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Case report

A very rare case of diphallia with anorectal malformation

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ABSTRACT

Introduction and importance: Diphallia is extremely rare urological anomaly with reported incidence of 1 in 5–6 million live births. It can present as complete or incomplete diphallia. In most of the cases it is associated with complex urological, gastrointestinal or anorectal malformations.

Case presentation: We report here case of a newborn who was presented to us at 1st day of life with diphallia and anorectal malformation. He had true diphallia with two separate uretheral orifices. Both phalluses were uncircumcised, phallus 1 was 2.5 cm in length while phallus 2 was 1.5 cm. Both phalluses had normal shaped glans with uretheral opening located at the normal place He had a single scrotum with two midline raphe and well-formed rouge. He was passing urine from both orifices. His ultrasonography of urological system showed two ureters and a single hemi bladder. He was admitted and operated upon and a sigmoid divided colostomy was constructed. Per-operatively congenital pouch colon (type 4) was identified. His post-operative recovery was uneventful. The patient was discharged on second post-operative day and called for follow up.

Clinical discussion: Diphallia is a rare congenital anomaly, which means two structurally and anatomically separate phalluses. Complete Duplication in Diphallia presents the kind of Diphallia in which both the phalluses have two corpus cavernosum and only one corpus spongiosum. As diphallia presents with a spectrum of diseases; therefore, it requires a multidisciplinary approach. A case of Diphallia may well present with complex urogenital, gastrointestinal or anorectal malformations. As in our case the patient had Diphallia with anorectal malformation. Hence he was operated upon and a sigmoid colostomy was constructed.

Conclusion: Diphallia is a very rare congenital anomaly which can occur in association with anorectal malformations. Management of such cases should be individualized depending upon the spectrum of disease.

1. Introduction

The duplication of urogenital system is extremely rare with the incidence of diphallia being 1 in 5–6 million live births [1]. The first case was reported in 1609 by Wecker in Bologna, Italy [2]. Until now only 100 cases have been reported in the literature [3]. In Pakistan the incidence is not known. The etiology of diphallia is complex and is due to alteration in genes sequencing during the embryonic development. However the extent of duplication varies, and is due to failure of mesodermal banding but is not well understood [4]. True diphallia refers to complete penile duplication, each with two corpora cavernosa and a corpus spongiosum, whereas bifid phallus is characterized by only one corpus cavernosum present in each penis [5]. We herein report case of a newborn with true Diphallia and anorectal malformation without fistula, who was admitted and managed at our facility. We report a case according to the Updating Consensus Surgical CAse REport (SCARE) 2020 guidelines [6].

2. Presentation of case

We report here case of a newborn who was presented to emergency on 1st day of life with complaints of inability to pass stool since birth and passing urine from two separate phalluses (diphallia). The child was born at 36 weeks of gestation via spontaneous vaginal delivery and was a product of consanguineous marriage. The parents had no family history of congenital anomalies. Upon arrival in the emergency department he was normothermic on room temperature with good hydration. On local examination no anal opening was identified (anorectal malformation) with two well-formed phalluses, both phalluses were uncircumcised, phallus 1 was 2.5 cm in length while phallus 2 was 1.5 cm.

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Both phalluses had normal shaped glans with uretheral opening located at the normal place. A single scrotum with two midline raphe and well-formed rouge (Fig. 1). Both lateral pouches had testis and the central pouch was empty. Patient was passing urine from both the uretheral openings. His x-ray abdomen/pelvis showed a gas shadow above the level of coccyx (anorectal malformation without fistula) with wide pubic diatheses. His ultrasonography of urological system showed two ureters and a single hemi bladder. He was admitted to the neonatal unit. He was operated upon and a sigmoid divided colostomy was constructed. Per-operatively he had dilatation of the rectum (Fig. 2) and a congenital pouch colon was identified (Fig. 3). His post-operative recovery was uneventful. The patient was discharged on second post-operative day and called for follow up.

3. Discussion

Diphallia is a rare congenital anomaly, which means two structurally and anatomically separate phalluses, discovered in 1609 by a Swiss doctor Johannes Jacob Wecker who found this condition while working on a cadaver [7]. Diphallia is most often associated with other congenital anomalies like hypospadias, ectopic scrotum, duplication of bladder, imperforate anus, duplication of colon, exstrophy of cloaca or bladder, ventral hernia and vertebral malformations [8]. Diphallia was initially categorized as True Diphallia, Bifid Phallus and Diphallia of glans by Schneider and the a 4th type Pseudo-Diphallia by Villanova and Raventos. True Diphallia is then further classified as Complete and Partial Duplication which are as a result of cleavage of the pubic tubercle and separation of pubic tubercle respectively [9].

