ORIGINAL ARTICLE

Rare emergency in children: Priapism and stepwise treatment approach

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ABSTRACT

BACKGROUND: Priapism is a rare condition in children and the treatment algorithm is controversial in this age group. Herein, we report eight cases with low-flow priapism and our stepwise treatment approach in light of literature.

METHODS: We present a simple stepwise treatment for low-flow priapism including five steps. Step 1: Cold compress and analgesia while evaluation the priapism and its etiology. Step 2: Corporal aspiration and adrenaline infusion in the ward. Step 3: Modified Winter shunt in the same place. Step 4: Ketamine application and caudal block in the operating room. Step 5: Sapheno-cavernous (Grayhack) shunt. Eight cases with low-flow priapism were reviewed retrospectively. Symptoms, duration of tumescence, the interventions, and step that provide detumescence were recorded.

RESULTS: The mean age of patients was 8.5 years (1–17 y). The median time of the priapism before admission was 15 h (4–165 h). The etiological factors were sickle cell disease, hemodialysis due to chronic renal failure, and factor V Leiden mutation in three patients. Detumescence was achieved in one patient at Step 2, in two patients at Steps 3, 4, and 5, respectively. Rigidity of cavernous body was observed in one patient in long-term follow-up.

CONCLUSION: Low-flow priapism is a urological emergency that may cause erectile dysfunction. Treatment options should be selected according to a protocol that prevents time loss and avoids more invasive treatment in unnecessary situations. Our algorithm with simple nature and its steps from less invasive to more invasive procedures may be an alternative for the treatment of low-flow priapism.

Keywords: Child; hemodialysis; priapism; sapheno-cavernous shunt; stepwise treatment.

INTRODUCTION

Priapism is a penile erection without sexual stimulus, lasting more than 4 h.[1] It is a very rare condition in children and the most frequent etiologic factor is sickle cell disease (SCD).[1,2]

It is a urological emergency and the treatment should be arranged urgently to avoid its sequelae such as erectile dysfunction (ED), shortening in penile length and deformity in penile shape.[3]

There are so many studies about adult population in this field,[4–7] but about pediatric priapism, due to its rarity, most of the studies in the literature are case studies.[8–13] The treatment algorithms of pediatric priapism that mentioned in the literature are mostly depend on adult outcomes.[3] There are no randomized controlled studies in pediatric population. Therefore, there is no consensus on the management of priapism in pediatrics. Herein, we report the management of eight cases with priapism and discuss our stepwise treatment approach in the light of the previous literature.

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MATERIALS AND METHODS

Institutional Ethical Board approval was obtained. Priapism was diagnosed with a history of more than 4 h erection without any stimulus. Until proven otherwise, all diagnoses of patients were considered low-flow priapism.

Steps of the Treatment Approach

We used a simple and straightforward management approach for priapism (Fig. 1). All steps of the approach were applied to all patients except Step 1 in the patients with SCD. The urgent evaluation of underlying pathology includes history, physical examination, urine analysis and culture, urine and blood tests, and penile Doppler ultrasound (USG). Blood tests include complete blood count, coagulation profile, hemoglobin electrophoresis, and toxicology tests. Hematological evaluation was performed by pediatric hematologist. First evaluation to differentiate ischemic priapism from non-ischemic priapism was performed by penile Doppler USG. If the ischemic (low-flow priapism) is detected, the steps were continue. While evaluating for an underlying pathology, the treatment was started with cold compress for ½ h without time loss (the first step). Cold compress was not applied in patients with SCD, because cold is contraindicated in patients with SCD. Analgesia was also applied simultaneously in the first step with paracetamol in dose of 10 mg/kg. The second step was corporal aspiration and adrenaline injection (CA-AI). An intravenous catheter or a needle was inserted through glans to the cavernous body and the blood was aspirated until the end of the dark blood coming out of the cavernous body. Blood gas analysis was performed to confirm the type of the priapism after corporal aspiration. This provides to remove the acidotic, hypercarbic, and anoxic blood from the cavernous tissue and reduce the pressure. Adrenaline solution (diluted adrenaline – 1 µg/ml) should be prepared before aspiration to avoid repeated needle insertion. 10–15 ml of adrenaline solution was used for each patient. For this step, the patient was taken to the ward and blood pressure and cardiac pulse should be monitored. Corporal aspiration and irrigation can be repeat 2 or 3 times and permanent detumescence should be achieved after the second step immediately. If the detumescence could not provide or tumescence is observed after transient detumescence, we passed the third step. The third step was modified Winter shunt, performed in the ward. Extra holes were created on a 16 gauge angiocatheter and the catheter was inserted through glans into the corpora with some holes aligned with the glans and some of them aligned with the corpora to achieve a distal spongio-cavernous shunt. The fourth step was ketamine anesthesia and placing caudal epidural catheter performed by anesthesiologist in the operating room. About 0.125% bupivacaine was applied for caudal block in 0.75 ml/kg dose. Fentanyl (200 µ/ml) and bupivacaine (5 mg/ml) solution was applied as an infusion through the caudal catheter in dose of 0.2 ml/kg/hr. Ketamine was applied in dose of 0.5 mg/kg intravenously. In case of persistent tumescence after these steps, sapheno-cavernous shunt (Grayhack shunt) was performed, as the fifth step. In this technique; the first incision is performed below the ligamentum inguinale, on the level of sapheno-femoral junction. Saphenous vein is ligated at the tenth centimeter distally (Fig. 2). A tunnel is created between penile shaft and inguinal region for the mobilization of the saphenous vein. The second incision is performed laterally on the penile body, and corpus cavernous

