CASE REPORT

A rare case of isolated complete diphallia and review of the literature

Vijay Kumar Kundal, Mufique Gajdhar, Arvind K Shukla, Raksha Kundal

Department of Pediatric Surgery, SMS Medical College, Jaipur, Rajasthan, India

Correspondence to

Dr Mufique Gajdhar, gajdhar@hotmail.com, gajdhar@gmail.com

SUMMARY

A 3-year-old boy presented with complaints of abnormal genitalia. On examination, there was penile duplication with two completely separate phalluses, unequal in size, one with hypospadias and the other with meatus at the tip of the glans. Voiding cystourethrography revealed a normal sized single bladder but two urethras with no associated vesicoureteral reflux.

BACKGROUND

This is a rare case of isolated complete penile duplication with no other associated systemic anomaly.

CASE PRESENTATION

A 3-year-old boy was admitted with complaints of abnormal genitalia. The child was born by vaginal delivery and his antenatal history was uneventful. There was no history of oral contraceptives intake by the mother, no radiation exposure or infection during gestation period, and no significant family history was reported. On examination, there were two completely separate phalluses, unequal in size, one with hypospadias and the other with normal meatus (figure 1).

There was a history of dribbling of urine from the hypospadias smaller sized phallus and continuous flow from the other phallus while micturating. Each phallus was found to have an apparently normal corpus cavernosum. The scrotum was well developed with palpable normal sized testis separated by soft tissue mass. The anal opening was at its normal site, and the anal sphincter had a normal tone.

INVESTIGATIONS

A karyotyping study showed 46XY. x-Rays of the spine and pelvis were normal. Abdominal ultrasonography and CT were normal. Intravenous urography (IVP) showed bilateral normal functioning of the kidneys. Baseline blood investigations were within the normal range. Voiding cystourethrography revealed no vesicoureteral reflux, a normal single bladder and two urethras (figure 2).

TREATMENT

Surgical procedure included the following steps: penile dissection up to the ischiopubic region; dissection of the hypoplastic urethra; amputation of the smaller phallus; urethral anastomosis and phalloplasty. The postoperative period was uneventful.

DISCUSSION

Diphallus or duplication of the penis is one of the very rare congenital anomalies of the urogenital system. It has an incidence of 1 in 5 million live-births.¹ Wecker² first reported diphallia in the literature in 1609. About 100 cases have been reported worldwide till now.³ The extent of duplication and the number of associated anomalies vary greatly from glans duplication arising from a common shaft to complete duplication of the penis accompanied by multiple anomalies including ventral hernia, bladder exstrophy, vesicointestinal fissure, bladder duplication, ureteral and renal defects, imperforate anus, ectopic scrotum, prepenile scrotum and meningomyelocele.^{4–8} According to Schneider,⁹ diphallia has been divided into three categories: (1) glans duplication, (2) bifid diphallia and (3) complete diphallia or complete penis duplication. Later, Vilanova and Raventos¹⁰ added a fourth category, namely pseudodiphallia.

All cases of diphallia need to be evaluated carefully and completely in view of the high incidence of systemic anomalies. Prognosis is largely dependent on whether it is a case of penile duplication with multiple associated anomalies or one of isolated penile duplication. The duplication of the phallus may be orthotopic or ectopic, symmetric or asymmetric in shape and size. Currently, widely accepted classification includes two main types: true diphallia and bifid phallus.¹¹ These two types are further divided into partial or complete. True complete diphallia or complete penile duplication is described as a case where each phallus has two corpora cavernosa and a corpus spongiosum. When one of the penises is smaller or rudimentary, it is called true partial diphallia. The term bifid phallus

Figure 1 Isolated complete diphallus with soft tissue mass in the scrotum.

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Figure 2 Voiding cystourethrograph showing two urethras with a single bladder and no vesicoureteral reflux.

is used when only one corpus cavernosum is present in each penis. When the degree of separation is complete up to the base of the shaft, it is called complete bifid phallus. Penile duplication is usually associated with other systemic anomalies, such as bladder and urethral duplication, cloacal exstrophy, imperforate anus, gastrointestinal duplications and vertebral anomalies, and it may also present as an isolated anomaly.^{8 12 13}

Detailed studies of embryological anatomy of the genital tubercle have thrown light on the understanding of diphallus. The development of the normal phallus begins with the joining of bilateral cloacal tubercles at the anterior end of pars phallic of the urogenital sinus.¹⁴ Mesodermal columns grow rapidly around the lateral margin of the cloacal plate from the genital tubercle.

Various possible embryological explanations for diphallia are as follows:

1. Historically, Cecil¹⁵ had suggested that penile and bladder duplication are considered the end result of incompletely fused bilateral anlagen or a form of atavism, because snakes and lizards possess a double penis.
2. Ravitch¹⁶ reported that diphallia could represent a teratoid structure or a form of incomplete twinning.
3. De vries and Friedland¹⁷ described that inhibited growth of the dorsal inferior wall of the urogenital sinus underlay accessory urethral development.
4. Das and Brosman¹⁸ hypothesised that abnormal Mullerian duct termination was responsible for urethral duplication.
5. Hollowell *et al*¹⁹ described that diphallia results from the failure of fusion of mesodermal bands.
6. Woodhouse and Williams²⁰ attributed it to an ischaemic process in embryogenesis.
7. Troyer²¹ proposed that maldevelopment of the distal portion of the urethra is because of the uncoordinated growth of the urogenital sinus and the urethral plate.

There are multiple embryological explanations for diphallia and systemic anomalies, but most researchers agree that it is a defect connecting the genital tubercle. It is hypothesised that diphallia during the gestation period occurs between 23 and 25 days, when the fetal caudal mass of the mesoderm is deeply disturbed by trauma, drugs or infections.

Management of diphallia poses a challenge in view of the medical, surgical and ethical decision-making. Patients with diphallia should undergo a thorough evaluation to rule out associated anomalies. Furthermore, a detailed anatomy of the

external genitalia also helps to classify not only the degree of penile duplication but also in the planning of surgical procedure. MRI is currently the non-invasive imaging for interpretation of the anatomy of complex genital anomalies.²² Treatment of isolated diphallus includes excision of the duplicated penile structure and its urethra.^{23 24}

Learning points

- Complete penile duplication is a rare congenital genitourinary condition.
- Patients of penile duplication should be evaluated in detail to rule out any associated systemic anomaly.
- Treatment includes excision of the duplicated penile structure and its urethra.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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