

Extreme penoscrotal transposition

Letter to the Editor

Penoscrotal transposition (PST) is an extremely rare congenital malformation, usually associated with a wide variety of other anomalies. We describe an exceptional case of a newborn with extreme PST and severe hypospadias but normal upper urinary tract and no other associated malformations.

A newborn male delivered at term with no reported antenatal complications. Three routine obstetrical sonography showed normal fetal development. There was no evidence of mother being exposed to teratogens during pregnancy. On physical examination, an abnormal appearance of the external genitalia could be seen. There was a complete rotation of the external genitalia with a normal scrotum and testis but a rudimentary penis with a severe proximal hypospadias and chordee just attached anterior to the anal margin. Urinalysis, renal and vesical sonography and voiding cystourethrography showed no other urological abnormalities. There were no associated cardiological or gastrointestinal anomalies.

Complete PST is a very uncommon heterogeneous condition in which the scrotum is positioned superior and anterior to the penis. It was first recognized in 1923 by Appleby in an adult patient.^[1,2] Less than 20 cases of extreme degree of PST with normal scrotum have been reported in the literature.^[1-7] PST results from abnormal genital tubercle development around the 6th week gestation. It is associated with delay in the midline fusion of the urethral folds. Although the occurrence of most reported cases of PST has been sporadic, other congenital anomalies like hypospadias, chordee and renal agenesis or dysplasia could be found in approximately 90% of patients.^[5] Gastrointestinal abnormalities, predominantly imperforate anus were found in 30% of cases.^[8] PST may present with a broad spectrum of abnormalities ranging from simple shawl scrotum (doughnut scrotum) to very complex extreme transposition with cardiac, gastrointestinal, craniofacial, central nervous system, genital and urological malformations associated.^[6] Growth deficiency and mental retardation have also been noticed in 60% of patients.^[9] Differential diagnosis must include pseudohermaphroditism, penoscrotal hypospadias, micro-penis, intrauterine penile amputation and especially penile agenesis with a midline skin tag anterior to the anus.

Surgery of the more complex cases of PST is technically challenging for pediatric surgeons. It is

usually performed between 12-18 months. The size of the phallus and its potential to develop at puberty into a sexually satisfactory penis are of paramount relevance when surgery is planned. In order to assess this situation, surgeon must carefully palpate the corporal bodies to determine their bulk and sometimes perform a testosterone test to demonstrate erections. Extreme penoscrotal transposition with severe hypospadias and chordee is difficult to differentiate from penile agenesis with a midline skin tag anterior to the anus (with penile atrophic body buried in the perineum). In both cases the penile reconstruction and repositioning are often unsatisfactory, and female gender reassignment, while ethically controversial, may be a prudent therapeutic in selected cases.

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