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Aphalia Penis Reconstruction with Tubing Radial Forearm Free Flap:

A Case Report

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***Abstract*—Aphallia Penis or penile agenesis is a condition where no penis is found in a man. This is caused by the failure of the development of the genital tubercle. This disorder is very rare, with an estimated incidence of 1 in 10 million to 30 million births. Treatment for aphallia must begin as early as possible, based on careful evaluation and rapid sex determination by the clinical team, including plastic surgeons, psychologists, endocrinologists, and urologists. Case: A man, 22 years-old-patient, from Bali came to the Reconstructive and Aesthetic Plastic Surgery Polyclinic Dr. Soetomo, Surabaya with the main complaint of not having male genitals from birth. The patient has been urinating through a hole under the clear yellow scrotum. Three operations have been carried out to reconstruct the penis. iscussion: In this case, the patient had reconstruction for aphalia penis. Three operations were performed. The first is a uretroplasty surgery in which the operation of the removal of the external urethra meatus from the perineal to the penoscrotal area The second stage of surgery was performed neorethra exploration in the antebrachial region, external genital reconstruction with radial forearm free flap sinister tubes, silicone implant insertions, Neophalus insertions, and split thickness skin grafts to close the defects in the antebrachia. In third stage surgery, uretroplasty surgery is performed again to move the external urethra meatus in the penoscrotal distal to the penis. Conclusions: Treatment for aphallia must begin as early as possible, based on careful evaluation and rapid sex determination by the clinical team, including plastic surgeons, psychologists, endocrinologists and urologists. After sex determination and consultation with a psychologist, after adulthood can be carried out a total phallic reconstruction using radial artery-based free forearm flaps or with pedicled anterolateral thigh flaps, where this method is known as a simple, fast, and well-reported method of phallus reconstruction.**

Keywords—aphalia Penis, tubing radial forearm free flap, phallus reconstructio

I. INTRODUCTION

Aphallia Penis or what is called penile agenesis is a condition in which the penis is not found in a man. This is due to the failure of the development of the genital tubercle. This disorder is extremely rare, with an estimated incidence of 1 in 10 million to 30 million births [1].

Some studies claim the absence of corpora cavernosa and corpus spongiosum as a prerequisite for the diagnosis of penile Aphallia. Patients usually have a 46XY karyotype. There are two main groups of penile Aphallia patients, those with congenital anomalies incompatible with life and those with penile Aphallia as a separate malformation [2].

Skoog and Belman mentioned three variants, based on the position of the urethra in relation to the anal sphincter, such as: Postsphincteric; Presphincteric (Prostato-rectal fistula) and Urethral atresia. The more proximal to the bladder tract, the more likely it is to develop an anomaly [3].

This rare occurrence should be differentiated in neonates with hidden penis, simple penis, micropenis, pseudo hermaphroditism, intersex, and intrauterine penile amputation. In the case of androgen insensitivity syndrome (AIS), which is a genetic condition in which androgen receptors are dysfunctional or ineffective and this causes partial or complete external feminization in infants whose XY chromosome and phallus may appear defective or absent. In some cases of adrenogenital hyperplasia in women with relatively prominent scrotal folds, the diagnosis can only be made by hormonal tests and chromosome studies. For older children who present with anomalous differentiation from acquired cases of penile ablation should be performed, in which the penis is traumatically or iatrogenically amputated after a problematic circumcision, in cases such as previous scarring or an incomplete phallus can be seen [3].

Treatment for aphallia should be started as early as possible, based on careful evaluation and rapid sex determination by the clinical team, including plastic surgeons, psychologists, endocrinologists, and urologists. For babies with aphallia, the recommended surgical treatment for patients with aphallia is the early determination of whether the sex is male or female. After sex determination and consultation with a psychologist, total phallic reconstruction can be performed

after adulthood using a radial artery-based free forearm flap or a pedicled anterolateral thigh flap, which is known as a simple, rapid, and well-reported method of phallus reconstruction [4].

II. Literature Review

A. Embriology

In a two-week-old fetus, there are two layers, namely ectodermal and endodermal. Then an indentation is formed in the middle, namely mesodermal which then migrates to the periphery, separating ectodermal and endodermal. The caudal side of the cloaca differentiates into the urogenital folds (precursors of male and female external genitalia). Between week four and week seven, mesodermal cells immigrate to the cranial side of the cloaca to form the genital tubercle. The cells from the endodermal originating from the cloaca will migrate along the ventral midline to become the urethral folds, at the same time the mesenchyme will proliferate to form the urogenital folds. At week seven, the genital tubercle will elongate and form the glans. It is the primordial form of the penis in males and clitoris in females [5].