Complete Duplication in Diphallia presents the kind of Diphallia in which both the phalluses have two corpus cavernosum and only one corpus spongiosum; whereas in Partial Duplication one of the phalluses has a rudimentary structure [9]. In Bifid Phallus there is only a single corpus cavernosum in each penis [10].

The French Association of urology (AFU) has their own classification of Diphallia based on histology and morphology of the associated organs into three types namely, Complete and Incomplete True Diphallia, Complete and Partial Bidfid and Pseudophillia as shown in Table 1 [11].

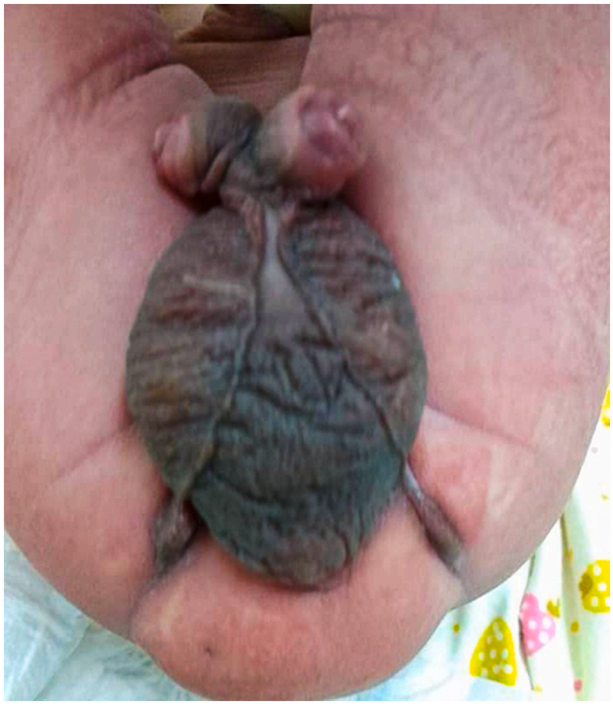

Fig. 1. Diphallia with two midline raphe and well-formed rouge.

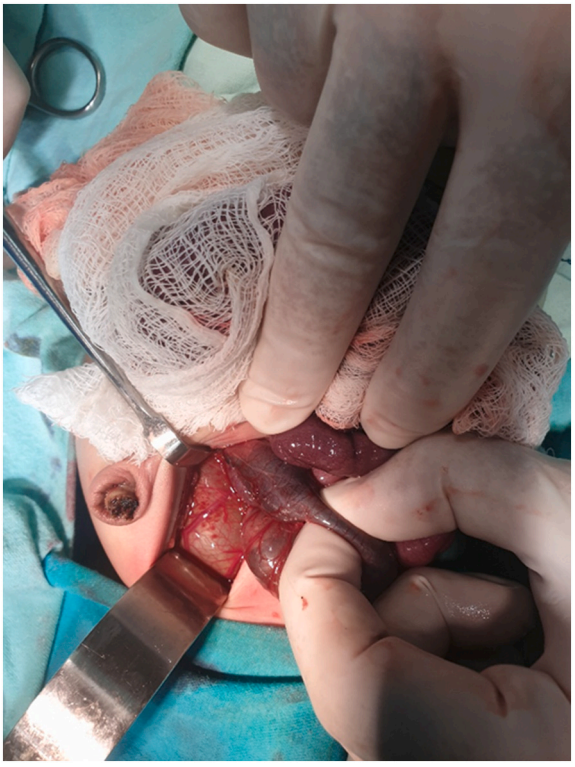

Fig. 2. Sigmoid colon ending into hugely dilated rectum.

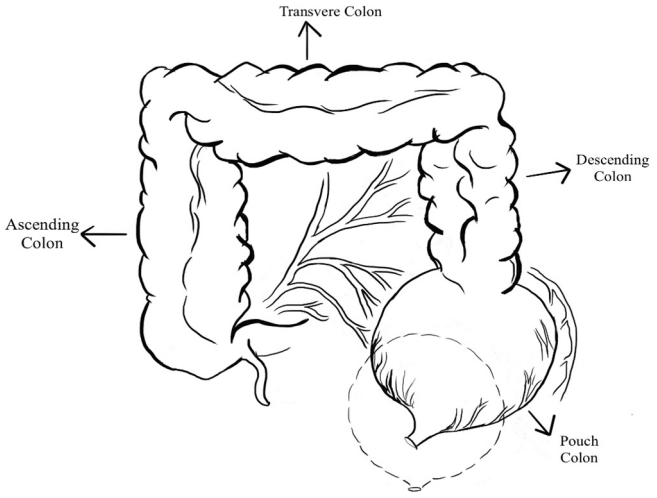

Fig. 3. Congenital pouch colon (Type 4).

