

MEGALOURETHRA

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Congenital megalourethra is defined as dilatation of anterior urethra due to absence of development or deficiency of erectile tissue of the penis. Since its first description by Nesbitt in 1955, 42 cases have been reported(1,2). The effects of megalourethra as such are not life threatening; they may be deformity of the penis (scaphoid megalourethra) or impotence (fusiform megalourethra). The associated anomalies are often life threatening and influence the management and prognosis(2). We present here a report of two cases of megalourethra—one case with serious life threatening urinary abnormalities and the other with associated urogenital, hindgut and extensive musculoskeletal abnormalities of previously unreported nature.

Case Reports

Case 1

A 1^{1/2}-year-old male child was admitted in September 1988 with upward angulation

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of penis and meatal stenosis. There was deficiency of corpus spongiosum in the distal part. Ultrasound scan and CT scan of abdomen showed normal left kidney and atrophic right kidney. Excretory urogram did not demonstrate the kidneys or ureters; but the urinary bladder was visualized, blood urea level was raised (46 mg/dl). Urine specific gravity indicated dilute urine (SG = 1010). Cystoscopy showed bladder trabeculation, type III posterior urethral valves, megalourethra and meatal stenosis. The posterior urethral valves were fulgurated. After 2 months, his blood urea (70 mg/dl) and serum creatinine (1.8 mg/dl) levels were higher than before. In March 1989, his creatinine clearance value was 4.4 ml/mm. Cystoscopy, division of urethral stricture and reduction urethroplasty and megalourethra by opening the skin and urethra in the sagittal plane were done. A urethral fistula that resulted was repaired later.

In October 1990 his blood urea was 100 mg/dl, and serum creatinine was 2.2 mg/dl. Voiding cystourethrogram showed no vesico-ureteric reflux. Isotope renogram showed contracted poorly perfused right kidney, small left kidney with delayed perfusion, delayed peaking and high background activity, suggestive of poor renal function. He was never proved to have urinary sepsis, and is now undergoing treatment with a nephrologist for chronic renal failure.

Case 2

A one day-old-male child of Muslim parents was admitted in January 1992 with multiple congenital anomalies. This was their 6th child. The mother had pre-eclamptic toxemia, but there was no other relevant family history or antenatal history.

It was a full term normal delivery. The mother was 34 years and father was 45 years old and they belonged to Trivandrum city. The child weighed 3 Kg. He had phocomelia of right upper limb, bilateral club feet, congenital dislocation of left upper limb, bilateral club feet, congenital dislocation of left hip, absent anal orifice and a globular swelling in the place of penis (*Fig. 1*) and scrotum. This fluid-filled swelling had an anterior orifice (urethral meatus) through which meconium stained urine was coming out. Testes were impalpable. Right kidney was palpable.

Since the child had anorectal malformation with rectourinary fistula, a high sigmoid colostomy was done as an emergency procedure. Detailed investigations were done after colostomy. Plain X-ray pictures

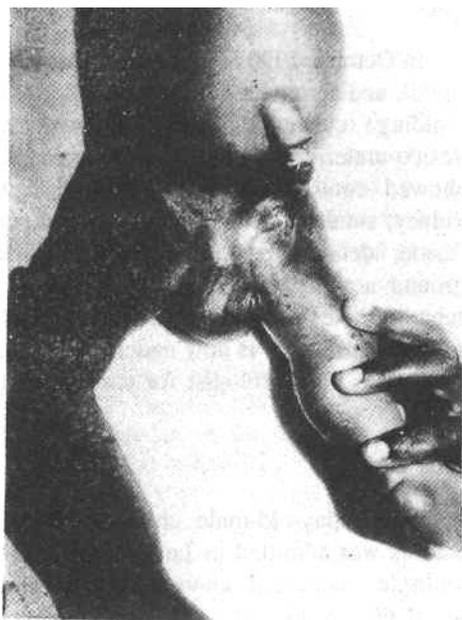


Fig. 1. Appearance of penis of Case 2.

showed hemivertebrae of T6 and T7. Retrograde urethrogram (*fig. 2*) showed hugely dilated anterior urethra and normal posterior urethra. The contrast entered the colon directly from the posterior urethra but the bladder was not visualized. This was obviously a case of fusiform megalourethra with high rectourethral fistula. The child died on the 3rd day of colostomy and hence further investigations could not be done.