In male fetuses with Y chromosomes, the SRY gene stimulates primitive sex cords to differentiate into Sertoli cells which will form fetal testes and Leydig cells. Leydig cells produce testosterone which will then be converted to dihydrotestosterone by 5α-reductase type 2. Dihydrotestosterone binds to androgen receptors which stimulate the lengthening and enlargement of the genital tubercle. Urogenital folds will migrate to the middle following the urethral groove, and genital swelling will become the scrotum. This process moves from proximal to distal [5].

In the third month of the fetus, the prepuce develops from the tissue at the base of the glans penis, which later grows to cover the dorsal part of the penis and surrounds the glans, and fuses at the centre to form the frenulum. Penile differentiation is complete by 12 weeks of gestation. During the second and third trimesters, penile growth proceeds with the help of fetal androgen hormones, which are produced due to stimulation from the fetal pituitary. There was a significant increase in penis size of nearly 20mm from 16 weeks of gestation to 38 weeks. Therefore, the true state of micropenis occurs due to hormonal disturbances that occur after 12 weeks of gestation [5]. There are several factors that affect the differentiation of male external genitalia, including: testosterone synthesis by fetal testes, conversion of testosterone to dehydro-testosterone by the 5α-reductase enzyme, and the presence of androgen receptors on target cells. In men who experience one of these disorders, the genital tubercle will become the clitoris, the genital folds will become the labia minora, and the genital swellings will become the labia majora [5,6].

B. Patophysiology

Fetal production of testosterone which is converted to dihydrotestosterone (DHT) is essential for the development of normal male genitalia. In early gestation, placental human chorionic gonadotropin (HCG) stimulates testicular development to produce testosterone by binding to luteinizing hormone (LH) receptors. Approaching 14 weeks' gestation, the fetal hypothalamic-pituitary-gonadal axis is active, and testosterone production decreases under the influence of luteinizing fetal hormone. Therefore, penile growth after the early trimester is dependent on fetal testosterone production. Shortly after birth, there is an increase in luteinizing hormone and testosterone that lasts for 12 hours, after which gonadotropin (LH-FSH) and testosterone production decrease. At the beginning of 1 week of age, levels of gonadotropins and testosterone begin to rise again until puberty levels, peaking at 1-3 months of age, then decreasing until prepuberty at 6 months of age. After 6 months of age, the next penis growth occurs slowly until it rises again at puberty until adulthood [5].

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C. Diagnosis

The diagnosis of penile aphalia begins with the absence of the corpora cavernosa and the copora spongiosum with the urethral opening at any point on the perineum in the midline, above the pubis, on the anterior aspect of the scrotum, or most commonly just the anterior anus and the anterior wall of the rectum. This rare occurrence should be differentiated in neonates with hidden penis, simple penis, micropenis, pseudo hermaphroditism, intersex, and intrauterine penile amputation. In the case of androgen insensitivity syndrome (AIS), which is a genetic condition in which androgen receptors are dysfunctional or ineffective and this causes partial or complete external feminization in infants whose XY chromosome and phallus may appear defective or absent. In some cases of adrenogenital hyperplasia in women with relatively prominent scrotal folds, the diagnosis can only be made by hormonal tests and chromosome studies [3].

For adolescents presenting with penile aphallia, a history should be taken, in which case a history of traumatic or iatrogenic amputation of the penis can be obtained after a problematic circumcision. In that case, a scar was found [3].

D. Management

Neonates with these lesions should be evaluated promptly by a karyotype and other appropriate examinations to determine if there are other malformations associated with the urinary tract or other organ systems. There is a clear link between aphallia and renal malformations [3,7].

Treatment for aphallia should be started as early as possible, based on careful evaluation and rapid sex determination by the clinical team, including plastic surgeons, psychologists, endocrinologists, and urologists. For babies with aphallia, the recommended surgical treatment for patients with aphallia is the early determination of whether the sex is male or female [4].

