International Surgery Journal
Emmanuel J et al. Int Surg J. 2021 Oct;8(10):3154-3156
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: https://dx.doi.org/10.18203/2349-2902.isj20214014

Case Report

Neonatal priapism: first reported case in South East Asia and literature review

John Emmanuel[1]*, Siti Maisarah Razali[1], Muhammad Asyraf Azmi[1], Banuprritaa Veejeyahshegarun[2], Michael Arvind[2], Chow Lai Yin[3]

[1]Department of Surgery, Hospital Raja Permaisuri Bainun, Ipoh, Malaysia
[2]Department of Surgery, [3]Department of Paediatrics, Hospital Teluk Intan, Malaysia

Received: 25 July 2021
Accepted: 02 September 2021

***Correspondence:**
Dr. John Emmanuel,
E-mail: johnegf82@gmail.com

ABSTRACT

Neonatal priapism varies significantly in aetiology and clinical course compared to older children and adults. This entity is rare and limited to case reports. Due to its rarity, management options are not well defined and this leads to variations in treatment practices. Almost all cases are idiopathic and achieve detumescence following a minimalist approach. We reported a case of neonatal priapism on day 1 of life and review the literature available.

Keywords: Neonatal, Priapism, Infant

INTRODUCTION

Neonatal priapism is a rare entity and with the cause being idiopathic in most cases. The true incidence is unknown. Most of these cases do not require intervention and detumescence occurs within days.[1]

CASE REPORT

Our baby was a case of birth before arrival (BBA) with a gestational age of 36 weeks and a birth weight of 2.59 kgs. Antenatally, mother had gestational diabetes mellitus (GDM) and was on oral hypoglycemic agents (OHA). He had a good cry at birth and was wrapped and attended to by ambulance personnel by about 10 minutes of life. Cord was cut by the house officer (HO) in the ambulance and child was taken to the hospital and nursed in the special care nursery (SCN). He was started on antibiotic coverage for presumed sepsis due to the late prematurity and BBA. Newborn examination was normal including a normal penile shaft, scrotum and both testes descended. At 25 hours of life, the child was noticed to have a persistent erection. Examination revealed an erect penis without cyanosis, discoloration or tenderness. The child was not in distress and was able to void frequently. Laboratory parameters including complete blood counts were within normal limits. Doppler sonography of the penis was normal. There was progressive detumescence and the penis was completely flaccid by day 5 of life. Outpatient follow up at one and three month revealed a normal penis.

DISCUSSION

Newborn males commonly have erections which were triggered by a distended urinary bladder, tactile stimulation, diaper changing, bathing and urethral catheterization.[1] These erections quickly subsided following withdrawal of the precipitating factor. These physiological erections also occured spontaneously and demonstrated that the nerves to penis were normal.[2]

Priapism is defined as a full or partial erection that persists more than 4 hours beyond sexual stimulation or

is unrelated to sexual stimulation.[3] In both adults and children, priapism is broadly classified into 3 types, ischaemic (low flow, veno occlusive), non ischaemic (high flow, arterial) and stuttering (intermittent, recurrent ischaemia).[4] Ischaemic priapism is always painful, however in non ischaemic priapism, pain is not a common feature.[5]

