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Isolated Primary Megalourethra: A Case Report

Sanjay Kumar Gupta, Shashidharan, Khalid Mahmood, Ahsan Ahmed, Atul Khandelwal, Vijoy Kumar, Mahendra Singh

Department of Urology, Indira Gandhi Institute of Medical Sciences, Patna, Bihar, India

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ABSTRACT

Megalourethra is a non-obstructive dilatation of the penile urethra. It is a rare congenital anomaly characterized by the congenital absence of the corpus spongiosum and/or corpus cavernosum, leading to dilatation of the urethra. Only 80 cases have been reported so far in English literature. Incidence is sporadic with no hereditary or racial predisposition. We report a case of six-year-old child who presented with ballooning of the phallus on micturition and post-void dribbling. The diagnosis of megalourethra was established on the basis of clinical and radiological findings. The patient was successfully managed by reduction urethroplasty. The patient is doing well after 4 years of follow-up.

CASE REPORT

A 6-year-old male child presented with ballooning of the phallus during micturition and post-void dribbling since birth. On physical examination, the phallus was swollen with diffuse sagging of the ventral aspect of the urethra, with dribbling of urine upon applying pressure. The meatus was adequate with normally developed scrotum and testes. An examination of the abdomen, chest, cardiovascular system, and musculoskeletal system was normal. On investigation, hemogram and renal function test was within normal parameters. Ultrasonography of the kidney, ureter, and bladder revealed normal sized kidneys with normal corticomedullary differentiation. The bladder was of normal capacity with no post-void residual urine. Retrograde cystourethrogram (Figure 1) demonstrated scaphoid dilation of the urethra, more marked in the penile region with a normal outline of the bladder. Cystourethroscopy showed marked dilatation of the anterior urethra with a normal posterior urethra and bladder. The patient was managed by 1 stage longitudinal reduction urethroplasty. After circumferential degloving of the penis, the redundant urethra was excised (Figure 2), and neourethra formation was done over a 10 Fr Foley catheter using vicryl 4/0 (Figure 3). The postoperative course was uneventful. The catheter was removed on the fourteenth day, with a normal cosmetic appearance of the penis (Figure 4). In the postoperative period, the patient voided with a good stream and without any cosmetic deformity and post-void dribbling. After 4 years of follow-up, the patient is doing well without any abnormality of the penile shaft.

DISCUSSION

Megalourethra was first coined by Nesbit (1955) who treated the first case of megalourethra at 9 months of age [1]. Embryologically, there is a congenital deficiency of mesodermal tissue of the phallus that leads to the absence or underdevelopment of the corpus spongiosum, and in extreme cases of corpus cavernosum, too. Exact etiology is not known. In megalourethra, urinary tract dilatation develops as a result of urinary stasis in the dilated penile urethra, which lacks adequate support and balloons during fetal micturition, thus causing passive obstruction of the urinary flow. This functional obstruction may also act as a valve-like flap mechanism in some cases, producing intermittent mechanical obstruction to the urine stream through the glandular urethra [2]. Although stricture or severe narrowing of the meatus may cause complete

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CORRESPONDENCE: Sanjay Kumar Gupta,\Department of Urology, Indira Gandhi Institute of Medical Sciences, Patna, Bihar, India (sanjayssgtamkuhi@yahoo.co.in)

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Figure 1. Retrograde cystourethrogram.

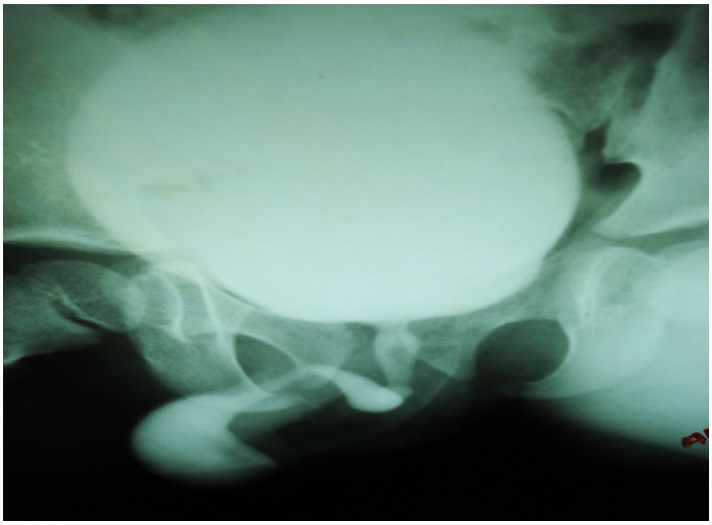


Figure 2. After circumferential degloving of the penis, the redundant urethra was excised.

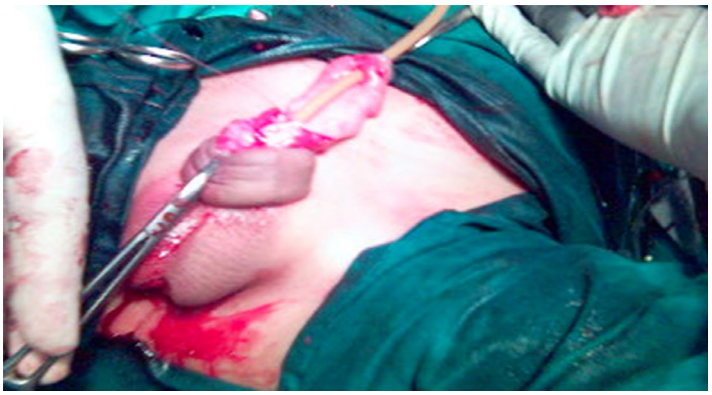


Figure 3. Neourethra formation was done over a 10 Fr Foley catheter using vicryl 4/0.

