

Case Series of Distal Epispadias with Intact Prepuce

Sanjay P Dhangar¹, Vaibhav A Thorat², Aman K Jasuja³

How to cite this article:

Sanjay P Dhangar *et al.* Case Series of Distal Epispadias with Intact Prepuce. *Uro, Nephro and Andro Int.* 2023; 8 (2):49-52.

Abstract

Epispadias represents one of the most severe forms of congenital penile anomalies. Classically, the urethra and the glans are opened dorsally with dorsal chordee and shortened penile body. It is usually found as a part of the epispadias-exstrophy complex. Isolated epispadias is rare. It has an incidence of 2.4 per 100,000 births. Distal epispadias and epispadias with intact prepuce are further rare. Only few cases have been reported of epispadias with intact prepuce. We here present our experience of 3 cases of distal epispadias with intact prepuce done by the authors at two different institutes. Post-operatively, all patients did well. After 3 months of follow up, the prepuce was retractable, urine stream was forward and downward and the look of the penis was acceptable cosmetically.

Keywords: Epispadias, Distal epispadias, Epispadias with intact prepuce, Reverse MAGPI.

INTRODUCTION

Congenital abnormalities of the genito-urinary tract involve functional and structural problems. Congenital abnormalities range from renal abnormalities to the urethral abnormalities. These represent 20-50% of all the congenital foetal abnormalities.¹⁻³ The incidence of these genito-

urinary abnormalities range from 0.42 to 4.2 per 1000 births in various studies.^{4,5}

Epispadias represents one of the most severe forms of congenital penile anomalies. Classically, the urethra and the glans are opened dorsally with dorsal chordee and shortened penile body. It is usually found as a part of the epispadias-exstrophy complex. Isolated epispadias is rare.⁶

Author Affiliation: ¹Assistant Professor, ^{2,3}Senior Resident, Department of Urology, Bharati Hospital & Research Center, Pune, Maharashtra, India, ⁴Consultant Urologist, Vishwaraj Hospital, Pune, Maharashtra, India.

Corresponding Author: Sanjay P Dhangar, Assistant Professor, Department of Urology, Bharati Hospital & Research Center, Pune, Maharashtra, India.

E-mail: drspdhangar@gmail.com

Received on: 29.05.2024

Accepted on: 29.06.2024



It has an incidence of 2.4 per 100,000 births.⁷ Just like hypospadias, epispadias can be divided into peno-pubic, shaft, coronal and glanular. It can be further divided into continent and incontinent variety. Distal epispadias (coronal and glanular) and epispadias with intact prepuce are usually seen as the continent varieties. Distal epispadias and epispadias with intact prepuce are further rare. Only few cases have been reported of epispadias with intact prepuce.⁸⁻¹⁰

The diagnosis of distal epispadias with intact prepuce is often missed at birth because the prepuce looks normal. Unless the spade like with broad glans and dorsal direction of the opening of prepuce are noticed and a few specific questions asked about the stream of urine, the diagnosis is not done. On further and specific clinical examination, we can palpate small gap between the two corpora cavernosa.

We here present our experience of 3 cases of distal epispadias with intact prepuce done by the authors at two different institutes.

CASE REPORTS

All the three patients presented late after the age of 5 years (Table). Two of them were brought by their parents for ballooning of the prepuce during micturition and the third one for the abnormal upward direction of the urinary stream. All patients were continent and there were no other urinary complaints. No other congenital anomaly noted clinically. Clinical examination of the penis revealed intact prepuce, prepucial slit oriented dorsally, wide open meatus upto corona, no chordee, bilateral scrotal testis and normal hernial sites. Therefore, clinical diagnosis of coronal epispadias with intact prepuce was made in all the cases. All the urinary, blood and sonological investigation were normal. We did not do any contrast study or urodynamic study as there was no doubt of any other urological abnormality.

After complete workup and fitness, all patients were operated. Epispadias repair was done the same way as we do for coronal hypospadias (Fig. 1-6), except the approximation of the two corpora over the newly formed urethral tube whenever needed. We used 5-0 PDS suture for urethral reconstruction over 10 Fr silicon catheter. Prepucioplasty can be done as a part of the procedure. The catheter was removed on 14th post-operative day. All patients passed urine from the newly formed meatus with anterior and downwardly directed urinary stream. There were no complications in all the patients.

All patients had minimum follow up of 1 year. After 1 year, the look of the penis was acceptable, the prepuce was retractable well with no further urinary or cosmetic complaints.



Fig. 1



Fig. 2



Fig. 3



Fig. 4



Fig. 5



Fig. 6

DISCUSSION

Epispadias is usually found in association with the bladder exstrophy as a part of the exstrophy-epispadias complex. Seventy percent of the

epispadias are complete. Isolated epispadias is very rare. Further rare is the epispadias with intact prepuce. Only few case reports and case series have been reported.¹¹⁻¹³

The embryonic development of prepuce and urethra is well correlated. It means that if there is a defect during development of urethra, the prepuce will also not develop on that side.^{14,15} This explains the typical deformation of the epispadias and hypospadias.

