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Case Report

A Two-Year-Old Boy with Diphallia and Multiple Congenital Anomalies

Laki-Laki 2 Tahun dengan Diphalia dan Kelainan Kongenital Multipel

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ABSTRACT

Diphallia is a rare urogenital defect and varies from a small accessory penis or duplication of the glans to complete penile duplication. Its incidence is 1 in 5.5 million live births. Only 100 cases were reported from 1609 until 2021. A two-year-old boy with diphallia and multiple congenital anomalies had an uncircumcised penis with two glans penis and two complete urethral orifices, accompanied by scrotum bifidum, asymmetric right buttock, with normal anorectal position. Spina bifida with lipomyelocele, tethered cord at the level of vertebra lumbar 3-4, scrotum bifidum, and left mild hydronephrosis were observed using Magnetic Resonance Imaging (MRI). Urethrocystoscopy of both complete urethra revealed meatal stenosis and bulbar urethral stricture, normal posterior urethral, and no verumontanum in the right urethral orifice and did dilatation. Whereas normal urethral orifice, anterior urethra, posterior urethra and verumontanum were found in the left side. Management for diphallia must consider associated anomalies and the goal of patient satisfaction. In this case, conservative treatment is the best choice.

Keywords: Diphallia, multiple anomalies

ABSTRAK

Diphallia adalah kelainan urogenital yang langka dan bervariasi yaitu dari penis yang kecil, duplikasi *glans penis*, hingga duplikasi penis lengkap. Insiden kejadiannya adalah 1 dari 5,5 juta kelahiran hidup. Dari tahun 1609 hingga 2021, hanya ada 100 kasus yang dilaporkan. Seorang anak laki-laki berusia dua tahun memiliki diphallia dan beberapa kelainan kongenital multipel. Anak memiliki dua glans penis yang belum disunat dan dua *meatus urethra eksternus*, disertai dengan skrotum bifidum, pantat kanan asimetris, dengan posisi anorektal normal. Pada pemeriksaan MRI ditemukan spina bifida dengan *lipomyelocele, tethered cord* setinggi vertebra lumbal 3-4, skrotum bifidum, dan hidronefrosis ringan kiri. *Urethrocystoscopy* pada kedua uretra ditemukan *meatal stenosis* dan striktur uretra pars bulbar, uretra posterior normal, dan tidak ada verumontanum pada uretra kanan. Pada sisi kiri ditemukan *meatus urethra eksternus*, uretra anterior, uretra posterior, dan verumontanum normal. Tatalaksana diphallia harus mempertimbangkan anomali yang saling berhubungan dan keinginan pasien. Tatalaksana konservatif adalah pilihan terbaik dalam kasus ini.

Kata Kunci: Anomali ganda, diphallia

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INTRODUCTION

Diphallia is a rare disorder in penile duplication. This anomaly varies from the addition to accessory penis, duplication of glans penis, or even duplication of complete penis. Global awareness regarding congenital anomaly has arisen as 260.000 deaths worldwide (about 7% of all neonates deaths) were related to congenital anomaly. Diphallia is considered extremely rare that it occurs in 1 neonate per 5.5 million live births and only 100 cases reported through 1609 until 2021 (1-3). The duplicated penis may be located in sagittal or frontal plane and symmetric or asymetric in shape or size (2).

Whereas in Indonesia congenital anomaly accounts for 5.7% of infant mortalities, in which this number increased to 19% in 2010. This also contributes to 4.9% deaths of children under-five. Most congenital anomaly cases (approximately 90%) arose from the developing countries with limited health facilities and resources (3). Nearly 10% of the cases were associated with various urogenital defects. However, factors and etiologies regarding these anomalies are yet to be understood (4).

Diphallia itself remains a challenge to be reconstructed because of number of varieties and every case is unique. This anomaly frequently presents with other urogenital anomalies, namely hypospadias, epispadias, or rudimentary penis. However, a type where diphallia presents with two completely developed penises, structurewise and functionwise, is extremely rare (5). Appropriate and thorough diagnostic modalities should be ordered to decide the proper treatment as it will affect the patient's quality of life. In this case report, we present a case of diphallia with two complete penises with other numerous congenital anomalies admitted to Saiful Anwar General Hospital Malang, which underwent conservative treatment.