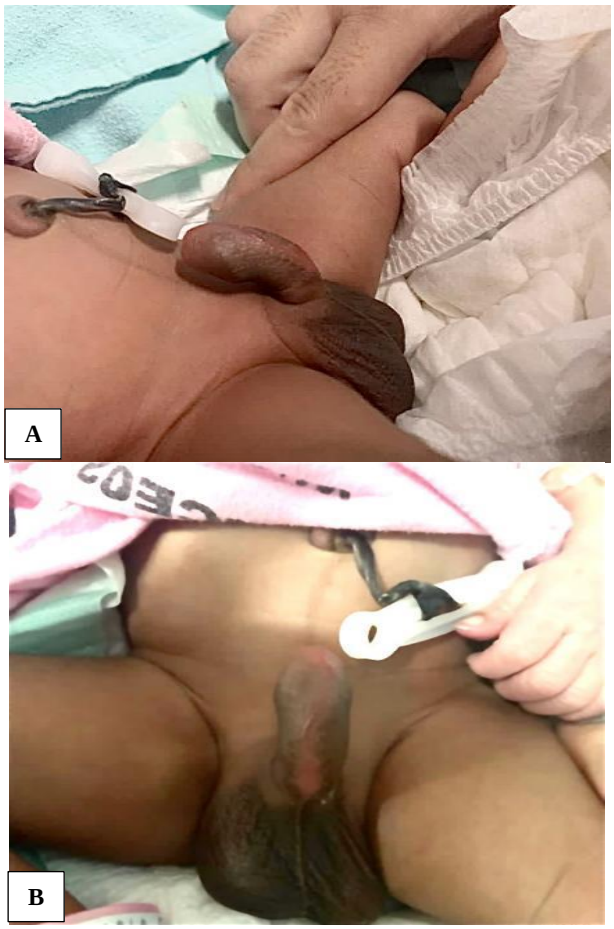

Figure 1: (A and B) Shows the priapsim experienced by the neonate in our case report; permission acquired from parents for publication of these photos.

Neonatal priapism was a rare phenomenon. Its true incidence was unknown.[6] In a small study, Merlob et al quoted a rate of 1 case in 6673 male newborns between 1974 to 1988.[7] Available literature on this pathology was limited to less than 30 case reports.[1] These reports were spread across the various continents including one in Asia. Sood et al reported a case of neonatal priapism in India in 2006.[8] However, our literature search did not reveal any reported cases from the South East Asian region. As most of these cases achieve spontaneous detumescence, it was likely that its rarity was contributed partially by under reporting of a condition with a benign course.

70% of paediatric priapism was caused by sickle cell disease which was characterised by sickle haemoglobin. However this was not applicable to neonates due to the abundance of fetal hemoglobin.[9] Priapism in neonates was generally idiopathic.[5] Other causes included polycythaemia, infection, cranial birth trauma, respiratory distress syndrome (RDS) and drugs (sildenafil, iNO).[4] The most common among the identifiable causes was polycythaemia.[10] Increased blood viscosity and sludging can impede outflow of blood from the penis which resulted in priapism. Most cases of neonatal priapism presented with similar characteristics. These erections began on day one or two of life and lasted an average duration of four to five days.[6,9] Genital examination revealed no discolouration of the penis and scrotum and the neonate did not seem to be in pain.

The diagnosis was mainly based on history (including drug aetiology) and clinical examination.[11] Common causes should be ruled out including polycythaemia and infection. First line investigations included blood counts and C-reactive protein.[1,4] Neonatal priapism was thought to be a form of non ischaemic priapism. Colour Doppler ultrasonography (CDU) and cavernous blood gas can be used to distinguish ischaemic from non ischaemic priapism. However, these were infrequently used in infants and may be difficult to perform.[11] Dust et al performed the first ever reported cavernous blood gas analysis in a newborn who had a persistent erection after 72 hours.[9] This resulted in a slight reduction of the erection. The benefit of this was debatable as a prior CDU indicated normal arterial and venous flow and the child was treated conservatively. Due to the favourable outcome of neonatal priapism, we advocated against doing invasive procedures like cavernous blood gas analysis. The non-invasiveness of CDU made it a safer option in neonates.

Management of these cases were not well standardized owing to its rarity. Lack of experience in evaluation and management caused a significant concern amongst doctors dealing with this entity. Majority of the reported cases were managed conservatively and achieved spontaneous detumescence within a period of 5 days.[1,10] Medical or surgical intervention should be reserved for scenarios where the priapism lasted more than 5 days.[6] Intravenous ketamine had been described as an option with the child achieving spontaneous detumescence.[10,11] Fannie et al cautions against the use of ketamine citing its possible role in neuroapoptosis in newborns.[1] Venesection and partial exchange transfusion were reported to be successful in polycythaemia associated priapism.[12,13] Alternatively, a conservative approach had also been successful in two cases associated with polycythaemia.[9] Hence, we proposed the theory that if given adequate time to resolve, the need for any intervention might be negated.

