauma, which creates a fistula between the cavernosal artery and corpus cavernosum. If conventional treatments fail, arterial embolization and ligation treatments are used [11].

Low-flow priapism, which is more common in pediatric patients, is caused by stasis in the cavernosum due to hematological, vascular, or neurological diseases or infectious, drug-related causes. Tissue ischemia caused by stasis causes cavernosal smooth muscle ischemia, pain, and corporal smooth muscle necrosis over time. Tissue fibrosis and permanent impotence may develop over time. Therefore, it is a urological emergency that requires rapid diagnosis and treatment [12]. Erectile dysfunction was observed in 14% of pediatric patients in whom detumescence could not be achieved [13]. In the present case, there was no hematological disease, malignancy, trauma, or drug use history that could cause priapism, and no complications were encountered during the 3rd month of follow-up.

In low-flow priapism, the aim is to provide venous outflow for the arterial blood supply of the corpus cavernosa with surgical treatment. Among the initial treatments, blood aspiration from the corpus cavernosum, saline irrigation applied when necessary, and intracavernosal phenylephrine/epinephrine injection were successful in 77% of the cases [9]. If those treatments fail, shunt surgery should be considered. In a previous case report, a 14-year-old male patient was successfully treated with intracavernosal tissue plasminogen activator after shunt surgery was unsuccessful [14].

Four shunt procedures have been described as a percutaneous distal (corporaglanular) shunts (Winter, Ebbehoj, Lue), open distal (corporoglanular) shunts (Al-Ghorab, Burnett), open proximal (corporospongiosal) shunts (Quackles, Sacher), and vein anastomoses/ shunts (Grayhack, Barry) [15]. In cases of priapism lasting more than 36 hours, the erectile tissue is generally impaired both structurally and functionally. These four shunt methods used to prevent structural deterioration have not been shown to be superior in terms of erectile function to each other [16]. The surgeon's experience and familiarity with the technique are important factors in deciding which shunt procedure to use. However, distal shunts are recommended because they are easier to apply and have fewer complications [9].

In a study of 13 adult male patients who underwent T-shunt surgery for priapism, 6 of them had a history of unsuccessful distal or proximal shunt surgery. After the operation, pain was reduced and penile detumescence was achieved in all patients, but only two patients' erectile function could not be preserved during follow-up. In this study, T-shunt was defined as an easy-to-use, reliable technique that provides rapid resolution of penile pain and rigidity [17].

In a 7-year-old patient with cerebral palsy who underwent surgery for an extra finger incision and developed propofol-induced priapism, corporal aspiration and intracavernosal epinephrine were used, and then distal shunt surgery was performed on the next day because of a persistent painful erection. The patient was followed up for 1 year after surgery, and penile erection was normal [12]. In another case report, a 7-year-old male patient with no comorbidity, drug history, or trauma history presented to the emergency department with a painful erection that began during the night, and abdominal direct X-ray revealed non-obstructive

gas and a dense stool pattern along the colon and rectum. After constipation treatment, the patient began to have bowel movements and after defecation, the erection spontaneously regressed due the resolution of the obstruction in the pelvic blood vessels, and no additional treatment was needed [18]. In the case report of Brönimann et al. [19], the complete blood count and bleeding parameters of a 12-year-old boy who had a painful erection for 24 hours were found to be normal. The patient was previously diagnosed with COVID-19 7 weeks ago and was in the subacute period. The COVID-19 PCR test result performed at admission was also positive; corporal aspiration was performed twice because of the recurrence of erection after 3 days, and detumescence was achieved. The patient was followed up after 8 weeks, and penile erection was found to be normal [19]. In the present case, the patient had a painful erection for approximately 1 week. Blood gas analysis could not be performed on cavernosal blood that had thickened due to long-term priapism. Doppler ultrasonography revealed low-flow priapism, and corporal aspiration was performed. However, distal shunt surgery, which is rarely performed in children, was performed owing to the development of penile rigidity the next day, and detumescence in the penis was achieved. The patient was followed up 3 months after surgery, and no loss of erection or recurrence of priapism was observed. The patient expressed that he was satisfied with his treatment because he did not have priapism and erection problems again.

Primary treatments are usually sufficient and successful in pediatric patients with priapism, but distal shunt surgery is rarely used in patients with prolonged and resistant priapism. In the present case, we wanted to emphasize when distal shunt surgery, which is rarely performed in pediatric patients, is necessary and how it is managed. Considering its serious potential complications, rapid differential diagnosis of high- vs. low-flow priapism and early intervention for low-flow priapism are necessary to reduce the rate of permanent sequelae. Informing physicians and families, especially in cases of recurrent priapism, is important to prevent delayed diagnosis and treatment.

Conflict of interest: No conflict of interest was declared by the authors.

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Informed consent: The patient and his parents gave informed consent for the publication.

Authors' contributions to the article

A.S. constructed the main idea and hypothesis of the study. S.B developed the theory and arranged/edited the material and method section. Discussion section of the article was written by M.B.D. and K.K.

Y.O. and S.C. reviewed, corrected and approved. In addition, all authors discussed the entire study and approved the final version.