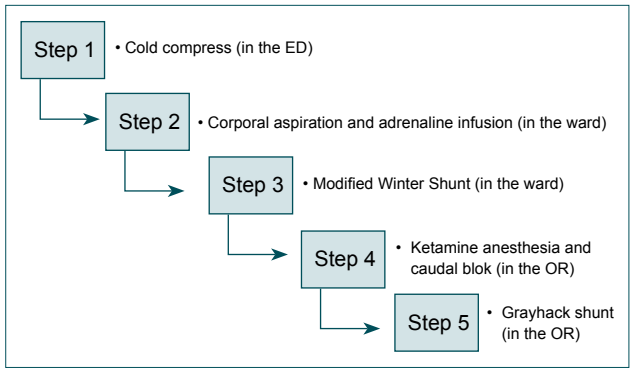

Figure 1. Treatment approach for priapism. ED: Emergency department; OR: Operation room.

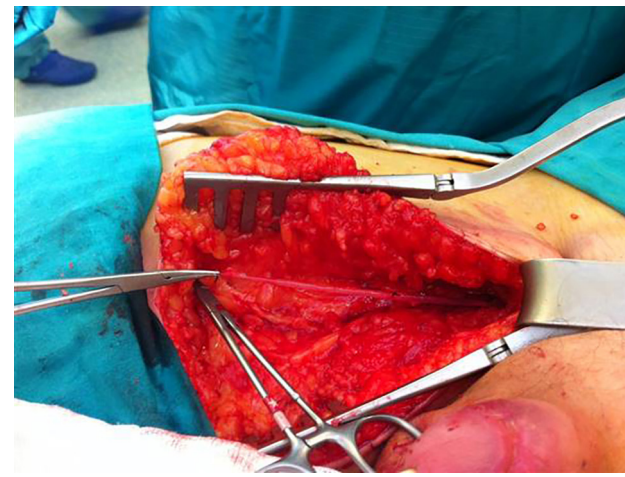

Figure 2. Dissection of saphenous vein.

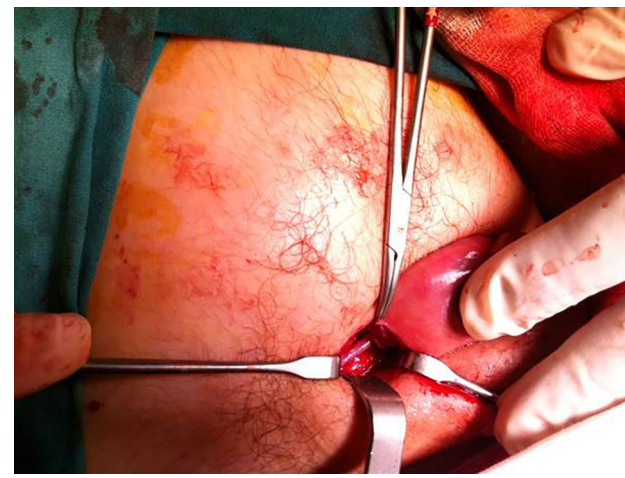

Figure 3. Anastomosis between saphenous vein and corpus cavernosum.

is dissected. The distal end of the saphenous vein is pulled through the tunnel and anastomosed to the cavernous body with 6/0 or 7/0 non-absorbable sutures (Fig. 3).

