**Ped Urol Case Rep 2015; 2(2):11-16** **DOI: 10.14534/PUCR.201529391**

PUCR
Ped Urol Case Rep

**PEDIATRIC UROLOGY CASE REPORTS**

**ISSN: 2148–2969**

**Journal homepage: http://www.pediatricurologycasereports.com**

# Congenital megalourethra in 2 weeks old boy associated with Prune-Belly syndrome

**Lawal Barau Abdullahi, Aminu Mohammad Mohammad, Lofty-John Chukwuemeka Anyanwu, Aliyu Umar Farinyaro**

*Pediatric surgery unit, Department of surgery, Aminu Kano Teaching Hospital, Kano, Nigeria.*

***Abstract*** The megalourethra is a rare congenital anomaly of the penile urethra. It is characterized by the congenital absence of the corpus spongiosum and/or corpus cavernosum. It is especially common associated with Prune-Belly syndrome, and with upper tract abnormalities. We present a 2 weeks old boy with congenital megalourethra because of its association with the Prune-Belly syndrome.

***Key words*** Congenital urethral abnormalities; megalourethra; mesoderm; Prune-Belly syndrome; urethroplasty.



***Corresponding Author***

*Lawal Barau Abdullahi, M.D.*

*Pediatric surgery unit, Department of surgery Aminu Kano Teaching Hospital, Nigeria*

*E mail: a.lawalbarau@yahoo.com*

*The paper was earlier presented at the association of pediatric surgeons of Nigeria annual general meeting and scientific conference, between 25th to 26th September 2014.*

*Accepted for publication: 2 February 2015*

## INTRODUCTION

Megalourethra is a rare congenital anomaly of the anterior urethra and Nesbitt in 1955 first defined it as "a congenital dilatation of penile urethra without distal obstruction" [1,2]. It is characterized by the congenital absence of the corpus spongiosum and/or corpus cavernosum [2,3]. Less than 100 cases have been reported in the English

**Cite this article as:** Abdullahi LB, Mohammad AM, Anyanwu LJC, Farinyaro AU. Congenital megalourethra in a 2 weeks old boy in association with the Prune-Belly syndrome. Ped Urol Case Rep. 2015;2(2):11-16.

literature until now [1,4,5]. In 1962 Dorairajan has been classified congenital megalourethra into two types, scaphoid and fusiform-according to urethrography findings [3]. However, it is now clear that patients can have intermediate forms and some authors consider megalourethra as a spectrum of disease [1,6,7]. Additionally, the specific embryological cause of the congenital megalourethra has yet to be defined. It is especially common in association with the Prune-Belly syndrome, the VACTER syndrome, and with upper tract abnormalities [1,8-10]. We report a case of congenital fusiform megalourethra because of its association with a Prune-Belly syndrome.

## CASE REPORT

A 2 weeks old boy the third child in the family was referred from a peripheral hospital. The patient presented with penile swelling and difficulty in passing urine. He had mild grade intermittent fever. On examination, he was noted to be febrile (37.8 C), his weight was 3.2 kg; he had a hypoplastic anterior abdominal wall with a dilated penile shaft covered by normal skin and bilateral nonpalpable testicles (Fig. 1). An assessment of Prune Belly syndrome in association with congenital megalourethra was made. Abdominal ultrasonography (USG) showed bilateral hydronephrosis.