CASE REPORT

A two-year-old boy came to the hospital and complained to have two glans penises (Figure 1). During neonatal state, he was able to urinate through both external meatuses. Two months later, he could only urinate from the left external meatus. There was no abnormality in voiding. According to the antenatal care history, the patient was delivered through cesarean section at the gestational age of 36 weeks. During pregnancy, his mother denied any history of consuming hormonal drugs, alcohol, or smoking. After birth, the patient immediately cried and only required routine care. Afterwards, the patient was admitted to the hospital due to diarrhea and undescended testicles, in which bilateral orchidopexy has been performed.

Physical and general examination showed that the patient was active and had good nutrition status based on his body mass index (BMI). Based on the urogenital status, two glans penises with two urethral orifices were identified with uncircumcised penis, scrotum bifidum (Figure 2), asymmetric right buttock (Figure 3), and normal anorectal position were observed. Karyotyping revealed 46XY. Urine was passed through both urethras. Further evaluations, including magnetic resonance imaging (MRI), urethrocystoscopy, and panendoscopy, were then planned



Figure 1. Urogenital examination



Figure 2. Scrotum bifidum revealed 2 glans penis and with 2 external urethral meatus



Figure 3. Asymmetrical right buttock and anal position

MRI examination showed spina bifida, lipomyelocele (Figure 5a), tethered cord at the level of vertebra lumbar 3-4, scrotum bifidum, and left mild hydronephrosis (Figure 4) and double corpus cavernosus (Figure 5b). Urethrocystoscopy on both penises revelaed meatal stenosis and bulbar urethral stricture (Figure 6), normal posterior urethra, and no verumontanum and did ureteral dilatation for bulbar ureteral stricture in the right urethral orifice (Figure 8), and normal urethral orifice, anterior urethra, posterior urethra (Figure 7), and verumontanum in the left counterpart. Two months after the surgery, the patient was only able to urinate from the left penis but still was not able to urinate from the right penis.



Figure 4. MRI examination shows (1) tethered cord L3-L4, (2) left anal sphincter deviation, and (3) left mild hydronephrosis



Figure 5. a.) MRI examination shows spina bifida with lipomyelocele. b.) MRI examination shows double corpus cavernosum



Figure 6. Right Urethrography examination. Urethrography showed bulbar urethral stricture



Figure 7. Left Urethrography examination: Urethrography showed normal anterior & posterior urethra



Figure 8. Urethrocystoscopy examination: In Right Penile, it was found Bulbar urethral stricture followed by the normal posterior urethra and no verumontanum

DISCUSSION

True diphallia with completely duplicated penises is a very rare anomaly and every case is unique as it is not identical to each other and has each unique anatomical variety. Often accompanying associated anomalies contribute to wider variations of clinical presentation. Diphallia is divided to two groups, which are true diphallia and bifid phallus. Each classification is then further classified to complete or partial duplication. In true complete diphallia, two completely duplicated penises are observed, each penis has one corpus spongiosum and two corpora cavernosa. In contrast, in bifid phallus, only one corpus cavernosa is present. Whereas in true partial diphallia, whether it is a true diphallia or bifid phallus, the duplicated penis has smaller or rudimentary structure (5).

Several congenital anomalies have been associated with true diphallia, including ectopic scrotum, bifid scrotum, colon duplication, double bladder, hypospadias, imperforate anus, vertebral defects, and bladder exstrophy (4). It is also reported to have association with numerous neurological defects such as lipomeningocele, tethered cord, low conus, dural ectasia, and spina bifida. In this case of true diphallia, several other defects, including spina bifida, lipomyelocele, tethered cord L3-L4, and scrotum bifidum, were found. The exact etiology and pathogenesis of the occurrence are still not understood clearly (4). However, some theories have been proposed. First, the defect is believed to be the result of malformation of embryonic development during 3th to 6th week of pregnancy (6). Second, abnormalities of paired mesodermal analgen migration during the 15th week of pregnancy was thought to play role in the defect.

