A case report of pediatric high-flow priapism: the management of a rare condition

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ABSTRACT

Background: High-flow priapism is extremely rare, especially in childhood and usually occurs after a penile or perineal trauma. It must be differentiated from low-flow priapism, which requires urgent treatment. Treatment options of high-flow priapism include conservative approach, embolization, and surgical ligation.

Case Presentation: A 6-year-old boy presented with a semi-rigid and painless penile erection. With the diagnosis of high-flow priapism, he underwent conservative therapy. After 4 weeks of conservative treatment, priapism was no longer existed, and no recurrence occurred during the 2-year follow-up.

Conclusion: Conservative approach should be applied before the invasive interventions as it is easy to apply and has no side effects.

Keywords: Priapism, child, high-flow, penile trauma, case report.

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Background

Priapism is defined as a penile erection that occurs independently of a sexual stimulation and lasts longer than 4 hours [1]. There are three subtypes: low flow (ischemic, veno-occlusive), high flow (non-ischemic, arterial), and stuttering (recurrent) priapism. Low-flow priapism is the most common subtype. It is associated with venous occlusion and may cause hypoxia, necrosis, and fibrosis in the cavernous tissue. It is, therefore, considered as a urological emergency. High-flow priapism is rare and is usually associated with penile or perineal trauma. It is characterized by arterio-cavernous fistula development and increased cavernosal blood flow. It is not a urological emergency since ischemia does not occur. Stuttering priapism is a recurrent and self-limiting condition and is often associated with sickle cell disease [1].

Treatment options of high-flow priapism include conservative approach, embolization, and surgical ligation [2]. To avoid possible side effects of invasive treatments in children with high-flow priapism, we emphasized the high success of the conservative approach.

In this study, we aimed to present a rare case of pediatric high-flow priapism and to review the differential diagnosis and treatment options.

Case Presentation

A 6-year-old boy was admitted to our clinic with the complaint of penile erection lasting 2 days. He had no history of systemic disease. In his story, it was learned that he fell off his bike 5 days ago and had horse-riding injury to perineum. Physical examination revealed a semi-rigid and painless penile erection. It was observed that detumescence was achieved with applying pressure to the perineum (Figure 1), and erection occurred again with the removal of the pressure (Figure 2) (Piesis sign). While erection was occurring, pulsation caused by arterial flow in the penis could be visualized. No murmur was detected with perineal auscultation. In penile color doppler ultrasonography (CDU), the peak systolic velocity in the right cavernous artery was measured as 150 cm/second (Figure 3) and an arterio-cavernosal fistula was detected (Figure 4). A pediatrician was consulted to rule out other possible causes. Sickle cell disease or leukemia was not detected. With the diagnosis of high-flow priapism developing after trauma, it was decided to apply conservative treatment, initially. His parents were asked to perform perineal compression until achieving detumescence and apply ice packs twice a day for 30 minutes. After 2 weeks, priapism was still present. In the CDU, it was observed

Figure 1. *Penile detumescence after perineal compression.*

Figure 2. *Penile erection occurs after the removal of perineal compression.*

that the peak systolic velocity in the right cavernous artery decreased to 25 cm/second, but the fistula formation persisted (Figure 5). Conservative treatment was applied for two more weeks. After 4 weeks of conservative treatment, priapism was no longer existed and normal cavernosal blood flow was observed in the control CDU. No recurrence occurred during the 2-year follow-up.

Discussion

Due to the differences in treatment approaches, the subtype of priapism should be determined initially. Low-flow priapism is an ischemic process and requires urgent treatment due to the risk of cavernous tissue fibrosis and permanent erectile dysfunction (ED) [1]. In high-flow priapism, venous flow is not impaired, and there