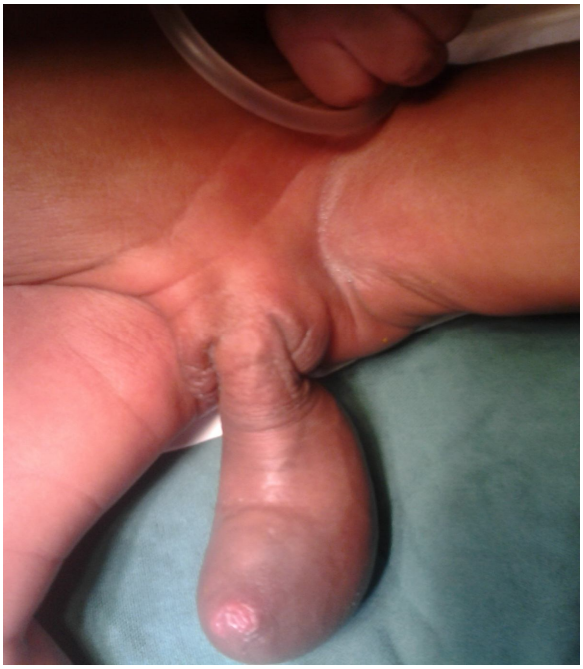

**Fig. 1.** Dilated penile shaft.

Intravenous urogram (IVU) showed bilateral hydrureteronephrosis with dilated calyces. Serum urea and electrolytes was normal. Parenteral antibiotic was commenced and the patient was prepared for surgery. The intraoperative findings were those of dilated penile urethra 4cm in diameter, hypoplastic corporal tissue and bilateral undescended testis. A longitudinal incision was made over the dilated urethra and the excess urethral tissue excised. The urethra was closed over a size 8 urethral catheter as a stent and the skin closed using vicryl 4/0 like the original Nesbitt repair. The catheter was left for 10 days, after which the catheter was removed

and the patient voided well from his neo-urethra. He did well, and was discharged home 15 days after surgery. Follow up after 4 month showed satisfactory wound healing with no meatal stenosis or any difficulty passing urine (Fig. 2).

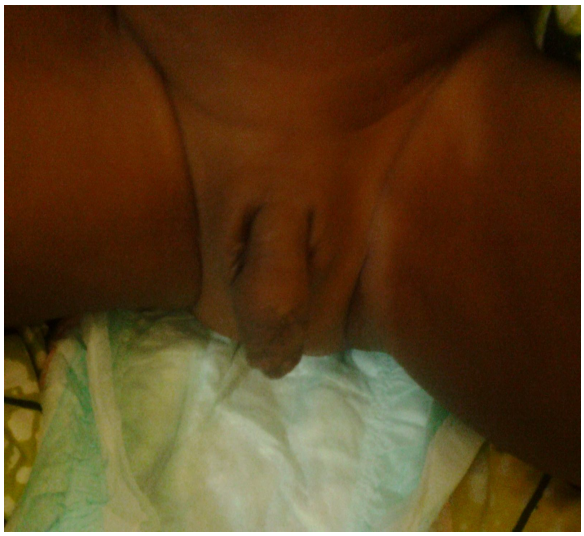

**Fig. 2.** Postoperative appearance of the penis.

## DISCUSSION

The embryological cause of megalourethra is not fully understood. The erectile tissues of the corpora cavernosa and corpus spongiosum arise from differentiation of mesodermal columns in the urethral folds [1]. It is proposed that a defect in the migration, differentiation, or development of the mesenchymal tissues of the phallus results in megalourethra (frequent association with Prune-Belly syndrome) [4,10]. Stephens *et al*. [11] suggested that delayed or deficient canalization of the glandular urethra results in transient urethral obstruction. Additionally, the severity of anomaly is related to the duration of the obstruction, with fusiform megalourethra affected more delayed canalization. There is a stasis of urine caused by functional obstruction due to the poor development of the corpus spongiosum especially in patients with scaphoid type megalourethra [11,12].

Age of diagnosis of megalourethra may change between 16 weeks gestation and 24-years-old [5,10]. Prenatal diagnosis of megalourethra is also possible and approximately 10 congenital megalourethra is diagnosed in neonatal period or early infancy. Sepulveda *et al.* [13] in 2005 described a series of four cases discovered between 20 and 24 weeks. They suggested that congenital megalourethra is associated with poor perinatal outcome. Termination of pregnancy may be advised in cases with other severe congenital anomalies and renal dysfunction [5].

Urethral anomalies such as urethral atresia, a web, duplication, and a diverticulum that affect the anterior urethra are considered in the differential diagnosis of megalourethra [14]. Additionally, some authors reported that megalourethra and diverticula have to be considered as a single malformation with a spectrum of presentations [15]. However, Apel *et al*. [16] suggested that congenital urethral diverticula are different from

megalourethra, because they have narrow orifices and cause obstruction by luminal compression upon filling, whereas the megalourethra never presents a true distal anatomic obstruction.

In approximately 85% of the reported megalourethra cases, coexisting congenital anomalies have been defined. Jones *et al*. [4], in an evaluation study identified associated anomalies in 66 of 78 reported cases of megalourethra. The most commonly associated genitourinary anomalies appeared within the spectrum of prune belly syndrome, although megalourethra has been associated with renal dysplasia/ hypoplasia, hydronephrosis, hydroureter, posterior urethral valves, urethral duplication, vesicoureteral reflux, megacystis, undescended testis, hypospadias and anorectal malformation [7].

