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CASE REPORT

Priapism in the newborn: management and review of literature

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Summary

Priapism is a pathological condition of a penile erection that persists beyond or is unrelated to sexual stimulation. Priapism is broadly classified into two types: (i) ischaemic priapism (veno-occlusive) (low-flow), (ii) nonischaemic priapism (arterial) (high-flow). We report the case of a newborn presenting with priapism on the first day of life and also review published data on the management and follow-up of this condition.

Introduction

Priapism is a pathological condition of a penile erection that persists beyond or is unrelated to sexual stimulation. Priapism is an important medical condition, which requires evaluation and may require emergency management (Berger *et al.*, 2001).

Priapism is broadly classified into two types. (i) Ischaemic priapism (veno-occlusive) (low-flow) is the most common form of priapism; it is usually a painful, rigid erection characterised clinically by absent cavernous blood flow. Ischaemic priapism beyond 4 h is a compartment syndrome requiring emergent medical intervention. (ii) Nonischaemic (arterial) (high-flow) priapism is a less common form of priapism caused by unregulated cavernous inflow. The erection is usually painless and not fully rigid. Nonischaemic priapism requires evaluation (Raveenthiran, 2008). High-flow priapism can cause persistent high oxygen tension and venoocclusion (similar to low-flow priapism) within the corpora, resulting in corporeal fibrosis and ultimately permanent erectile dysfunction (Sandler *et al.*, 2008).

Preservation of normal erectile function is a major goal in the management of priapism (Meijer & Bakker, 2003; Wisard *et al.*, 2007). We report the case of a newborn presenting with priapism on the first day of life and also review published data on the management and follow-up of this condition.

Case report

The patient was born to a 30-year-old gravida 2, para 1, mother. Routine pre-natal screens had been normal including no evidence of blood group incompatibility. The vaginal delivery was uncomplicated. The newborn's birth weight was 3100 ***g*** and Apgar scores were eight and nine at 1 and 5 min respectively. The initial physical examination was normal. The testes were descended and the penile shaft and scrotal were normal in colour and appearance. One day after birth, we had noticed that the infant had a persistent erection (Fig. 1). The physical examination revealed an erect penis without signs of cyanosis or tenderness. Serum platelet count, white blood cell count, glucose, electrolyte and bilirubin were within normal limits. The newborn voided frequently with a good stream and was in no apparent distress. The priapism disappeared after 4 days of observation. The patient continued to show repeated erections upon the slightest

stimulation following flaccid states within several minutes. The physical and neurologic examinations at the time of discharge were normal. The penis was normal at multiple follow-up visits at 12 months after birth.

Discussion

Newborn males will frequently have erections with the slightest tactile stimulation. Erections are usually triggered by a full bladder. Typically, this physiological erection lasts a couple of minutes and disappears quickly after the withdrawal of the stimulus (Burgu *et al.*, 2007). Erections demonstrate that the nerves to the penis are normal. Merlob & Livne (1989) discussed that the term 'neonatal priapism' was not appropriate and they recommended to call this condition as 'prolonged penile erection in the newborn'.

Most of the newborns with persistent penile erection are likely to have an idiopathic aetiology. However, when we consider the identifiable aetiologies, polycythemia may be the most common. Polycythemia can result in hyperviscosity and subsequent sludging in the microvasculature. It may be this sludging that results in decreased penile venous outflow and persistence of a penile erection. In older children, reported associations include sickle cell disease, birth trauma, respiratory distress syndrome, umbilical artery catherisation, metabolic hypoxia, parenteral nutrition and drugs (heparin, hydrochlorothiazide and others) (Walker & Casale, 1997).

Two forms of priapism can be distinguished: ischaemic (veno-occlusive) (low-flow) and nonischaemic (arterial) (high-flow). Two forms of priapism can be discriminated by performing a duplex Doppler ultrasound examination of the penis or by blood gas analysis of aspirate from the cavernosal bodies. The main concern in the management of neonatal priapism is the preservation of erectile function (Meijer & Bakker, 2003).

The first case of priapism in a neonate was described by Nuckols (1876), which was thought to be related with congenital syphilis. Since 1876, 15 cases of priapism or prolonged penile erection in the newborn have been reported. The onset, treatment, duration of priapism, proposed cause and follow-up of all 15 cases are presented in Table 1.

In these 15 cases, there was no pain or distress reported at the time of priapism. In 12 cases, including two cases with polycythaemia and the case with priapism after a blood transfusion, the condition was managed by observation alone.