The follow-up included the visits at the 1st week, 1st month, and every 6 months postoperatively in long term till 18 years of age. The physical examination was performed to determine the functional and cosmetic sequelae of the penis such as deformity, asymmetry of the penile shape, and rigidity of corpora.

Eight cases with low-flow priapism were reviewed retrospectively. There was no patient with high-flow priapism in our study period. Symptoms, duration of tumescence, the interventions, and step that provide detumescence were recorded. The mean, median, minimum, and maximum values were given for the data that show normal and abnormal distribution of numeric values.

RESULTS

The mean age of patients was 8.5 years (1–17 y). The median time of the priapism before admission was 15 h (4–165 h). The median time for resolution of tumescence was 8.5 h (1–48 h). The mean follow-up time was 30.63 months (12–52 m). The etiological factors were SCD, hemodialysis due to chronic renal failure, and factor V Leiden mutation in three patients and priapism was idiopathic in five patients. The priapism was observed during hemodialysis in Case 2. The clinical and etiological features of the cases are summarized in Table 1.

Spontaneous resolution was observed in one patient. In one patient that admitted after the 6th h of priapism, tumescence was resolved in the second step. We performed CA-AI in seven patients and blue dot sign was observed on the glans as a minor complication in two of them. No patient had penile hematoma due to corporal aspiration and irrigation. In two patients, the tumescence was resolved by modified Winter shunt (Step 3). In Case 3 who admitted with priapism lasting 165 h, detumescence was achieved in the third step but recurrence occurred after a few hours and ketamine anesthesia and caudal block was performed. No complication was observed associated with the modified Winter shunt or ketamine anesthesia and caudal block. Four patients underwent ketamine anesthesia and caudal block, in two of them resolution were achieved. In the remaining two patients, Grayhack shunt was performed. After this procedure, detumescence was provided in both patients. These patients were discharged with anticoagulant treatment. One of these two (Case 6) is a candidate for ED in the future due to his cavernous body rigidity on long-term follow-up. The other patient who underwent Grayhack shunt had uneventful follow-up.

The first three steps did not provide sufficient detumescence in Case 6. At that point, parents did not give consent for further treatment and hospitalization. One day later, the patient was readmitted. Sapheno-cavernous shunting was performed at his 59th h of tumescence. After successful shunting, detumescence was achieved immediately.

DISCUSSION

Priapism is a rare condition in childhood compared to adulthood. While the incidence of priapism in adult males is 2.9/100.000/year,[2] the incidence of this disease in children is uncertain.[3] There are two randomized controlled studies among adolescents and adults with recurrence priapism but no randomized control study about low-flow priapism.[4,5] Pediatric priapism is a challenging emergency with its unclear mechanism due to the absence of evidence-based data in pediatric literature.

Table 1. The clinical and etiological features of the cases

	Age (years)	Etiology	Time of tumescence (hour)	Treatment	Follow up (month)
Case 1	10	Idiopathic	24	Step 1–3	48
Case 2	8	CRF + hypertension Hemodialysis*	6	Step 1–4	18
Case 3	7	Idiopathic	165	Step 1–4	28
Case 4	17	Sickle cell disease	18	Step 2–5	48
Case 5	1	Idiopathic	4	Spontaneous resolution	12
Case6	10	Mutation of factor V, MTHFR (A1298C), PAT-1 4G/5G	32	Step 1–5	52
Case 7	6	Idiopathic	6	Step 1–2	22
Case 8	9	Idiopathic	12	Step 1–3	17

*Priapism was observed during hemodialysis. CRF: Chronic renal failure; CA-AI: Corporal aspiration-adrenalin injection. Step 1: Cold compress; Step 2: Corporal aspiration and adrenaline infusion; Step 3: Modified Winter shunt; Step 4: Ketamine anesthesia with caudal block; Step 5: Grayhack shunt.