After sex determination and consultation with a psychologist, total phallic reconstruction can be performed after adulthood using a radial artery-based free forearm flap or a pedicled anterolateral thigh flap, which is known as a simple, rapid, and well-reported method of phallus reconstruction [4].

Experimental data and several clinical observations have established the role of androgens in the formation of male gender identity. Postnatal and early postnatal androgen exposure greatly contributes to male psychosexual development and identity, which cannot be changed by neonatal castration, hormonal treatment, phenotypic adjustment, or psychosexual manipulation. Therefore, male sex should be maintained in patients with penile agenesis [8].

In the literature there are almost no data on total phallus construction in children with congenital malformations. In contrast, phalloplasty for traumatic penile amputation has been reported in children and adolescents using different techniques. The most commonly used procedure for phalloplasty in postpubertal patients is a microvascular radial forearm flap. This complex procedure requires a skilled team of doctors, including plastic surgeons, psychologists, endocrinologists, and urologists and is currently only performed in a handful of specialized centres that can guarantee high standard results [7].

The main disadvantage of the radial forearm flap is a large, unpleasant donor scar. While we believe that this method should be considered in patients born with aphallia [7].

III. Case Report

A 22-year-old male patient from Bali came to the Polyclinic of Reconstructive and Aesthetic Plastic Surgery at Dr. Soetomo, Surabaya with the main complaint of not having male genitalia since birth. The patient has been urinating through a hole under the scrotum, which is clear yellow in colour. Defecate every day and normal.

On physical examination, there was no penis, but a scrotum was found. The external urethral meatus was found in the perineal area (figure 1).

Fig. 1. External urethral meatus.

Performed three times operation. The first operation was a urethroplasty operation in which the external urethral meatus was transferred from the perineal to the penoscrotal area (figure 2).

Fig. 2. Uretroplasty operation.

While the second stage operation was performed 4 months after the first operation, neourethra explorations with a silicone catheter of size 10 in the antebrachii region (figure 3), reconstruction of the external genitalia with radial tubing forearm free flap left (figure 4), inserting silicone implants, inserting Neophalus, and split thickness skin graft to close the defect. on the antebrachii (figure 5). In stage three surgery, a uretroplasty was performed again to move the external urethral meatus at the penoscrotal distal to the penis. Penoscrotal fistula repair was also performed with donor scrotal skin.

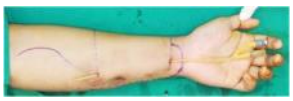

Fig. 3. Antebrachii region.

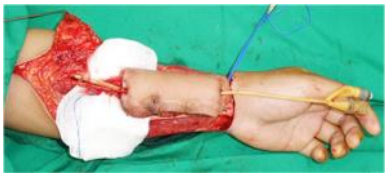

Fig. 4. Reconstruction of the external genitalia with radial tubing forearm free flap left.

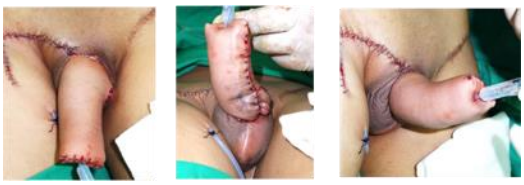

Fig. 5. Neophalus, and split thickness skin graft to close the defect on the antebrachii.

IV. Conclusion

Aphallia Penis or what is called penile agenesis is a condition in which the penis is not found in a man. This is due to the failure of the development of the genital tubercle. This disorder is extremely rare, with an estimated incidence of 1 in 10 million to 30 million births [1].

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After sex determination and consultation with a psychologist, total phallic reconstruction can be performed after adulthood using a radial artery-based free forearm flap or a pedicled anterolateral thigh flap, which is known as a simple, rapid, and well-reported method of phallus reconstruction [4].

Management of the patients in this case report was to carry out 3 stages of surgery. The first is uretroplasty in which the operation involves transfer of the external urethral meatus from the perineal to the penoscrotal area. While the second stage of surgery was performed neourethra exploration with a silicone catheter of size 10 in the antebrachii region, reconstruction of the external genitalia with a radial tubing forearm free flap left, inserting silicone implant, inserting Neophalus, and split thickness skin graft to close the defect in the antebrachii. In stage three surgery a uretroplasty was performed again to move the external urethral meatus at the penoscrotal distal to the penis. Also performed penoscrotal fistula repair with donor scrotal skin.

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