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CASE REPORT

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Diphallus associated with a third ectopic urethra

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Abstract A case of diphallus is reported in a Saudi male born with two independent phalli with two urethras, the right urethra being joined by a third urethra opening on the perineum. In addition, the child had an absent right 1st rib, a 3rd thoracic hemivertebra, an absent left kidney, and a spinal lipoma.

Key words Penis · Duplication · Diphallus

Introduction

Diphallus is a rare congenital anomaly. In this paper we consider the investigations that should be carried out before surgery is undertaken at the age of six months.

Case report

A 27-day-old infant was referred to the endocrinology clinic with abnormal external genitalia. He was the product of a full-term pregnancy complicated by gestational diabetes, for which the mother was using insulin.

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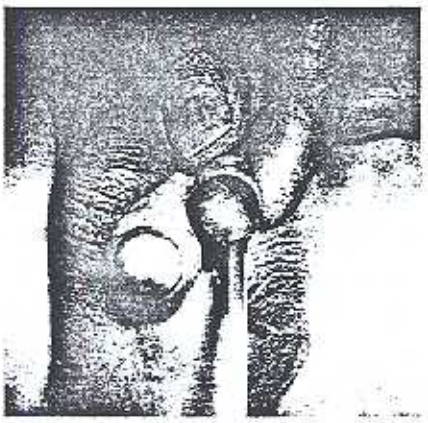

Fig. 1 Diphallus with left glans

The baby was delivered by cesarean section due to cephalopelvic disproportion. The birth weight was 3.2 kg and he had no immediate postnatal problems. Both parents were healthy, the father 33 and the mother 31 years of age, and there was no consanguinity. They had a normal 2-year-old son and the family history was negative for any congenital anomaly. The patient's weight at 27 days was 3.7 kg (25th percentile), his length 49.5 cm (just below the 5th percentile), and his head circumference 36 cm (10th percentile). He showed slightly dysmorphic facial features with a mildly depressed nasal bridge and high-arched palate. His developmental milestones were appropriate for his age.

Examination of the genitalia showed two phallia, the right measured 3.7 cm, with a small meatal opening, while the left was 3.3 cm with a normal meatal opening. Both were erectile by history and clinical examination. The mother stated that the child passed urine normally through the left phallus with a few drops being passed simultaneously from the right side (Fig. 1). The scrotum was bifid with palpable testicles, which were completely descended, on either side. There was no hernia and the anal opening appeared normal. The hips were also clinically normal. There was a soft swelling at the back of the scrotum that extended into the perineum anteriorly and had the consistency of a lipoma. The right lower limb was 0.5 cm shorter than the left, the right foot was markedly smaller than the left and had mild equinovalgus and overlapping toes. The neurological examination was normal.

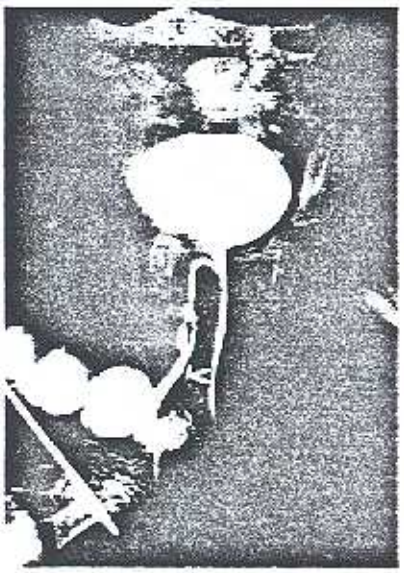

Fig. 2 Cystourethrogram showing third urethra (*arrow*)

Ultrasonography (US) of the abdomen revealed an absent right kidney, which was confirmed by a DMSA scan. Two testicles were seen in the scrotum. For a micturating cystourethrogram, it was only possible to cannulate the left meatus, which showed a normal urethra ending in the bladder. The right bifurcated distally into two urethras, one ending subcutaneously (Fig. 2). A skeletal survey showed an upper dorsal hemivertebra and absent 1st rib. A computed tomography

Table 1 Classification of diphallus

1. True diphallia
   a) Complete: two independent penises, each with two corpora cavernosa and a corpus spongiosum
   b) Partial: rudimentary penis represented by a glans or small erectile tissue.
2. Bifid phallus
   a) Partial: bifid glans
   b) Complete: bifid penis with one corpus cavernosum in each penile branch
3. Pseudodiphallia
   Rudimentary atrophic penis, existing independently of the normal penis

scan showed a neural arch defect. Chromosomal analysis revealed a normal male karyotype (46 XY); serum electrolytes were normal. Cystoscopy through the left urethra revealed a normal urethra; it was not possible to pass the cystoscope beyond the urethral bifurcation on the right. Excision of the right accessory phallus was carried out through an elliptical incision, the dissection being carried down to the base of both corpora. The third urethra was mobilized from the subcutaneous tissue and excised with the accessory phallus.

Discussion

Diphallus is one of the rarest congenital anomalies, the incidence being 1 in 5–6 million births [11]. It was first reported in a cadaver by Wecker in 1609; since then approximately 100 cases have been reported [1–11]. The first case in an Arab child was reported in Egypt in 1968 (complete diphallus) [3]; a further three cases were reported from this institution in 1985 [6]. The site of the accessory phallus can be anywhere: the penile shaft, perineum, groin, or inguinal canal. Embryologically, it is believed that this anomaly is due to failure of fusion of the paired genital tubercles (cloacal membranes).

Diphallus has been classified by Aleem [2] as shown in Table 1. Associated anomalies have also been described. In 90% of cases there are urological anomalies, e. g., double bladder, double or triple urethra, absence of both urethras, bladder exstrophy, vesicoureteric reflux, hypospadias, epispadias (common), undescended testicles, horseshow or single kidney, and absent prostatic tissue. Gastrointestinal anomalies, e. g., complete duplication of the hind-gut and anorectal malformations have also been described, as have musculoskeletal anomalies such as spina bifida, meningocele, hemivertebrae, short sacrum, thoracic scoliosis, omphalocele with wide separation of the symphysis pubis, and segmented spleen.

This is the eight case report in an Arab child [6]. In this case, however, there were different associated anomalies, including three urethras, an absent right 1st rib, and vertebral anomalies in addition to an intraspinal lipoma. A similar case in a 15-year-old patient was recently reported [7], although in this case the third urethra originated from the bladder neck.

Despite the social implications of this condition, surgery for the anomalies should be delayed until 6 months of age so that an accurate assessment can be made and a proper cystoscopic examination done, identifying the verumontanum with its ejaculatory duct. US evaluation of the diphallus has recently been recommended [9], and in our patient this showed two corpora cavernosa and one corpus spongiosum in each penis. This provides an additional tool for ensuring a normally functioning penis prior to excision of the other phallus.

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