There are three types of priapism: Low flow (ischemic and veno-occlusive), intermittent (stuttering and recurrent ischemic), and high flow (non-ischemic and arterial). The main goal of the evaluation of the patient with priapism is to distinguish the ischemic (low flow) priapism from non-ischemic (high flow) priapism. History of the patient is important. Ischemic priapism is usually painful while non-ischemic priapism usually is not. History of using vasoactive agents for intracavernous injection therapy and history of pelvic, genital or perineal trauma, especially a perineal straddle injury, is usually seen in high-flow priapism. In the physical examination of patients with low-flow (ischemic) priapism, the corpora cavernosa are often completely rigid, while in patients with high-flow priapism, the corpora may not be completely rigid. Physical examination of pelvis and perineum is also important to determine the evidence of trauma. Cavernosal blood gas analysis and color Doppler USG of penis are the most reliable diagnostic methods to differentiate ischemic from non-ischemic priapism. Cavernosal blood gas analyses in patients with ischemic priapism reveal a PO_2 <30 mmHg, PCO_2 greater than 60 mmHg, and pH <7.25. Cavernosal blood gas analyses in patients with non-ischemic priapism are similar to arterial blood gas levels. Color Doppler USG is performed in the lithotomy or frog-leg position. Patients with ischemic priapism have little or absent of blood flow in the cavernosal arteries, while patients with non-ischemic priapism have normal or increased blood flow in the cavernosal arteries on Doppler USG.[14]

Low-flow priapism is the most common type seen in children.[3] Low-flow priapism is characterized by venous occlusion and vascular stasis and mainly caused by hematologic disorders such as SCD, leukemia, and hypercoagulable states or adverse drug reactions, while high-flow priapism is mainly caused by penile or pelvic trauma, cavernous vasoactive drug injections, bites of scorpion, spider or snake, some medications such as psychoactive medications, anticoagulants, and propofol, and local infections (cavernositis) in children.[8,9] Stuttering priapism is a distinct pathology that shows short and self-limiting tumescence episodes. SCD is usually the etiologic factor[15] and sometimes require acute medical intervention in adult population in this type of the priapism but in childhood, it is less understood.[16,17] Although corporal aspiration and adrenaline infusion (CA-AI) were reported 95% effective in resolution of priapism in SCD patients,[18] it does not always provide normal erectile function in the future. Anele et al.[19] reported that patients with SCD are at higher risk for ED compared to non-SCD patients (47.7% vs. 21.1%). In our report, one of eight patients had SCD and was managed with open surgical shunting in the fifth step of our treatment approach successfully and did not observed any sequela in the follow-up.

In another patient with hematological disease in our serial, hypercoagulation was detected due to mutation of factor V (MTHFR (A1298C), PAT-1 4G/5G). There are only two adult cases in the literature with priapism due to factor V mutation.[10,20] One pediatric case, with factor V Leiden mutation, experienced thromboembolic events including purple toe syndrome and skin necrosis was reported in the literature.[11] Warfarin treatment was applied and the patient experienced priapism at the 18th day of warfarin medication while his coagulation parameters were longer than normal.[11] The authors of that case had considered that priapism was a complication of warfarin therapy[11] as seen in the adult literature. To the best of our knowledge, Case 6 is the first pediatric case in the literature with priapism due to factor V Leiden mutation. We performed all steps of the treatment to achieve detumescence in this patient. Erectile failure and rigidity of cavernous bodies were observed in follow-up. We think that the main factor for this morbidity was the long duration of priapism (>24 h).

Priapism associated with hemodialysis has been reported very rarely in the adult literature,[21] and in the pediatric literature, there are only two adolescent cases reported by Shih and Wong.[22] Mechanism of the hemodialysis-associated priapism is unclear. The most commonly accused mechanisms were high hemoglobin levels after erythropoietin administration and some drugs.[23] It is also unclear whether the elevated serum testosterone and sex hormone binding globulin levels play a role in hemodialysis-associated priapism or not.[23] In this study, we report a pediatric case (8 y) with hypospadias, chronic renal failure, and hypertension, who was under alpha-methyldopa and nifedipine medication, presented with priapism during hemodialysis. His hemoglobin level was normal. To the best of our knowledge, this is the only preadolescent case and the youngest case in the literature with priapism associated with hemodialysis.

Despite all investigations, 10–50% of pediatric priapism cases remain idiopathic.[3] In our series, we could not determine an etiological factor in five patients despite our evaluation protocol.

The management of priapism should be performed with an algorithm to prevent time loss and unnecessary invasive procedures. Our treatment approach with steps is simple, useful, and beginning from less invasive method. Cold compression and analgesia (the first step) begin while the diagnostic evaluation is run. Cold provides analgesia and cytoprotective effect by limiting ischemic process and vasoconstrictive effects.[1] Paracetamol is the first choice for analgesia but opiate analgesia may also be required. Analgesia alone may provide detumescence.[24] After evaluation, if high-flow priapism is detected, this treatment approach was abandoned.

