



# Management of Neonatal Priapism: Report of Two Cases and Review of the Literature

## *Yenidoğan Priapizmine Yaklaşım: İki Olgu Bildirimi ve Literatür Derlemesi*

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### ABSTRACT

Priapism in a newborn is a rare entity with only 15 cases reported in the literature since 1879. The most commonly detected etiologic factor is polycythemia, but most of the cases are idiopathic. Conservative treatment seems to be sufficient and surgical approach is usually unnecessary. Here, we present a prolonged erection in two newborns which occurred at the first day of life and detumescence was achieved with conservative approach at the 6<sup>th</sup> and 7<sup>th</sup> day of life, respectively. We also reviewed the literature to investigate the most adequate evaluation and management criteria.

### Keywords

Idiopathic, conservative, priapism, neonatal, management

### ÖZ

Yenidoğan priapizmi nadir görülen bir hastalık olup 1879'dan şimdiye kadar literatürde 15 olgu bildirimi yapılmıştır. En sık rastlanan etiyolojik faktör polisitemi olsa da, çoğunlukla idiyopattır. Yenidoğan priapizmi genellikle iskemik olmayan tipte seyredir. Tedavide çoğu zaman cerrahi müdahaleye gerek kalmaz. Konservatif yaklaşım en iyi seçenek olarak görülmektedir. Bu olgu sunumunda konservatif izlem ile yaşamın 6. ve 7. günlerinde gerileyen iki yenidoğan priapizm olgusunu sunulmaktadır. Olgu sunumu ile beraber en uygun yaklaşım ve değerlendirme için yol göstermesi amacıyla literatür derlemesi de sunulmuştur.

### Anahtar Kelimeler

İdyopatik, konservatif, priapizm, yenidoğan, yaklaşım

## Introduction

Preservation of normal erectile function is the major goal in the management of priapism. Ischemic priapism is a urological emergency due to compartment syndrome after 4 hours of prolonged erection. However, ischemic priapism has not yet been reported in a neonate. Priapism in newborn is challenging situation for pediatric urologists because of the lack of experience due to paucity of cases, and well-established guidelines. Therefore, evaluation and management strategy remains unclear. Here, we present the latest two cases of newborn priapism and review of the literature.

## Case Presentation

First case was the second child of a 31-year-old woman. The newborn was delivered by cesarean section at the 39<sup>th</sup> week of gestation with a birth weight of 2870 g. Appearance, Pulse, Grimace, Activity, Respiration (APGAR) test scores were 9/10 and 9/10 at 1 and 5 minutes, respectively. There was no evidence of blood group

incompatibility and hematological pathology in the newborn. Blood count, biochemical analysis, and physical examination revealed no abnormality and, neurological examination was normal. Maternal hypothyroidism, for which she used levothyroxine, and a history of smoking during pregnancy was noted. Routine prenatal examinations were performed regularly. The patient was referred to the pediatric urology department at the second day of life with prolonged erection which was noticed at the first day of life by the parents. Physical examination showed rigid penis without cyanosis and tenderness (Figure 1). The testes were descended bilaterally and normal on physical examination. Projectile voiding was observed without restlessness and, defecation was normal. Penile Doppler ultrasound scan revealed normal arterial and venous flow and there was no evidence of ischemic priapism. Polycythemia and other hematological disorders were ruled out after pediatric hematology consultation. The patient underwent conservative follow-up. Spontaneous detumescence was achieved at the 6<sup>th</sup> day of life without any surgical intervention. His course was uneventful. No recurrence and abnormality was observed during 6 months of follow-up.

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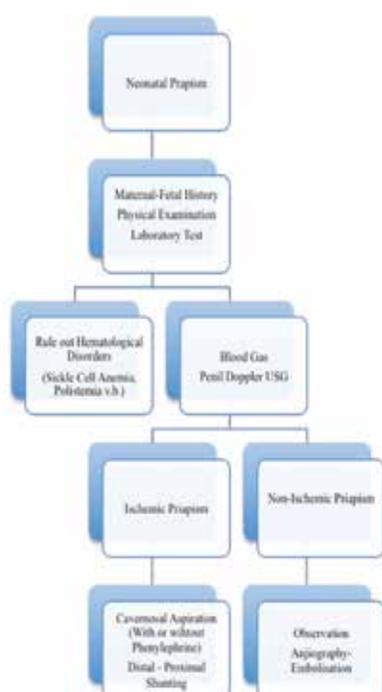
The second case was the first child of a 28-year-old woman. The newborn was delivered vaginally at the 40<sup>th</sup> week of gestation with a birth weight of 3280 gr. APGAR scores were 9/10 and 10/10 at 1 and 5 minutes, respectively. Routine neonatal evaluation was normal. Maternal smoking history was denied. The patient was referred to the pediatric urology department at the first day of life with prolonged erection. Physical examination showed a rigid penis without cyanosis or tenderness (Figure 1). The testes were descended bilaterally and were found to be normal on physical examination. Hematological evaluation showed no pathology. Penile Doppler ultrasound imaging was not performed for this case. Finally, the detumescence was achieved on the 7<sup>th</sup> day of life without any surgical intervention. No recurrence and abnormality was observed during 3 months of follow-up.