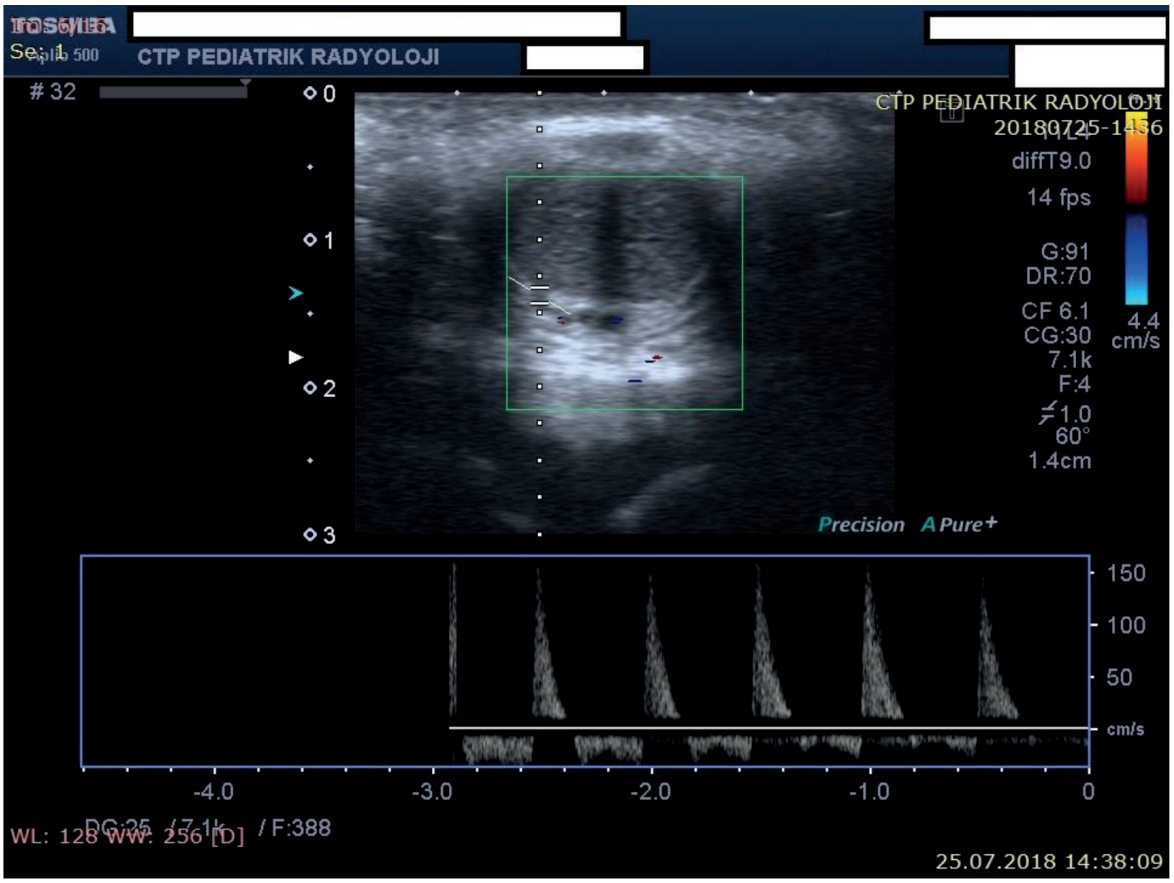

Figure 3. *Intracavernosal increased arterial blood flow [Peak systolic velocity (PSV): 150 cm/sn].*