CA-AI is the second step. Aspiration and/or irrigation treatment is the most common method used for treatment. Corporal blood gas analysis is recommended as an initial test to differentiate low- or high-flow priapism in adult population.[1] In children, color Doppler USG while applying cold com-

press and analgesia (Step 1) may be more sensible due to risk to convert low-flow priapism to high flow by insertion of a needle for sampling[3] and also prevents unnecessary invasive intervention to a child. Therefore, we prefer to determine the type of priapism with color Doppler USG before an invasive attempt. After corporal aspiration, blood gas analysis was performed to confirm the diagnosis of low-flow priapism. If high-flow priapism was detected, conservative management would be the first choice instead of CA-AI. Corporal aspiration by inserting needle to corporal body laterally on the midshaft may cause injury of urethral or neurovascular bundle which was reported up to 13% of cases in the literature.[18] For that reason, we prefer to insert intravenous catheter through glans which may cause less hematoma.[25] Infection and non-ischemic priapism are other rare complications associated with corporal aspiration. It has been reported that CA-AI is successful in more than 75% of cases, especially for priapism lasting shorter than 12 h.[14] Irrigation with cold saline after aspiration is still controversial. AUA guideline stated that there was no sufficient data to support aspiration and irrigation of the cavernous bodies instead of aspiration alone.[14] AUA guideline also emphasized that after corporal aspiration, sympathomimetic injection was significantly more effective than aspiration alone in solving priapism.[14] In our report, CA-AI was performed in eight cases with one minor complication and provides successful detumescence only in one patient. The duration of tumescence was 6 h in that patient. The median duration of the priapism in our serial was 15 h (4–165 h). We think that the low success rate at this step may be result of the high duration time of tumescence or underling pathologies.

Shunts are more invasive options used for severe cases and reported as distal or proximal shunting in the literature. Distal spongio-cavernosal shunt may be performed by percutaneous (Winter shunt, T shunt, Ebbehoj, and Lue) or open (Al-Ghorab and Burnett) techniques. Proximal shunts may be spongio-cavernosal (Quackels and Sacher) or cavernovenous (by saphenous vein – Grayhack and by superficial or deep dorsal vein – Barry).[1]

Since proximal shunting is more challenging and associated with an increased number of complications, such as fistula formation and thromboembolic events, distal shunting is preferred more frequently.[14] Proximal spongio-cavernous shunting is usually preferred when distal shunts fail because of its complications such as urethral strictures, cavernourethral fistula, and cavernositis.[14]

The third step of our treatment is modified Winter shunt. In original Winter technique, distal spongio-cavernosal shunting is performed by inserting a biopsy needle percutaneously through glans to distal corpora and excision a piece of tissue from tunica and corpora to provide shunt between glans and corpus cavernosum.[26] Reoperation rate was reported 50% following spongio-cavernosal shunt in adult patients.[27] In modified Winter shunt, 16 gauge angiocatheter with additional lateral holes on it, inserting from glans through the cavernous body was used to perform a distal spongio-cavernosal shunt between glans and corpus cavernous. This approach is performed in the ward. In our series, among six patients, modified Winter shunt resolved the tumescence in 2 children (33.3%).

The fourth step is intravenous ketamine infusion and caudal epidural block. Ketamine is a detumescence agent that provides dissociative anesthesia and also analgesia and reported as an effective agent for priapism in adult literature.[3] There are case reports, including a newborn, and one series of four patients with priapism resolving with ketamine in pediatric literature.[12,13] Ketamine has an alpha-agonistic effect on peripheral vasculature besides its anesthetic and analgesic effects. This may help to normalize penile blood flow. The dissociative effect may contribute solving tumescence by blocking limbic system that involved in erectile function.[13]

Although enlargement on penile shaft after caudal block was reported in the literature,[28] caudal or epidural block with or without infusion of local anesthetics was recommended in priapism literature.[3] In our report, four children underwent ketamine anesthesia and caudal block; successful detumescence was achieved in two of them. In this step, it is unclear whether the main effect on detumescence was due to ketamine or caudal block.

Grayhack shunt (the fifth step) has been defined as a safe and effective surgical procedure and suggested as the treatment of choice for refractory low-flow priapism.[6] Complications of Grayhack shunt are hematoma, urethrocutaneous fistula, bilateral thrombophlebitis with transient pulmonary embolism, and reduction of sensibility in the perineal area.[7] In pediatric cases, Grayhack shunt is also performed successfully without major complications.[29] In our report, Grayhack shunt was required and successfully performed for resolving priapism in two children. Both of these patients had late admission after onset of symptoms. Fibrosis of the corpora and erection failure observed in one of them in long-term follow-up.