The severity of associated anomalies tends to be greater with fusiform type and, associated anomalies were observed in 80% of scaphoid type and 100% of fusiform type [4]. Other system abnormalities associated with megalourethra have been identified as VATER (vertebral, anal atresia, trachea-esophageal fistula, and renal anomalies) and VACTERL (vertebral, anal atresia, cardiac, trachea-esophageal fistula, renal, and limb deformities) [14].

The scaphoid type of megalourethra can be treated by performing a longitudinal reduction urethroplasty described by Nesbitt [2] or a technique of urethral plication described by Heaton *et al.* [17]. The management of fusiform variant is technically challenging for the surgeon because of major phallic reconstruction [4]. However, there have been reports of phallic reconstruction with satisfactory cosmetic and functional outcome [18]. These patients are followed for long-term with regard to the erectile function and fertility potential [14].

As a result, megalourethra is a rare and surgically treatable anomaly of the anterior urethra, as in our case. The prognosis is generally dependent on the type of the anomaly. Early diagnosis and management affords a long lasting cure to the patients. In view of associated anomalies, the workup of megalourethra should include renal function tests and imaging of upper and lower urinary tracts. Then, the status of upper urinary tract will determine the ultimate outcome and the quality of life.

**Acknowledgements**

The author(s) declare that they have no competing interests and financial support.

## REFERENCES


1. Ozokutan BH, Küçükaydin M, Ceylan H, Gözüküçük A, Karaca F. Congenital scaphoid megalourethra: report of two cases. Int J Urol. 2005;12(4):419-21.
2. Nesbitt TE. Congenital megalourethra. J Urol. 1955;73(5):839-42.
3. Dorairajan T. Defects of spongy tissue and congenital diverticula of the penile urethra. Aust N Z J Surg. 1962;32(3):209-14.
4. Jones EA, Freedman AL, Ehrlich RM. Megalourethra and urethral diverticula. Urol Clin North Am. 2002;29(2):341-8.
5. Mohan CG, Prakash SV, Kumar VV, Babu RG. Isolated megalourethra: A rare case report and review of literature. J Indian Assoc Pediatr Surg. 2014;19(3):178-80.
6. Adamson AS, Burge DM. Megalourethra- a spectrum of disease. Pediatr Surg Int. 1990; 25(5): 449–50.
7. Khan M, Qadri SJ, Rather AA, et al. 11 year old boy with megalourethra. JK-Practitioner. 2011;16 (1-2):85-6.
8. Romero Otero J, Gómez Fraile A, Feltes Ochoa J, et al. [Megalourethra in association with VACTER syndrome]. Actas Urol Esp. 2006;30(4):412-4.
9. Ardiet E, Houfflin-Debarge V, Besson R, Subtil D, Puech F. Prenatal diagnosis of congenital megalourethra associated with VACTERL sequence in twin pregnancy: favorable postnatal outcome. Ultrasound Obstet Gynecol. 2003; 21(6):619-20.
10. Sharma AK, Shekhawat NS, Agarwal R, et al. Megalourethra: a report of four cases and review of the literature. Pediatr Surg Int. 1997;12(5-6):458-60.
11. Stephens FD, Fortune DW. Pathogenesis of megalourethra. J Urol. 1993;149(6):1512-6.
12. Wakhlu AK, Wakhlu A, Tandon RK, Kureel SN. Congenital megalourethra. J Pediatr Surg. 1996;31(3):441-3.
13. Sepulveda W, Elorza C, Gutierrez J, Vasquez P, Castro V. Congenital megalourethra: outcome after prenatal diagnosis in a series of 4 cases. J Ultrasound Med. 2005;24(9):1303-8.
14. Wax JR, Pinette MG, Landes A, Cartin A, Blackstone J. Prenatal sonographic diagnosis of congenital megalourethra with in utero spontaneous resolution. J Ultrasound Med. 2009;28(10):1385-8.
15. Savanelli A, Schiano A, Esposito C, Russo S, Dolezalova H. Congenital megalourethra associated with urethral duplication and imperforate anus. Pediatr Surg Int. 1998;13(8):607-9.

16. Appel RA, Kaplan GW, Brock WA, Streit D. Megalourethra. J Urol. 1986;135(4):747-51.
17. Heaton BW, Snow BW, Cartwright PC. Repair of urethral diverticulum by plication. Urology. 1994;44(5):749-52.
18. Locke JR, Noe HN. Megalourethra: surgical technique for correction of an unusual variant. J Urol. 1987;138(1):110-1.

**Access this article online**

**http://pediatricurologycasereports.com**

**Quick Response Code**