In one polycythaemic newborn, the choice of treatment was phlebotomy and a total of 50 ml blood was drawn from a peripheral vein (Humbert *et al.*, 1969). In another polycythaemic newborn, 40 ml of blood was exchanged for an equivalent amount of plasma protein fraction. Immediately after completion of the exchange transfusion, detumescence was noticed (Walker & Casale, 1997).

In children, the most common cause of priapism is sickle cell disease. This is characterised by predominance of sickle haemoglobin. Priapism develops in 27.5% of children with sickle cell disease (Mantadakis *et al.*, 1999). The priapism is generally related to a low-flow state, secondary to sickling of red blood cells within the sinusoids of the corpora cavernosa, resulting in venous stasis. This situation results in decreased local oxygen tension and pH, which potentiate further stasis and sickling (Bruno *et al.*, 2001). The pain that is experienced, is a sign of ischaemia. In sickle cell patients, tricorporeal priapism has also been described. Intervention in the form of icepacks, analgaesia, sedation, hydration, oxygenation and blood transfusion may be helpful in the initial 10–12 h of

Table 1 Clinical data of 15 neonates with priapism (this table was adapted with additional two new cases from reference 9)

Reference	Onset	Treatment	Duration of priapism (days)	Proposed cause	Follow-up
Humbert *et al.* (1969)	Day 1	Observation	2	Polycythaemia	Normal
	Day 4	Phlebotomy	5	Polycythaemia	Normal
Larocque & Cosgrove (1974)	Not reported	Observation	4	Polycythaemia	Not reported
Amlie *et al.* (1977)	Day 37	Observation	12	Blood transfusion	Normal
Leal *et al.* (1978)	At birth	Observation	6	Idiopathic	Normal
Shapiro (1979)	At birth	Observation	3	Idiopathic	Not reported
Merlob & Livne (1989)	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 & Ritchie (1992)	At birth	i.v. ketamine	3	Idiopathic	Normal
Walker & Casale (1997)	At birth	Exchance transfusion	4	Polycythaemia	Normal
Meijer & Bakker (2003)	Day 1	Observation	4	Idiopathic	Normal
Burgu *et al.* (2007)	Day 1	Observation	3	Idiopathic	Normal
Present case	Day 1	Observation	4	Idiopathic	Normal

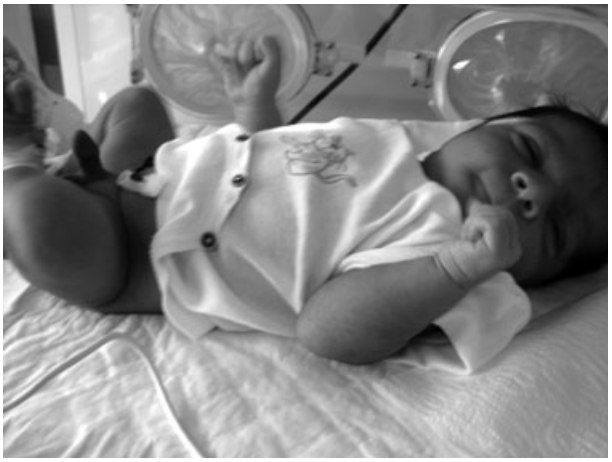

Fig. 1 Priapism in a newborn.

symptoms. If the priapism remains unresolved, surgical intervention is indicated (Shah *et al.*, 2004).

Stothers & Ritchie (1992) administered 1% ketamine hydrochloride solution intravenously (total amount of 0.3 ml), under anaesthetic supervision in an operating theatre, in a 3.6-kg newborn. Treatment of idiopathic priapism with intravenous ketamine hydrochloride produced rapid detumescence. It has been described as an alternative method of management for this condition.

In 13 cases, information about follow-up was provided, 10 of these cases were managed by observation alone. The longest follow-up was 2–8 years in the literature and this follow-up revealed no urogenital problems with normal erections in the pre-pubertal period, but a longer period is needed (at least until puberty) (Merlob & Livne, 1989).

Conclusion

Management of priapism in the newborn by observation alone seems to be the best approach. If newborn's priapism is associated with polycythaemia, red cell volume reduction should be the choice of treatment. Intravenous administration of ketamine hydrochloride could be considered before surgical intervention under anaesthetic supervision. Informed consent must include the possibility of surgical intervention to avoid any medicolegal complications.

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