

**CASE REPORT****CONGENITAL SCAPHOID MEGALOURETHRA: REVISITING A RARE ANOMALY****Ilyas Zahoor, Mohammad Asif, Sarhad Khan, Naveed Haroon**

Department of Urology, Lady Reading Hospital, Peshawar-Pakistan

Megalourethra is an infrequent malformation of the anterior urethra that is caused by the lack of corpus spongiosum and in severe cases is accompanied by the lack of corpora cavernosa as well. We report a five-year-old boy presented to us with scaphoid variety of megalourethra having complaints of ballooning of urethra during voiding. He was investigated with urethrogram and cystoscopy and subsequently repaired with Nesbitt Longitudinal Reduction Urethroplasty. He had smooth recovery postoperatively with normal voiding stream on follow up.

**Keywords:** Megalourethra; Corpora cavernosa; Urethroplasty

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**INTRODUCTION**

Megalourethra is an infrequent malformation of the anterior urethra that is caused by the lack of corpus spongiosum and in severe cases is accompanied by the lack of corpora cavernosa as well. There is no mechanical or functional obstruction of the urethra in congenital megalourethra, but there is still ballooning of the urethra due to the absence of the mentioned structures which differentiates this unique entity from a urethral diverticulum.<sup>1</sup>

The disease is classified in two forms;

1. A milder more common Scaphoid type which lacks the corpora spongiosum
2. A severe less common fusiform type which lacks both the corpora spongiosum and corpora cavernosa.

The Scaphoid type manifests as bulging of the urethra on the ventral aspect while Fusiform type as circumferential expansion of the urethra. This classification is based on the findings of urethrography.<sup>2</sup> The Fusiform type is frequently related to other developmental abnormalities like prune belly syndrome, VATER (vertebral, anal atresia, trachea oesophageal fistula, and renal anomalies) and VACTERL (vertebral, anal atresia, cardiac, trachea-oesophageal fistula, renal, and limb deformities), posterior urethral valves, undescended testis and hypospadias.<sup>3</sup> Due to scarcity of literature on this rare urological anomaly we needed to highlight its important management aspects and in the end bolster our argument with histopathologic evidence.

**CASE PRESENTATION**

A five-year-old male child was referred to our Urology clinic at Lady Reading Hospital with chief complaints of swelling/ballooning of the urethra during micturition and urinary dribbling after passing urine. His antenatal and neonatal history was unremarkable. Upon questioning it was discovered that the complaint has

been existent since birth. On examination the patient was uncircumcised with a normal external urethral meatus with slight anterior urethral ballooning and both the testes were palpable in the scrotum [Figure-1].

Retrograde urethrogram was performed which confirmed the existence of scaphoid variety of megalourethra [Figure-2]. His ultrasound KUB showed normal upper tracts without associated anomalies. Cystourethroscopy was performed with 6Fr ureterorenoscope. An outpouching was visibly confirmed on the ventral aspect of the penile urethra about 3cm proximal to the external urethral meatus, after irrigation was instilled [Figure-3]. The bulbar and posterior urethra were found to be normal and a mildly trabeculated urinary bladder was observed.

Following cystourethroscopy, a Nesbitt longitudinal reduction urethroplasty was performed in the same session [Figure-4]. A sub coronal circumferential incision was made with meticulous degloving performed till the penile base exposing ballooned urethra. The redundant urethral tissue was excised and vicryl 4/0 was used to fashion a neo-urethra over an 8Fr silicon catheter with intervening Dartos and fascial water proofing layers. Excess skin was also trimmed and reconstructed over neo-urethra.

The patient had a normal recovery and the catheter was removed on 14th post-op day. Afterwards, the child passed urine with healthy forward directed stream [Figure-5]. There was no more ballooning of the urethra on micturition and the child did not complain of post micturition dribbling anymore. The patient had his first follow up visit after 3 months and he underwent circumcision 6 months after his initial surgery.

