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MEGALOURETHRA ASSOCIATED WITH PRUNE-BELLY SYNDROME*

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A 14-day-old male infant with megalourethra is presented because of the rarity of the anomaly and its association with prune-belly syndrome. The lax, wrinkled appearance of the abdomen, bilateral cryptorchidism and severe dilatation of the urinary system are features included in the classic triad of the prune-belly syndrome. Our patient had the scaphoid variety of megalourethra since the penis appeared elongated and floppy in the fusiform form. *Key words: megalourethra, prune-belly syndrome, congenital anomaly.*

Megalourethra is a very rare congenital anomaly characterized by dilatation of the penile urethra. It is secondary to absent corpus spongiosum. In this report, we describe a case of megalourethra associated with prune-belly syndrome[1-5].

Case Report

A 14-day-old male infant was referred to our clinic for evaluation of an abnormally large phallus. A marked ventral bulging of the penis was noted, which increased with voiding. The abdomen was lax and there were several wrinkles on the abdominal wall (Fig. 1). Both kidneys were palpable, the right measuring 10×10 cm in size. The testes were nonpalpable. There was no neurological deficit and examination of the fundus was within normal limits. *E.coli* was isolated from the urine and appropriate antibiotic therapy was initiated. A huge urethra was seen on the urethrogram (Fig. 2). An antegrade cystography was performed by means of a percutaneous tube which demonstrated bilateral vesicoureteral reflux and hydroureteronephrosis with massive ureteral dilatations (Fig. 3). The finding of a lax and wrinkled abdominal wall, bilateral cryptorchidism and dilatation of the urinary collecting system were considered to be the classic triad of the prune belly syndrome. Cutaneous vesicostomy was performed to facilitate emptying of the bladder. Reconstruction of the urethra was scheduled to be performed at the end

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of the first year, but the baby was never brought in for a follow-up, and we learned that he had died six months after discharge.

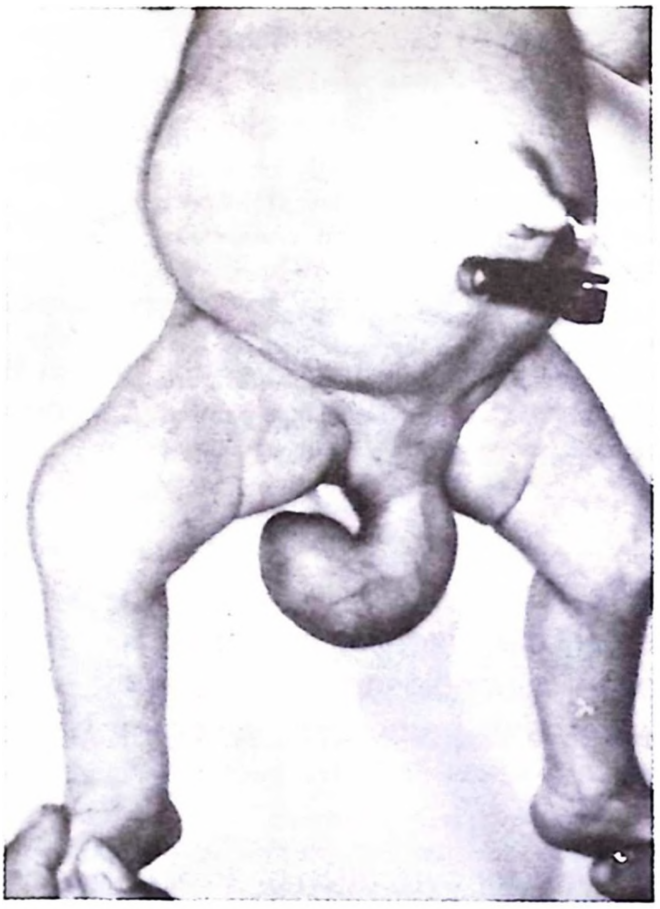

Fig. 1: An abnormally large phallus.