The developmental defect of the exstrophy-epispadias complex is well explained, but there is no well documented explanation for the epispadias with intact prepuce. Few authors suggested that it is because of the non-closure of the urethral plate due to developmental arrest.^{13,16} While others correlate it with megameatus intact prepuce hypospadias variety as many cases of epispadias with intact prepuce were glanular.¹⁷

The real diagnosis of epispadias is usually missed at the time of birth. Under-reporting is expected.¹² This is due to the normal appearing prepuce and hidden urethra inside the intact prepuce. Same happened in our cases. Most of the cases are diagnosed later in life when the child grows and child or the parents notice ballooning of prepuce or abnormal upward urinary stream or the child is planned for circumcision.¹⁸ In our cases also, two children were brought by parents due to abnormal ballooning of the prepuce and the third one brought due to upward urinary stream. No further investigations are required in these patients as the diagnosis is usually clinical except ultrasound of the abdomen and pelvis to look for any other congenital abnormality of the genito-urinary tract. In our cases also, the ultrasound scans were normal in all the patients.

The treatment of this condition needs surgical correction. In cases of distal epispadias specially, glanular and coronal epispadias with intact prepuce, simple approximating urethral reconstruction can be done, with or without prepucioplasty or reverse MAGPI procedure known as IPGAM can also be done. We also did the same in all our cases. Other procedures for more proximal epispadias are Cantwell-Ransley or modified Cantwell-Ransley or complete penile disassembly technique by Mitchell can be done.

Surgical results for repair of distal epispadias give good results as in our cases. Complications are common in more proximal or the classical epispadias repair.

CONCLUSION

The diagnosis of concealed epispadias is prone to be missed and therefore under-reported. Proper diagnosis requires awareness to the clinician as well the parents, high index of clinical suspicion and thorough and complete external genitalia examination.

Financial interests: None

Conflicts of interest: None

Ethical committee approval: Not required

REFERENCES

- Melo BF, Aguiar MB, Bouzada MC, et al. Early risk factors for neonatal mortality in CAKUT: analysis of 524 affected newborns. *Pediatr Nephrol* 2012; 27:965-972.
- Mizuno R. Increase in male fetal deaths in Japan and congenital anomalies of the kidney and urinary tract. *ReprodToxicol* 2010; 30:405-408.
- Queisser-Luft A, Stolz G, Wiesel A, et al. Malformations in newborn: results based on 30,940 infants and fetuses from the Mainz congenital birth defect monitoring system (1990-1998). *Arch GynecolObstet* 2002; 266:163-167.
- You-Lin Tain, Hsing Luh, Ching-Yuang Lin, Chien-Ning Hsu. Incidence and Risks of Congenital Anomalies of Kidney and Urinary Tract in Newborns: A Population-Based Case-Control Study in Taiwan. *Medicine (Baltimore)*. 2016 Feb 8;95(5):e2659.
- Li, Zy., Chen, Ym., Qiu, Lq. et al. Prevalence, types, and malformations in congenital anomalies of the kidney and urinary tract in newborns: a retrospective hospital-based study. *Ital J Pediatr* 45, 50 (2019).
- Dees JE. Congenital epispadias with incontinence. *J Urol* 1949; 62:513e22.
- Epidemiology of Bladder Exstrophy and Epispadias(1987): A Communication from the International Clearinghouse for Birth Defects Monitoring Systems. *Teratology*, 36, 221-227.
- Bhattacharya V, Sinha JK, Tripathi FM. A rare case of epispadias with normal prepuce. *Plast ReconstrSurg* 1982;70: 372-4.
- Sina A, Alizadeh F. Concealed male epispadias, a rare form of penile epispadias presenting as phimosis. *Urol J* 2011;8: 328e9.
- Saurabh Garge. Concealed Epispadias: Report of Two Cases and Review of Literature. *Urology*.2016 Apr;90:164-8.
- McCahill, P.D., Leonard, M.P. and Jeffs, R.D. (1995) Epispadias with Phimosis: An Unusual Variant of the Concealed Penis. *Urology*, 45, 158-160.
- Bos, E.M., Kuijper, C.F., Chrzan, R.J., Dik, P., Klijn, A.J. and de Jong, T.P. (2014) Epispadias in Boys with an Intact Prepuce. *Journal of Pediatric Urology*, 10, 67-73.
- Merlob, P., Mor, N. and Reisner, S.H. (1987) Epispadias with Complete Prepuce and Phimosis in a Neonate. *Clinical Pediatrics (Phila)*, 26, 43-45.
- Cold CJ and Taylor JR The prepuce. *BJU Int*. 1999;83:34-44.
- Stephens FD, Hutson JM. Differences in embryogenesis of epispadias, extrophy-epispadias complex and hypospadias. *J Pediatr Urol*. 2005;1(4): 283-8.
- Ebert, A.K., Reutter, H., Ludwig, M. and Rosch, W.H. (2009) The Exstrophy-Epispadias Complex. *Orphanet Journal of Rare Diseases*, 4, 23.
- Maitama, H.Y., Ahmed, M., Bello, A. and Mbibu, H.N. (2012) Epispadias with Complete Prepuce: A Rare Anomaly. *African Journal of Urology*, 18, 90-92.
- Waziri, A.M., Abubakar, B.M., Adamu, S., Sulaiman, I.E. and Gashua, M. (2016) Epispadias in a Child with Intact Prepuce: A Rare Congenital Abnormality. *Open Journal of Urology*, 6, 19-22.

