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LETTER TO EDITOR (VIEWERS CHOICE)

A RARE CASE OF CONGENITAL COMPLETE DIPHALLIA

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A twin male neonate was born at 36-weeks-gestation via vaginal delivery, to a gravida 4 para 1 woman with a spontaneous dichorionic twin pregnancy. The neonate's Apgar scores were 9 and 10 at 1 and 5 minutes, respectively and birthweight was 2245 gm. The 22 weeks prenatal ultrasound (USG) scan showed fetus 1 with a 10 mm anechogenic cyst next to a normal penis and fetus 2 with normal appearing male genitalia. On physical examination, twin 1 had two penises with distinct and independent urethral meatus and no hypospadias (Figure 1), which resulted from an asymmetric sagittal duplication, with the dorsal glans smaller than the ventral glans. Scrotum was normal with bilateral testicles in the scrotum. Anus was normal and there was a small red bump on the sacral region suggesting a hemangioma. Twin 2 had no abnormality on physical examination. Scrotal USG of twin 1 revealed signs of diphallia - both penises with two cavernous bodies adjacent to each ischium, and normopositional testicles in both scrotal sacs separated by the lower penis. Renal USG did not show any abnormality. A voiding cystourethrography identified a single and morphologically normal bladder with two non-communicating urethral paths. The upper urethra arising from the bladder neck (usual topography of the urethra) had a mild proximal stenosis. The lower urethral path corresponding to the ventral penis showed an ectopic location with lateralized implantation in the bladder. Vesicoureteral reflux was not observed. Karyotype was 46XY. Excision of the ventral penis with the ectopic urethra was performed and dilation guided by cystoscopy of the orthotopic urethral stenosis was done, with an excellent aesthetic and functional result. Currently, twin 1 is being followed up in Pediatric Urology clinic and has a slight asymmetry of the scrotum due to penoscrotal transposition.

Diphallia is an extremely rare urogenital malformation with an estimated incidence of 1 in 5 million neonates.[1] The embryological cause of diphallia is not completely known and most explanations are controversial.[2] A more plausible theory is that diphallia results from a defect in the development of the genital tubercle: it arises during separation of the pubic tubercle, in which case each phallus will contain only one corporal cavernous body and one urethra, or during cleavage of the pubic tubercle, in which case each phallus will contain two corporal cavernous bodies and two urethras.[3] Clinical presentation is variable. In cases of complete duplication, other genitourinary malformations such as urinary tract duplication and renal agenesis are usually present.[2,4] Abnormalities of the gastrointestinal system (rectosigmoid duplication, anal atresia) or musculoskeletal malformations such as vertebral deformities may occur.[5,6,7,8] Treatment should be individualized, and it depends on the type of congenital abnormalities detected. The main objective of surgical correction is the reconstruction of the external genitals, ensuring the preservation of continence and erectile function.[2,8] In our patient, due to the ectopic location of the urethra, the excision of the ventral penis was performed, which went without any complications and with an excellent aesthetic and functional result. To date, no other genitourinary or gastrointestinal malformations have been detected.

Figure 1. Two penises with distinct and independent urethral meatus and normal scrotum

The prognosis depends on the number and extent of malformations. All patients should be carefully evaluated due to the high incidence of other associated systemic abnormalities and maintain a long-term follow-up to avoid the appearance of complications.

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