## Discussion

Priapism is rare in children, especially in neonates. Fifteen cases have been reported in the literature since 1879 (1,2,3,4,5,6,7,8,9,10).



**Figure 1.** Prolonged penile erection of the first case at the 4<sup>th</sup> days of life (A) and prolonged penile erection of the second case at the first day of life (B)



**Figure 2.** Algorithm of neonatal priapism management

Estimated incidence of idiopathic neonatal priapism is 15 per 100,000 live births based on a surveillance study performed between 1974 and 1988 (1). Current experience on neonatal priapism is based on mostly similar individual case reports. Thus, evaluation, treatment and follow-up are challenging in pediatric urology practice. We present two cases of neonatal priapism treated in our department. These cases represent our first experience on this rare disorder. Since a limited number of cases have been reported, no clinical guideline has been established for neonatal priapism. We review the literature to propose a clinical approach for these cases (Table 1).

The most common cause for newborn priapism is idiopathic (8,9). However, other etiologic factors should be excluded. The possible causes include blood dyscrasias, malignancy, trauma, neurological pathologies and drug-related side effects (11). Although, sickle cell anemia is common in children, it is negligible in neonates because of the predominance of fetal hemoglobin. Among them, polycythemia is the most known detected etiology (3,7). In the literature, there are 4 reported cases of neonatal priapism attributed to polycythemia and one to blood transfusion, where all other 10 cases were idiopathic (Table 1). Polycythemia was ruled out in our two cases and there was also no any other significant finding in evaluation. Therefore, we also defined our cases as idiopathic. We detected maternal hypothyroidism and a history of maternal smoking during pregnancy in our first case. Our management of neonatal priapism algorithm is shown in Figure 2.

Neonatal priapism was non-ischemic in all reported cases. Our two cases were also non-ischemic priapism. Increased blood flow may be the cause of priapism in cases with polycythemia. But non-ischemic course of priapism was seen in the reported 4 cases with polycythemia. We support the hypothesis of Meijer and Bakker (2) that neonatal priapism should be the form of non-ischemic priapism because this form presents with a painless erection and conservative management is possible. Therefore, Doppler ultrasound and cavernosal blood gas analysis can be reserved for cases suspicious for ischemic priapism.

Treatment of this pathology is not clear yet. Suggested management options include observation, intravenous ketamine admission, phlebotomy and exchange transfusion. Majority of cases were managed with observation, and spontaneous detumescence was achieved. However, intravenous ketamine admission in one case (3), phlebotomy in one case and exchange transfusion in one case (4) have been reported. Intravenous ketamine admission and exchange transfusion resulted in immediate detumescence where it occurred four days after phlebotomy. In all other cases including ours, detumescence was achieved mostly in 5 to 6 days. Follow-up showed no abnormality, although we don't have long term results. Therefore, observation should be the first approach in idiopathic neonatal priapism.

## Ethics

Informed Consent: Consent form was filled out by all participants. Peer-review: Internal peer-reviewed.

## Authorship Contributions

Surgical and Medical Practices: Haydar Kamil Çam, Cem Akbal, Mehmet Özyay Özgür, Çağrı Akın Şekerci, Concept: Cem Akbal, Çağrı Akın Şekerci, Design: Cem Akbal, Çağrı Akın Şekerci, Data Collection or Processing: Çağrı Akın Şekerci, Farhad Talibzade, Analysis or Interpretation: Çağrı

Table 1. Data of 17 cases [Rearranged with additional four cases from reference (2) and two cases from reference (5)]

Reference	Onset	Treatment	Duration of priapism (days)	Proposed etiology	Follow up
Humbert et al. (12)	Day 1	Observation	2	Polycythemia	Normal
	Day 4	Phlebotomy	5	Polycythemia	Normal
Larocque and Cosgrove (6)	Not reported	Observation	4	Polycythemia	Not reported
Amlie et al. (7)	Day 37	Observation	12	Blood transfusion	Normal
Leal et al. (8)	At birth	Observation	6	Idiopathic	Normal
Shapiro (9)	At birth	Observation	3	Idiopathic	Not reported
Merlob and Livne (1)	At birth	Observation	6	Idiopathic	Normal
	Day 1	Observation	5	Idiopathic	Normal
	Day 5	Observation	4	Idiopathic	Normal
	Day 1	Observation	5	Idiopathic	Normal
Stothers and Ritchie (10)	At birth	Intravenous ketamine	3	Idiopathic	Normal
Walker and Casale (3)	At birth	Exchange transfusion	4	Polycythemia	Normal
Meijer and Bakker (2)	Day 1	Observation	4	Idiopathic	Normal
Burgu et al. (4)	Day 1	Observation	3	Idiopathic	Normal
Aktoz et al. (5)	Day 1	Observation	4	Idiopathic	Normal
Marmara	Day 1	Observation	6	Idiopathic	Normal
Marmara	Day 1	Observation	7	Idiopathic	Normal

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