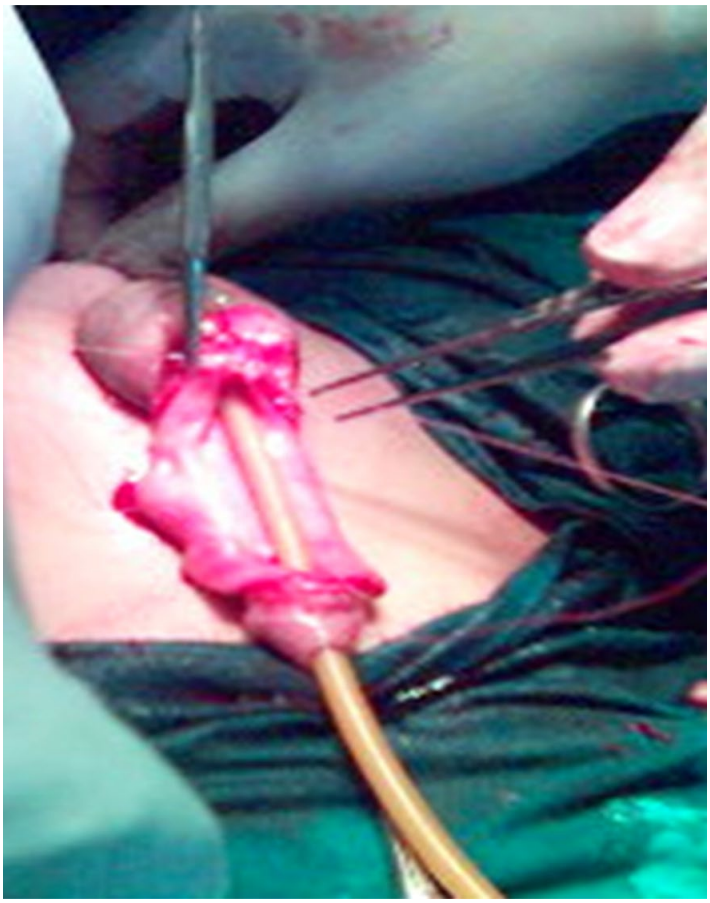
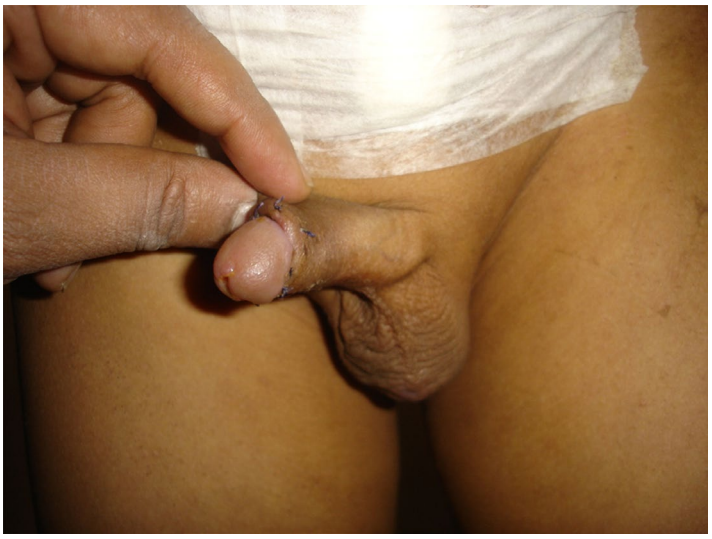


Figure 4. The catheter was removed on the fourteenth day, with a normal cosmetic appearance of the penis.



anatomic urethral obstruction, no true anatomic obstructive defect has been identified in cases of megalourethra [2,3]. Based on urethrography, there are 2 types of megalourethra [4]. First is the scaphoid type in which there is attenuation of the corpus spongiosum and thus bulging of the ventral urethra. The second is the fusiform type. The absence of both the spongiosum and the cavernosum leads to the circumferential expansion of the urethra. Of cases, 50% are associated with Prune Belly Syndrome [5]. There may be association with genitourinary or extra genitourinary anomalies, including vacteral syndrome. Associated anomalies are more common in the fusiform type and the prognosis is worse. Prenatal diagnosis of megalourethra

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relies on sonographic dilatation of cystic dilatation of the penis with or without the presence of obstructive uropathy [6,7]. In neonates with severe fusiform deformity presenting with urosepsis, patients may need a diversion vesicostomy followed by definitive urethroplasty. Diagnosis after birth is mainly clinical. Urethrogram differentiates it from large anterior urethral diverticulum. Some workers diagnosed megalourethra antenatally by ultrasonography (USG).

Ardiet et al. reported a case of complete sequence of vacteral anomalies with megalourethra in one of the twins of antenatal pregnancy [8]. At birth, the penis was enlarged and covered with wrinkled skin. The testes were undescended and the anus was imperforate. Also noted were a single umbilical artery, a patent urachus, and a type-III esophageal atresia. Echocardiography was performed and interventricular communication was discovered. Transfontanellar echography appeared normal but the vertebral echography demonstrated a tethered spinal cord. Sindic et al. reported a complex case of complete urethral duplication with dorsal megalourethra that was severely stenotic in its bulbar part, and the meatus, with the ventral urethra atretic distally, and dilated proximally while missing the corpus spongiosum and cavernosum. Urethral patency was restored successfully by meatoplasty, staged buccal mucosa graft urethroplasty, and tailoring of the megalourethra [9]. Our case was of the scaphoid type with a large, thin, and baggy urethra with attenuated spongiosum. We managed the case by simple reduction urethroplasty. Rarity of the defect precludes any generalization with regard to surgical management. Every case must be considered individually.

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