One of the major concerns in neonatal priapism was the preservation of erectile function. Mismanaged and recurrent priapism can lead to erectile dysfunction, however these had not been reported in the neonatal cohort. A review by Meijer in 2003 documented a follow

up period between 4 weeks to 8 years.[5] All these newborns had normal erections throughout their follow up period. However, these and other available case reports did not document follow up into the adolescent period. Despite this, it was our believe that a case of erectile dysfunction secondary to neonatal priapism would had been reported had it occurred.

Irrespective of the cause, duration or treatment, reported cases showed a favourable outcome. Hence, some authors have proposed to use the term persistent penile erection of the newborn to accurately reflect the benign course of the condition in reported cases.[7,12] However proponents of this had pointed to the limited case reports available and the short follow up period to safely use the term (dust).

CONCLUSION

Most cases of neonatal priapism achieve spontaneous detumescence without the need for intervention. Masterly inactivity should be the initial approach when managing these cases with early escalation of referral to a paediatric urologist/surgeon when in doubt. Erectile dysfunction has not been reported in the follow up of these newborns however further long term studies are warranted to accurately define this positive outcome.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Fanni C, Marcialis MA, Pintus MC, Loddo C, Fanos V. The first case of neonatal priapism during hypothermia for hypoxic-ischemic encephalopathy and a literature review. Italian J Pediatr. 2018;44(1):1-7.
2. Aktoz T, Tepeler A, Gündogdu EO, Ozkuvanci U, Müslümanoglu AY. Priapism in the newborn: management and review of literature. Andrologia. 2011;43(1):65-7.
3. Broderick GA, Kadioglu A, Bivalacqua TJ, Ghanem H, Nehra A, Shamloul R. Priapism: pathogenesis, epidemiology, and management. J Sex Med. 2010;7:476-500.
4. Donaldson JF, Rees RW, Steinbrecher HA. Priapism in children: a comprehensive review and clinical guideline. J Pediatr Urol. 2014;10(1):11-24.
5. Meijer B, Bakker HHR. Management of priapism in the newborn. Urology. 2003;61(1):224.
6. Kuwano AY, Cavalcante A, Costa-Matos A, Spanholi EF, Souza FM. Management in neonatal priapism: case and review. Urol Case Rep. 2017;14:48-9.
7. Merlob P, Livne PM. Incidence, possible causes and followup of idiopathic prolonged penile erection in the newborn. J Urol. 1989;141(6):1410-1.
8. Sood R, Wadhwa SN, Jain V. case report neonatal priapism associated with spontaneous bilateral pyocavernositis. Ann Acad Med Singap. 2006;35(6):425-7.
9. Dust N, Daboval T, Guerra L. Evaluation and management of priapism in a newborn: a case report and review of the literature. Paediatr Child Health. 2011;16(1):6-8.
10. Burgu B, Talas H, Erdeve O, Karagol BS, Fitoz S, Soygur TY. Approach to newborn priapism: a rare entity. J Pediatr Urol. 2007;3(6):509-11.
11. Wang LL, Berrondo C. Idiopathic non-ischemic priapism in an infant: a case report. Urol Case Rep. 2020;33:101428.
12. Walker JR, Casale AJ. Prolonged penile erection in the newborn. Urology. 1997;50(5):796-9.
Humbert JR, Abelson H, Hathaway WE, Battaglia FC. Polycythemia in small for gestational age infants. J Pediatr. 1969;75(5):812-9.

Cite this article as: Emmanuel J, Razali SM, Azmi MA, Veejeyahshegarun B, Arvind M, Yin CL. Neonatal priapism: first reported case in South East Asia and literature review. Int Surg J 2021;8:3154-6.