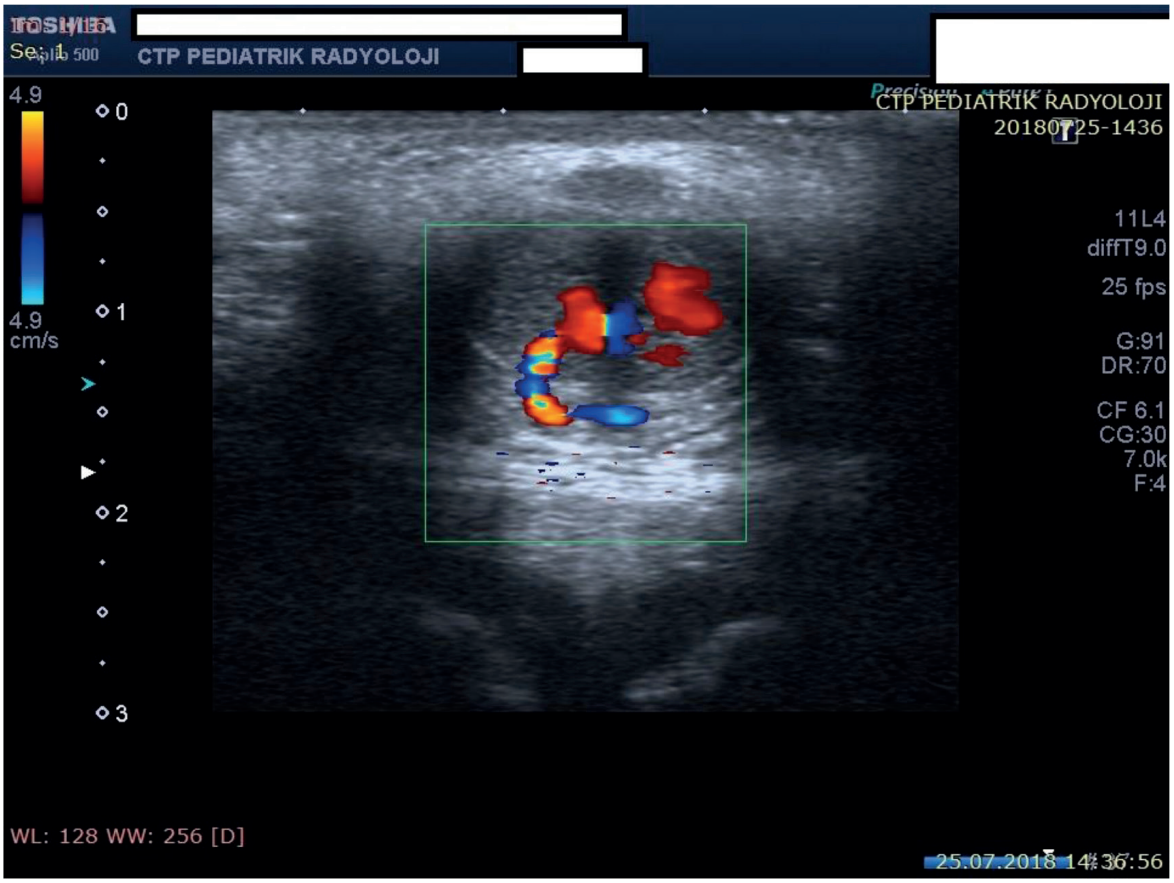

Figure 4. *Arterio-cavernous fistula.*

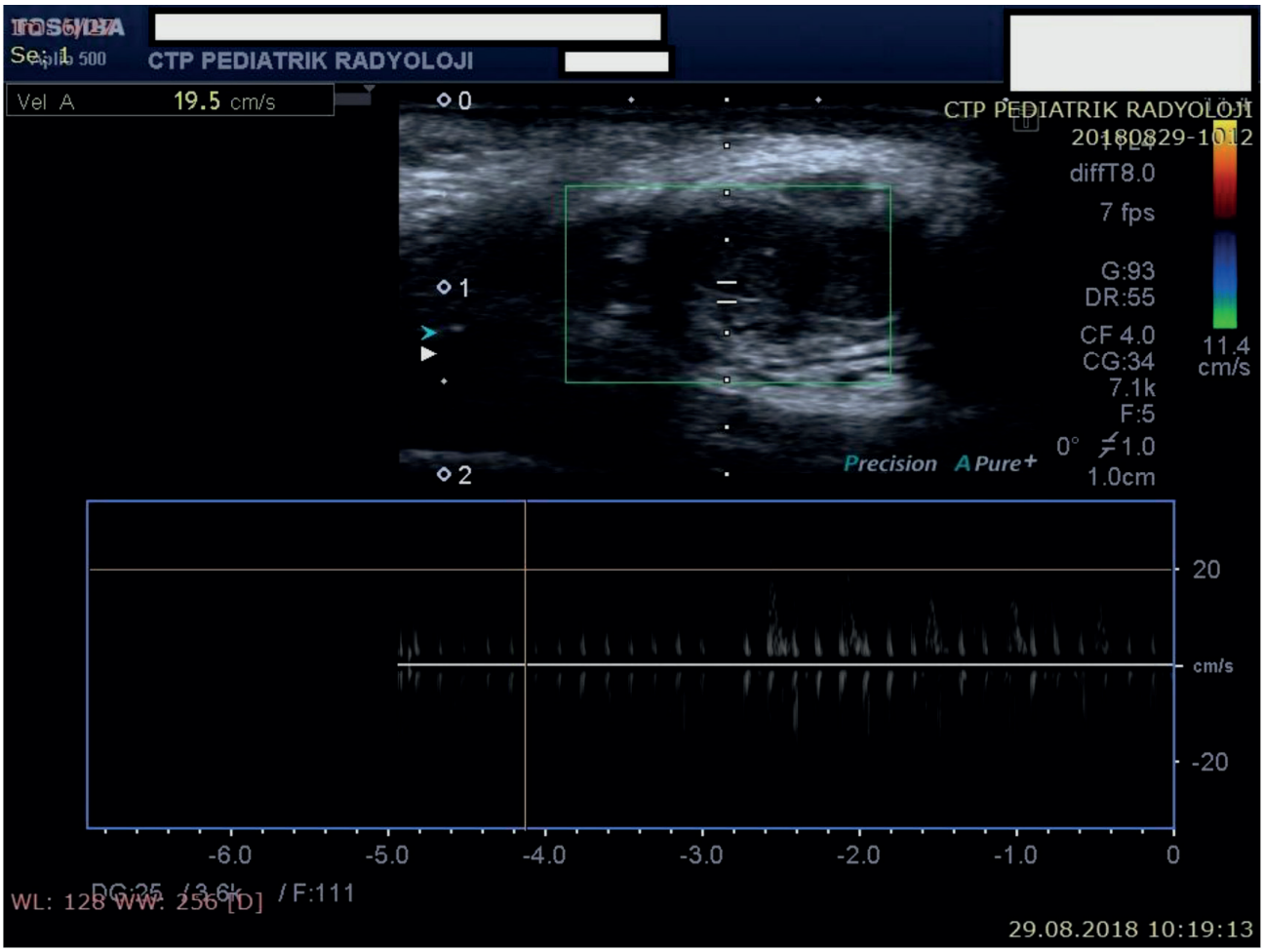

Figure 5. *Intracavernosal arterial blood flow after 2 weeks of conservative therapy (PSV: 25cm/sn).*

is an increase in arterial flow. Therefore, erectile tissues are well oxygenated and do not require an urgent management [3]. History, physical examination, cavernosal blood gas analysis, and penile CDU are useful for differential diagnosis. Pain is the most important symptom in low-flow priapism. The cavernous tissues have a rigid erection, while the glans and cancellous tissue are flaccid [4]. In high-flow priapism, there is no pain, and the erection is semirigid. It usually occurs after a history of penile or perineal trauma. Priapism is observed a few hours or days after the trauma [5]. In cavernosal blood gas analysis, hypoxia, hypercapnia, and acidosis are observed in low-flow priapism, while high-flow priapism has increased partial O_2 pressure with arterial blood characteristics [6]. High blood flow in the cavernous body and arterio-cavernous fistula may be shown in high-flow pripaism with CDU. In low-flow priapism, blood flow can not be monitored due to venous occlusion [7]. Penile arteriography can also be used to show arterio-cavernous fistula or pseudoaneurysm, but it is an invasive procedure and should be applied if embolization is preferred for the management.

