ISSN: 2573 - 9611

Case Report

Journal of Pediatrics & Neonatal Biology

# Congenital Megalourethra: A Rare Entity

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**Submitted**: 29 Mar 2020; **Accepted**: 04 Apr 2020; **Published**: 09 Apr 2020

## Abstract

*Congenital megalourethra (CMU) is a dilatation of penile urethra without any distal obstruction. It occurs due to one or both corporal defect. About 80-100% cases of CMU are accompanied by multiple systems malformations which make the treatment more challenging. Cysto-urethrogram is necessary for radiological diagnosis and surgical planning. Depending on the type of the CMU and associated congenital anomalies, reduction urethroplasty or major reconstructive surgery is fashioned. We report a case of scaphoid variety of CMU in a 2 year child who was managed by reduction urethroplasty with additional dartos double breasting to prevent postoperative urethrocutaneous fistula (UCF).*

**Keywords:** Congenital Megalourethra, Congenital Anomalies, Reduction Urethroplasty, Scaphoid, Fusiform

## Introduction

CMU results from underdevelopment or deficiency of one or both erectile tissue of the penis. Clinically, it is of two varieties namely; scaphoid and fusiform. Radiological diagnosis by micturating cystourethrography (MCU)/ intravenous urography (IVU) must be performed before any corrective surgery [1]. Associated congenital anomalies along with varying degree of renal and pulmonary dysplasia often affect the surgical outcome. Urethral tapering and tube urethroplasty is the standard operative method for scaphoid variety. While in fusiform variety, urethral reconstruction is much more complex and individualized depending on intraoperative and endoscopic findings, condition of phallus and presence of associated multiple systemic anomalies [2].

## Case history

An 11 month old child presented with tortuous swelling of undersurface of penis and dribbling of urine during micturition since birth. Micturition was never completed unless mother squeezed the penis to empty the urethra. Surprisingly, the child did not have any episode of urinary tract infection. On examination, the child had a tortuous swelling in the ventral surface of penis which was soft and on squeezing of which urine droplets came out from the meatus. The child had no other congenital deformities. He was tried to be evaluated by MCU, but, 6 fr. Infant feeding tube could not be negotiated into the bladder as it coiled inside the dilated part of the urethra. On pushing dye, the dye was pooled up in the dilated urethra. An intravenous urography (IVU) showed dilated and elongated penile urethra. Ultrasonography was normal. With the provisional diagnosis of scaphoid megalourethra (SMU), the child was taken up for surgery.

After degloving of penis by circumcoronal incision, dilated penile urethra was found (Figure 1). We excised the redundant urethral tissue and tubularized the remaining urethral strip over a 6 French nelaton catheter (Figure 2). A second layer of imbricating suture was used to achieve a watertight closure. Per-urethral catheter was kept for 14 days. Patient passed urine normally after catheter removal. There was no evidence of recurrence or fistula in 2 year follow-up.

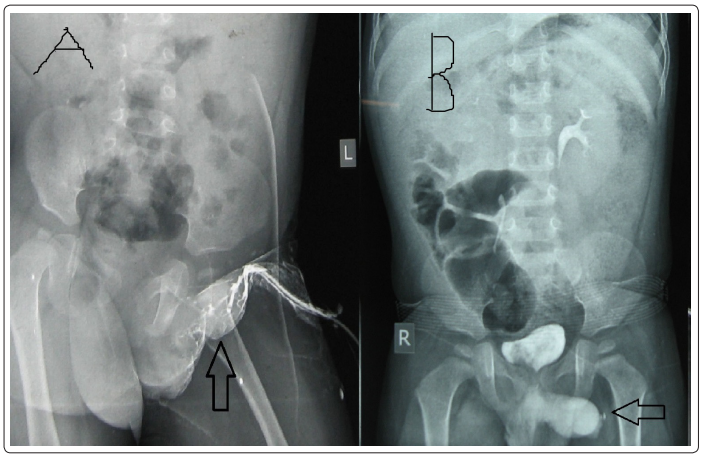


**Figure 1:** Micturating cysto-urethrogram (A) and intravenous urography (B) in an 11 month old child showed coiling of urethral catheter and dilated penile urethra.

## Discussion

In 1989, prenatal diagnosis of CMU was described by Benacerraf [3]. Postnatal CMU was first reported by Obrinsky in 1949 while its surgical treatment was the first described by Nesbitt in 1955 [4]. It is extremely rare in female. Only a few cases of female CMU are reported in literature as an associated anomaly of disorders of sexual differentiation, anorectal malformation and Prune belly syndrome [5]. According to severity, CMU is classified as (a) SMU, a milder form and occurs due to absence or hypoplasia of corpora spongiosum only. 80% cases of SMU have congenital anomalies and they usually have normal amniotic fluid volume with preserved renal and lung function and thus have low mortality rate (13%) [6]. (b) Fusiform megalourethra (FMU), the severe form which occurs due to defect in both corpus cavernosum and spongiosum. Associated anomalies like; renal dysplasia, hydroureteronephrosis, vesicoureteric reflux, prune belly syndrome and urethral duplication are present in 100% cases [7]. Prenatal oligohydramnios, renal dysplasia and pulmonary hypoplasia cause high mortality in this type of CMU. Apart from history and clinical findings, radiological diagnosis by MCU is mandatory. MCU often become difficult as urethral catheter may get coiled into the dilated penile urethra. In such situation, IVU is done which also depicts the megalourethra as well as any upper tract abnormalities. Nesbitt first proposed a longitudinal reduction urethroplasty for SMU. Urethral plication is another viable option which was described by Heaton but both techniques have more or less same rate of postoperative UCF [8]. Now a days, an extra layer of double-breasted dartos tissue after urethroplasty is recommended for prevention of postoperative UCF [9]. Management of FMU is complex, because of lack of supportive corporal tissue. Moreover, associated life threatening anomalies often influence the management as well as the prognosis [10, 11]. Additionally, sex reassignment or placement of penile prosthesis is often required in cases of FMU [12]. Though, successful cosmetic and functional reconstruction of few FMU cases have been reported but, long term follow-up in respect to the urinary continence, erectile function and fertility potential are not established yet.


## Acknowledgement

Dr. Ranjit Kumar Das [MS, M.Ch], Professor & Head, Department of Urosurgery, R G Kar Medical College, Kolkata, India.


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