Table 1
Different types of Diphallia.

Schneider classification	Vilanova classification	French association of urology (AFU)
Complete Diphallia	Complete Diphallia	Complete & Incomplete True Diphallia
Bifid Diphallia Diphallia of glans	Bifid Diphallia Diphallia of Glans Pseudo-diphallia	Complete & Partial Bifid Penis Pseudo-diphallia

Diphallia is thought to occur in a fetus at approximately 4th week of gestation. The exact cause of Diphallia remains uncertain; however, there are numerous theories that postulate the true cause of Diphallia.

One of the theory suggests that it happens because of partial coalescence of Anlagen because of incomplete development of urogenital sinus' dorsal inferior wall and causes accessory urethral growth [12]. Another theory states that urethral duplication occurs because of inability of fusion of mesodermal bands [13]. Mullerian Duct Termination is also considered to be one of the causes of Diphallia by one theory. One of the theories attributes Ischemia during the development of fetus to the development of Diphallia [14]. The anomalies of other associated organs occur when the genital tubercle has a defect in its connection. Depending on the type of Diphallia, urination can occur from both or one penis and scrotum can either be completely developed or bifid.

Neonatess born with Diphallia have varied degrees of erectile function with either completely intact erectile function in both penises or one of them or even no erection in either one. Patients with Diphallia are at a greater risk of death than normal infants owing to their anomalous renal, colorectal and genital systems [15].

As diphallia presents with a spectrum of diseases; therefore, it requires a multidisciplinary approach. A case of Diphallia may well present with complex urogenital, gastrointestinal or anorectal malformations. As in our case the patient had Diphallia with anorectal malformation. Hence he was operated upon and a sigmoid colostomy was constructed. Ultrasonography is the modality of choice but MRI has far more precision. Since we do not have the facility of MRI at our setup an ultrasonography of urological system was performed which revealed two separate ureters and a single hemibladder [16].

Surgery is the mainstay of treatment; for both the congenital anomalies. For anorectal malformations a sigmoid divided colostomy is performed in the neonatal age. Then distal contrast study is performed highlighting the type of anorectal malformation. After four to eight weeks a definitive procedure (anorectoplasty) is performed via posterior sagittal or abdomino-perineal approach. The Pena's dilation protocol is followed until the adequate size is achieved. Which is followed by Colostomy reversal in four to eight weeks of time [19].

The higher complexity and mortality associated with Diphallia is because of the accompanying anomalies of urogenital and gastrointestinal system [17]. The goal of the surgery is to restore the normal functional anatomy, a cystouretherogram is performed outlining the type of anomaly, whether it is complete or incomplete uretheral duplication along with bladder duplication. Hence the primary step is preserving penis with the orthotopic urethra to perform penectomy, resection of the replicated, less functional penis [18]. Along with Penile resection, staged reconstruction is performed including creating neourethera, which is followed by scrotoplasty at a later stage, in order to yield the best possible cosmetic and functional outcomes and to preserve the maximum possible erectile, ejaculatory, urinary functions. Bhat et al. reports a similar where successful correction of true diphallia in a 1-year-old child, and later on at 5-years his scrotoplasty was performed [20]. Rawat et al. presents a case similar to ours of diphallia with anorectal malformation in a 2-days-old neonate where early reconstruction was performed. A divided sigmoid colostomy was made in neonatal life followed by both penile reconstruction, and anorectoplasty simultaneously at 6 months of age and the patient was followed up with good bowel and bladder control at 10 months [9].

4. Conclusion

Diphallia is a very rare congenital anomaly which can occur in association with anorectal malformations. Management of such cases should be individualized depending upon the spectrum of disease.

Ethical approval

This study does not require institutional ethical review board approval in accordance to institutional policies.

Patient consent

Written informed consent was obtained from the patient's parents/legal guardian for

publication of this case report and accompanying images. A copy of the written consent

is available for review by the Editor-in-Chief of this journal on request.

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Acquisition of data: Murad Habib, Hajira Fazeelat Bajwa.
Interpretation of data: Murad Habib, Muhammad abbas.
Intellectual content: Muhammad Abbas, Hajira Fazeelat Bajwa.
Supervision: Muhammad Amjad Chaudhary.

Declaration of competing interest

There are no conflicts of interest.

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