High-flow priapism is an extremely rare condition in childhood. It often occurs after a penile or perineal trauma [5]. In sickle cell disease, recurrent attacks of priapism are usually observed as low-flow priapism with venous occlusion, while less frequently high-flow priapism may also occur. Therefore, every patient should be consulted with a pediatric hematologist [8]. The pathophysiology is arterial wall rupture caused by trauma, development of fistula formation and high arterial blood flow into the cavernous body [3]. The increase in the partial O_2 pressure increases the synthesis and release of nitric oxide in the corpus cavernosum [9]. Priapism occurs a few hours or days after trauma. Usually a nocturnal erection is the trigger [5]. In our case, priapism occurred 48 hours after perineal trauma. It is typically painless with a semi-rigid erection. Piesis sign (detumescence with perineal compression and re-erection with removal of compression) is typical for high-flow priapism [10]. In cavernous blood gas analysis, pH> 7.3, pO_2> 50 and blood is bright red in color. However, since it is an invasive procedure, it should be applied if low-flow priapism is suspected in the differential diagnosis [6]. Penile CDU is a non-invasive test that may show high blood flow and fistula within the cavernous body. Typically, there is a continuous arterial blood flow and no occlusion of the venous flow [7].

In our case, we did not require cavernosal blood gas analysis because of typical examination and penil CDU findings.

Treatment options include conservative approach, embolization, and surgical ligation [2]. The aim is to provide detumescence and preserve erectile function. As the conservative approach, ice packs and perineal compression are used to reduce cavernosal blood flow velocity and it is thought that this will result in thrombus formation within the fistula [4]. Success rates up to 62% have been reported in the literature. [2]. There is no consensus on how long this approach should be applied. Corbetta et al. [11] successfully treated three patients with conservative approach and reported the treatment durations of these three patients as 14, 27, and 36 days, respectively. Burns et al. [12] also stated that the conservative approach can be applied safely for 6 weeks. In our case, we discussed the treatment options with the family and preferred conservative approach as initial treatment. We achieved full detumescence after 28 days.

Selective arterial embolization may be preferred as a second-line treatment in cases where conservative approach is not successful. The success rate in the first session was found to be 80% in 30 patients, and full detumescence was achieved in 5 of 6 patients in repeated sessions. Therefore, the overall success rate of embolization was reported as 97% [4]. For embolization, autologous clot, gel foam or sponge, or a more permanent material microcoils may be used. It is thought that the risk of permanent ED is less with the temporary materials, as they will melt over time and allow re-flow of blood in the embolized artery and cavernous tissue. However, there is no prospective study about which embolic agent is superior [13]. Although repeated sessions may be needed, complication rates and risk of ED after embolization are quite low [14]. The disadvantages of embolization include being invasive, needing re-embolization sessions, requiring anesthesia and exposure to ionizing radiation.

Surgical ligation may be considered as the last option in cases where full detumescence cannot be achieved despite repeated embolization sessions. This technique, which has a success rate of 63%, should always be considered at the end of the treatment algorithm because it is highly invasive and has a high risk of ED [13,15].

Conclusion

High-flow priapism is not a urological emergency. Therefore, low-flow priapism, that requires urgent treatment, should be ruled out initially. Conservative approach can be applied safely for 6 weeks before the invasive interventions. As seen in our case, it is easy to apply, has no side effects and has high success rates. If complete detumescence can not be achieved with conservative approach, selective arterial embolization should be preferred prior to surgical ligation due to its high success rates and low risk of ED.

What is new?

The authors present a rare case of pediatric high-flow priapism and review the differential diagnosis and treatment options. To avoid possible side effects of invasive treatments in children with high-flow priapism, emphasized the high success of the conservative approach.

List of Abbreviations

CDU Color doppler ultrasonography
ED Erectile dysfunction

Conflicts of interest

The authors declare that there is no conflict of interest regarding the publication of this case report.

Funding

None.

Consent for publication

Written consent was obtained from the parents of the patient.

Ethical approval

Ethical approval is not required at our institution to publish an anonymous case report.

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Summary of the case

1	**Patient (gender, age)**	Male, 6 years old
2	**Final diagnosis**	High-flow priapism
3	**Symptoms**	Semi-rigid and painless penile erection
4	**Medications**	None
5	**Clinical procedure**	Conservative approach applied with ice packs and perineal compression
6	**Specialty**	Pediatric urology