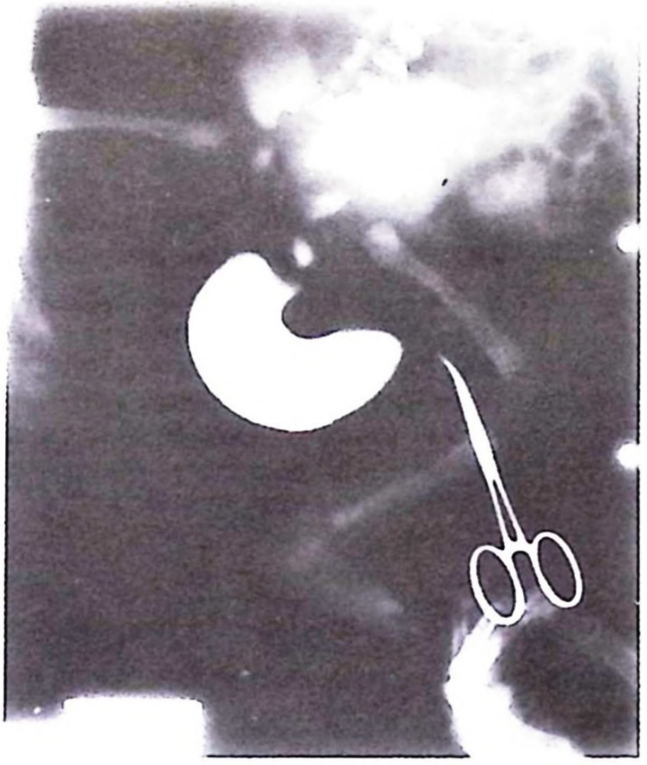

Fig. 2: Retrograde urethrogram.

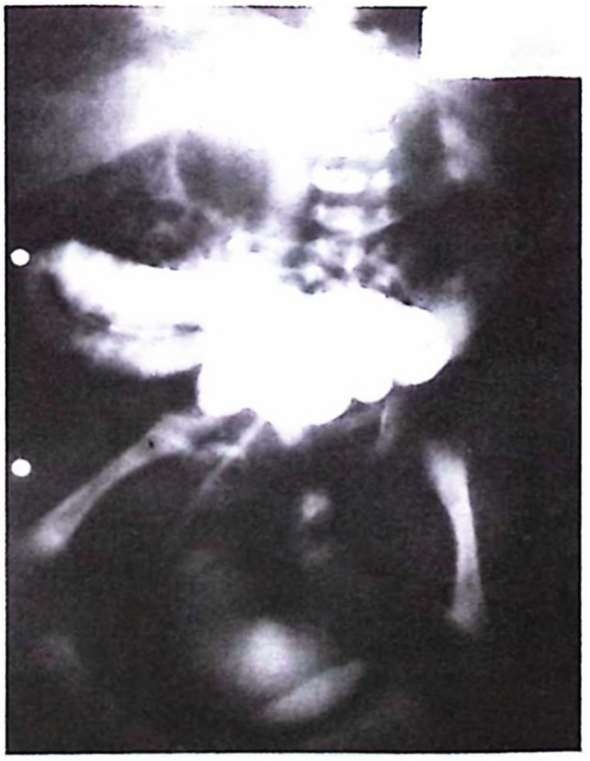

Fig. 3: Antegrade cystography.
Bilateral vesicoureteral reflux, hydroureteronephrosis and antegrade filling of the urethra are seen.

Discussion

Megalourethra is a very rare congenital malformation characterized by dilatation of the penile urethra. Forty-one cases have been reported so far in the literature[1,2,4,5]. The term megalourethra to describe this anomaly was originally suggested by Nesbitt. The etiology of megalourethra is unknown, but it is presumed to result from an arrest in the embryogenesis of the corpus spongiosum at an early stage. In some cases, the corpora cavernosa are affected as well, resulting in their absence or atresia. Therefore, megalourethra has been subdivided into scaphoid and fusiform types. The fusiform variety represents the severest form of this anomaly in which there is a complete absence of the corpora cavernosa and corpus spongiosum. A more commonly encountered form of megalourethra is the scaphoid variety. In this type, there is a loss of the corpus spongiosum over the distal part of the urethra, but the corpora cavernosa are normally developed. It is generally accepted that the scaphoid and fusiform types of megalourethra represent varying degrees of a single disorder rather than two separate entities. Congenital urethral diverticula are not part of this anomaly since they are usually located in the bulbar urethra and cause obstruction. There is no obstruction in megalourethra.

Megalourethra may be associated with prune-belly syndrome since mesenchymal development is defective in both these disorders. The association of megalourethra with upper urinary tract anomalies (such as hydroureteronephrosis, azotemia, cystic displasia) has been reported[3]. This point must be stressed because the status of the upper urinary tract determines the ultimate outcome. This anomaly has also been reported in association with posterior urethral valves, hypospadias and imperforate anus.

Treatment consists of urethroplasty with resection of the excess urethral tissue and reconstruction of the urethra. Prognosis depends mostly on the upper urinary tract status and the severity of the associated anomalies.

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