Penis development commences with the concretion of

bilateral cloacal tubercles at the anterior end of pars phalica located in the urogenital sinus. Column of mesoderm grows rapidly at the lateral margins of the cloacal plate, forming the genital tubercle. Collateral urethral duplication is also thought to result from longitudinal duplication of the cloacal membrane, allowing three or four columns of primitive streak mesoderm to migrate ventrally around the two cloacal membranes and form two genital tubercles (5,6).

Failure of genital tubercles fusion during the 23rd to 25th day of gestation is the main etiopathogenesis of diphallia. Karyotype is normal in most of the cases, where karyotyping shows karyotype of 46XY. However, exception of karyotype abnormality, which was a balanced translocation (1,4) (p36.3; q24.3), was reported by Karna et al Multiple imaging modalities have been studied to evaluate diphallia, such as ultrasonography and MRI (7). Utilizing the aforementioned imaging modalities might precisely identify the presence of corpora cavernosa, hence the distinction of complete bifid phallus and true complete duplication. MRI is advantageous in evaluating diphallia as it has greater tissue resolution and may evaluate structures through different planes. MRI has high sensitivity and specificity in differentiating types of diphallia. It also provides information regarding urethral course, its length, as well as its relationship to surrounding corporal bodies. Furthermore, generally, MRI is very effective in evaluating detailed pelvic anatomy structures. Pre-operative planning may also be aided with MRI (7,8). Abdominal ultrasonography is utilized to identify congenital malformations located in the abdomen. Another modality, urethrocystoscopy, is useful in locating functional urethra as well as differentiating it from congenital urethroperineal fistula. Moreover urethrocystoscopy also helps evaluate the bladder (9,10).

Complete diphallia poses a complex treatment problem in many aspects. Basically, surgical repair is the main procedure to correct the anatomical problems so that proper urinary continence and erection can be achieved. The procedure also aims to restore esthetic aspect of the penis and proper urinary stream. Other accompanying anomalies are also able to be repaired through surgical procedure. Penile urethroplasty with augmentation of the ventral penis with excision of the shorter duplicated penis might be the treatment of choice for selected cases (4,11,12). However, surgical approach may differ from one another, depending on the anomalies occurring in each

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patient. Thus, the principle of reconstructing diphallia lies in the anatomical repair of the penis while maintaining the continence, erectile function, and goal of patient satisfaction (4). In this case, conservative management was done, in which observation of dilatation of urethral stricture without excision of the other glans was performed. As a result, the patient showed viable glans, meatus, and tissues, hence an excellent outcome of urinary function. Conservative treatment with panendoscopy and routine follow-up was considered the best treatment in this case as duplicated penile excision may pose a risk of permanent stricture to the glans and meatus.

Diphallia is a rare and unique urogenital defect, in which no cases are believed to be identical. The variety ranges from a small accessory penis or duplication of the glans to complete penile duplication. Diphallia is divided into true diphallia and bifid phallus. Imaging modalities, such as ultrasonography, MRI, and urethrocystoscopy, are necessary and beneficial in diagnosing and evaluating congenital defects. In this case, conservative management was chosen by observing urethral dilatation with routine follow up as both of the glans, meatus, and tissues are still viable. The patient had excellent urinary function outcome. Therefore, conservative management is the preferred treatment of choice. Follow-up is still mandatory as erectile and excretory function is still necessary to be evaluated.

DATA AVAILABILITY

All data underlying the results are available as part of the article, and no additional source data are required.

COMPETING INTERESTS

No competing interests were disclosed.

CONSENT

Written informed consent for publication of their details and clinical images was obtained from the patient.

ETHICS

Ethical approval for this case report was obtained from The Ethics Committee of Saiful Anwar General Hospital, Malang, with approval number 400/04/K.3/CR/302/2020.

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