Even though intervention after 48–72 h may provide detumescence, it does not mean it could provide normal erectile function in all cases.[1] Adult literature demonstrated that priapism lasting longer than 24 h is strongly associated with ED (90%).[1] Corporal interstitial edema was found at the 12th h and necrosis of smooth muscle cell at the 48th h of ischemic priapism.[30] Evidence-based data about ED in pediatric patients in long-term follow-up are insufficient. In one patient of our serial (Case 3), the priapism lasting for 165 h was resolved firstly at the third step, but recurrence was observed after few hours. Erection failure and fibrosis were not detected in his follow-up. We think that the favorable result of this patient may be explained with partial detumescence periods during this 165 h. In our serial, one patient (case 6)

admitted with 32 h tumescence, leaved the hospital, and returned 1 day later and detumescence was achieved in his 59th h of priapism. Despite successful shunting, fibrosis of corporal bodies was observed.

The limitations of this study are the relatively small number of cases comparing with adult literature due to its rarity in pediatric population and limited follow-up time in the pediatric period. Another limitation of this study is that the erectile function could not assess properly in pediatric patients to determine the sequela.

Conclusion

Understanding and management of pediatric priapism is still a challenge due to the absence of evidence-based data. Our simple treatment approach could save time with its easy understandable and memorable structure. The steps are in order from less invasive intervention to more invasive one. Although this management seems long to maintain, steps are joined and sort by the place where the intervention performed. As Donaldson et al.[3] mentioned that "priapism is the myocardial infarct of the penis," duration of tumescence is very important. Low-flow priapism should be treated urgently with a management protocol. Therefore, we recommend our simple and useful step by step treatment approach in this emergency.

Ethics Committee Approval: This study was approved by the Marmara University Faculty of Medicine Ethics Committee (Date: 04.12.2020, Decision No: 09.2020.1334).

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ORİJİNAL ÇALIŞMA - *ÖZ*

Çocukluk çağının nadir bir acili: Priapizm ve basamaklı tedavi yaklaşımı

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AMAÇ: Priapizm çocukluk çağında çok nadir görülen bir hastalıktır. Hasta sayısının azlığı nedeniyle tedavi algoritması da halen literatürde netleşmemiştir. Bu çalışmada, priapizmli sekiz çocuk hasta ve kliniğimizde uyguladığımız basamaklı tedavi yaklaşımımız literatür ışığında sunulmaktadır.
GEREÇ VE YÖNTEM: Uyguladığımız tedavi beş basamaktan oluşmaktadır: 1. basamak; etiyoloji değerlendirmesi yapılırken soğuk kompres ve analjezi uygulaması. 2. basamak; korpus kavernozuma korporal aspirasyon ve adrenalin infüzyonu. 3. basamak; Modifiye Winter şunt. 4. basamak; ameliyathanede ketamin infüzyonu ve kaudal blok uygulaması. 5. basamak; safeno-kavernöz (Grayhack) şunt. Düşük akımlı priapizmi olan sekiz hastanın verileri geriye dönük olarak ele alındı. Başvuru şekli, tumesans süresi, yaşı, yapılan müdahaleler ve hangi basamakta detumesans sağlandığı kaydedildi.
BULGULAR: Yaş ortalaması 8.5 (1–17) yıl idi. Başvuru öncesindeki ortanca tumesans süresi 15 saat (4–165) idi. Üç hastada etiyolojik faktörler; orak hücreli anemi, kronik böbrek yetersizliği nedeniyle yapılan hemodiyaliz ve faktör V Leiden mutasyonu idi. Bir hastada ikinci basamakta detumesans sağlandı. 3, 4, 5. basamakalarda detumesans sağlanan 2'şer hasta oldu. Uzun dönem kontrolünde bir hastada kavernöz cisim rijiditesi görüldü.
TARTIŞMA: Düşük akımlı priapizm ürolojik bir acildir ve erektil disfonksiyona neden olabilir. Tedavi seçenekleri, zaman kaybını ve gereksiz invaziv işlemleri engellemek için bir algoritma veya bir tedavi protokolü ile uygulanmalıdır. Kliniğimizde uygulanan tedavi protokolü basit ve daha az invaziv işlemden daha invaziv işleme doğru gitmektedir ve düşük akımlı priapizm tedavisinde iyi bir alternatif olabilir.
Anahtar sözcükler: Çocuk; hemodiyaliz; priapizm; safeno-kavernöz şant; step-wise treatment.

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