Discussion

There are 2 varieties of congenital megalourethra—scaphoid and fusiform. In the scaphoid variety, the corpus spongiosum is deficient or absent, whereas in the fusiform type both the corpus spongiosum and the corpora cavernosa are deficient or absent. Most recently, the two varieties are viewed as a spectrum of presentations of a single entity(1). The fusiform variety is more serious than the scaphoid variety and is associated with more serious anomalies than those associated with scaphoid type(3,4). Hence the former carries a poorer prognosis(1). Out of the previously reported 42 cases of megalourethra, 29 had urinary tract anomalies, 12 had anomalies of other systems and 12 had prune belly syndrome. Both the present cases had urinary tract anomalies; one had anomalies in other systems also. Thus, about 71% had urinary tract anomalies, 30% had anomalies of other systems and 27% had prune belly syndrome. The urinary tract anomalies were hydronephrosis, renal hypoplasia, absent kidney, cystic dysplasia, or pyelonephritis. Ten out of the 42 reported cases were fusiform type and 7 out of those 10 were azotemic or dead; whereas 19 out of the rest 32 reported patients with scaphoid variety were azotemic or dead. The other urinary tract anomalies were megaureter, megacystis, vesicoureteric reflux, bladder

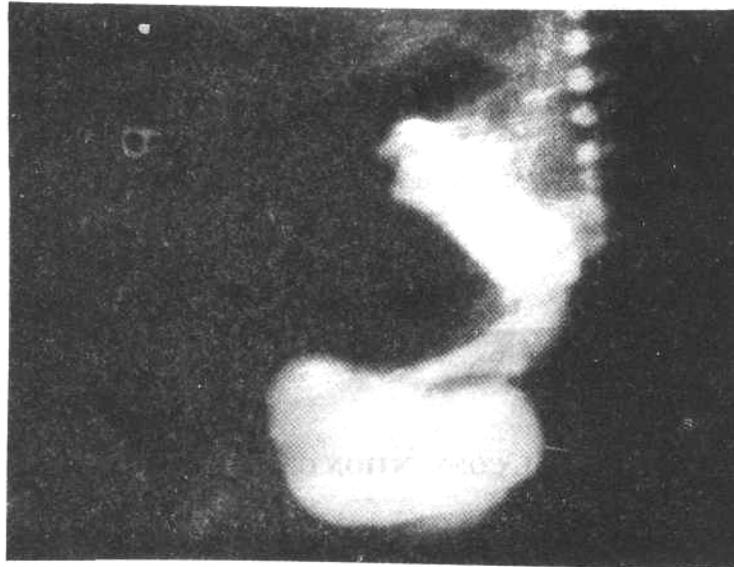


Fig. 2. Retrograde urethrogram of Case 2. The anterior urethra is hugely dilated. The dye is seen entering the rectum.

diverticula, hypospadias, manifestations of intersex, urethral valves, as well as defects in other organs such as teeth, heart and intestines[^]), mongolism⁽⁵⁾ and undescended testes. Six cases of imperforate anus were reported out of which one was with colourethral fistula, one other case with pure rectourethral fistula, one case each of sacral agencies, scoliosis, bilateral club feet; congenital dislocation of hip and cleft palate.

It has been suggested that sac-like dilatations of distal urethra caused proximal obstruction with resultant dilatation of upper urinary tract. This assumption is supported in a case⁽⁶⁾ which the urinary tract returned to normal after correction of megalourethra. Limb anomalies associated with megalourethra have been reported by others^(1,7), But the extensive variety of

limb abnormalities including phocomelia have not been described.

Detailed investigations of the upper and lower urinary tract should be carried out at the earliest to detect anomalies and to treat them early to reduce morbidity and mortality. Instrumentation of urethra is to be avoided⁽¹⁾ as there is a high chance of urinary infection and pyelonephritis, which may prove fatal.

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