**DISCUSSION**

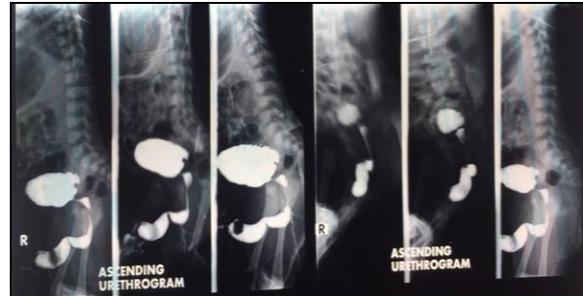
Congenital megalourethra results from the lack of development of mesenchymal tissue of the phallus.<sup>4</sup> In

literature two theories are usually proposed to explain the aetiology and pathogenesis of congenital megalourethra. One suggests a failure of migration of the mesenchymal cells of the primitive streak, during 3<sup>rd</sup> week of embryogenesis, around the cloacal membrane that later on develops into the cloacal folds.<sup>5</sup> The other proposes the delayed or deficient canalization of the glandular urethra<sup>6</sup> fusiform type of megalourethra are complicated by other anomalies<sup>7</sup> However, in our case none of associated anomalies were found hence indicating the rarity of this isolated scaphoid megalourethra. Our patient presented at five years of age far later than other reported cases in literature. It largely goes unnoticed perhaps mainly because of the rarity of the disorder, lack of basic understanding of problem on the part of parents and physicians alike particularly in this part of the world. There are a couple of other anomalies which merit consideration while going ahead with the work up for megalourethra for instance urethral atresia, urethral duplication and urethral diverticulum.<sup>2</sup> The role of voiding cystourethrogram (VCUG) is helpful in establishing the diagnosis of congenital megalourethra especially in cases where it is accompanied by other abnormalities of upper or lower urinary tract.<sup>4</sup> An extensive search on the topic was conducted only to find dearth of reported cases in Pakistan and few from around the world.

Nesbitt in 1955 performed the first reduction urethroplasty for a congenital megalourethra and also coined the term.<sup>8</sup> The aim of the surgery is to excise the redundant part of the urethra and to restore the urethra to its normal caliber. In our case there was an isolated scaphoid congenital megalourethra and we were successful in treating it with a single stage Nesbitt longitudinal reduction urethroplasty with excellent outcome. For a better understanding of the disease, we went ahead to prove histopathological absence of corpus spongiosum as well, thereby corroborating the theory of lack of mesenchymal tissue.



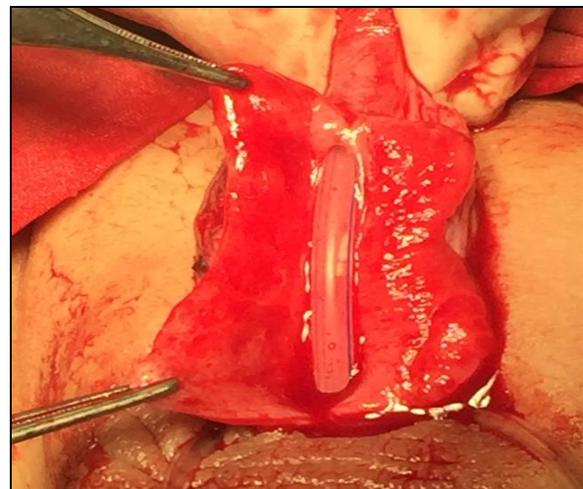
**Figure-1: Post micturition picture of congenital megalourethra showing excessive urethral tissue**



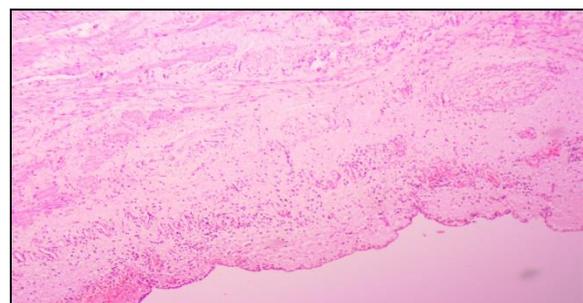
**Figure-2: Boat shaped scaphoid variety on ascending urethrogram**



**Figure-3: Cystoscopic picture showing characteristic trans-illumination of anterior urethra**



**Figure-4: Intra-operative picture after longitudinal division of urethra**



**Figure-5: Histopathology block showing absence of corpus spongiosum around urethra (Blue Arrows mark urethral margin)**



**Figure-6: Postoperative picture showing patient's urinary stream**

## CONCLUSION

Management of megalourethra varies from case to case and treatment is tailored according to the nature and complexity that the case presents. This case study ascertains particularly the non-existence of corpus spongiosum and that single stage longitudinal urethroplasty can successfully treat non-complex and isolated cases of congenital megalourethra.

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### Address for Correspondence:

**Naveed Haroon**, Department of Urology, Lady Reading Hospital, Soekarno Road, Peshawar-Pakistan

**Email:** haroon.naveed@gmail.com