Rare Phallus Malformations in Children

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Abstract

Introduction: Penile agenesis (PA) and diphallus (DP) are extremely rare genital anomaly in children. Numerous associated anomalies have been described with these malformations. These patients need several investigations and finally surgical intervention.

Material & Methods: In a retrospective study, 14 patients who were treated for phallus malformation in Mofid Children's Hospital from January 2004 to December 2013 were studied. Detailed history was taken and para clinical examinations were performed in each patient and diagnosis was confirmed by laboratory tests, abdominal ultrasonography, voiding cystourethrography (VCUG), and karyotype study. Data regarding age, sex, clinical findings, associated anomalies and treatment were collected.

Results: From 14 patients, eight cases had PA (aged 2 to 4320days), and six had DP (aged 2 to 5040 days).Karyotype in all PA's patients was 46XY. Five of DP cases had completed, and one had bifid phallus type. All patients were treated surgically after complete investigations, and followed within period of study.

Conclusion: All patients with phallus anomalies need extensive evaluations and surgical intervention. No surgical intervention should take place without counseling the parents.

keywords

- penile agenesis
- diphallus
- treatment
- outcome

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Introduction

Agenesis of the penis is an extremely rare genitourinary anomaly, with the incidence rate of one in 30 million live-births.[1, 2] Penile agenesis (PA) results from either absence of the genital tubercle or its failure to develop.[3] Patients with PA usually have 46XY karyotype.[4, 5] These patients have two major groups of: 1- with congenital anomalies incompatible with life and 2- with solitary malformation.[6, 7] More than half of these patients have associated anomalies.[8] Those with solitary malformation have absent penis and usually normal appearing scrotum which contains palpable testicles with normal function.[3, 7, 9, 10] Skoog and Bellman [8] introduced a classification on the basis of the relationship between the urethral meatus and the anal sphincter:1-Post sphincteric with anterior perianal urethra, 2- pre-sphincteric with urethra-rectal fistula, and 3-urethral atresia with Vesico-rectal fistula. Opening of the urethra is either over the pubis bone or on the perineum (mostly, in anterior wall of the rectum). [8, 11] Treatment for these patients is controversial (Surgical intervention during infancy).[8] Duplication of the penis or diphallus is very rare and occurs once in every 5 to 6 million live births. [12] Approximately 100 cases have been reported after the first case published by Wecker in 1609.[13] Extent of the duplication and associated anomalies are very diverse.[14] Embryologically a diphallus deformity can occur in two ways; 1: "separation" of the pubic tubercle, in which each phallus has the same set of corporal body and urethra, or 2: "cleavage" of the pubic tubercle in which each phallus has a unique set of corporal cavernous bodies and urethras. [15] Diphallus has been classified in different ways.[6] Most cases of Diphalia have the same corpus cavernosum in each organ, and duplicated urethras usually may be associated with diphallus.[17-19] We herein discuss our six diphallic patients and their associated anomalies. We also evaluated the meatus type, scrotum and testis position. Finally all cases underwent surgery to reconstruct diphallus and its associated anomalies.

Material and Method

In a retrospective study, 14 patients who have been treated for phallus malformation were evaluated in Mofid Children's Hospital from January 2004 to December 2013. Detailed history was taken and para clinical examinations were performed in each patient and diagnosis was confirmed by laboratory tests, abdominal ultrasonography, IVP, voiding-cysto-urethrography (VCUG), and karyotype study. Data regarding age, sex, clinical findings, associated anomalies and treatment were gathered.

Results

From 14 patients, eight cases had PA (aged 2 to 4320 days), and six had DP (aged 2 to 5040 days). Karyotype in all PA patients was 46XY. Five of DP cases had completed, and one had bifid phallus type. All patients were treated surgically after complete investigations, and followed within the period of study. Of eight cases with penile agenesis, 3 patients had no associated anomalies, but anomalies in the other 5 was as follows: hydronephrosis was seen in one patient, Vesico-urethral reflux in 2 cases, mono-orchid in one patient, imperforate anus one, anal stenosis in one, cleft palate one , and cleft lip in one case. These eight patients finally underwent multiple surgical procedures: cystoscopy + cysto-urethrography in 3 cases, urethral transposition in 5 patients, bilateral orchiectomy+ labioplasty in 5 cases, anti-reflux procedures in 2, Vaginoplasty in 5 , urethroplasty+ phalluplasty one, vesicostomy one, anoplasty one, and cleft lip +cleft palate repair in one patient.

Of six patients with diphallus 3 had complete type and normal meatus, and 2 had complete type but abnormal meatus (one Hypospadiac, one Epispadiac) and one had bifid phallus with Hypospadiac meatus. Associated anomalies in these 6 patients was as follow: two imperforate anuses, 3 bladder duplications, one colon duplication, one inguinal hernia, one single kidney, one bifid scrotum, one hemivertebra, one bladder extophy, 2 Hypospadiac meatus, and one Epispadiac meatus. These patients underwent multiple surgical procedures as follow: colostomy in 2 cases, cysto-urethroplasty in 2 cases, phalloplasty in 5 cases, Scrotoplasty in 4 cases, colon resection +anastomosis in one, PSARP 2 cases, cystoplasty in 3 cases, bladder Extrophy repair in one, Mitrofanoff in 1 case.

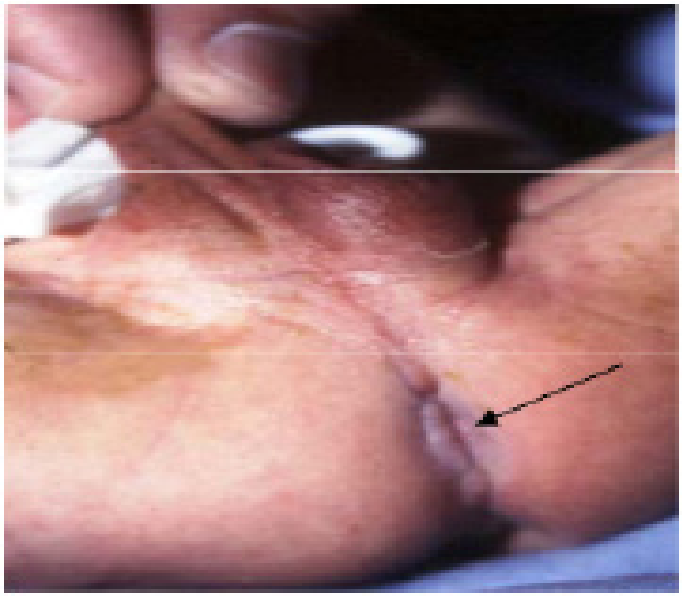

Figure 1: A neonate with penile agenesis and anterior located urethrorectal fistula.

Figure 2: A case with penile agenesis (neonate)

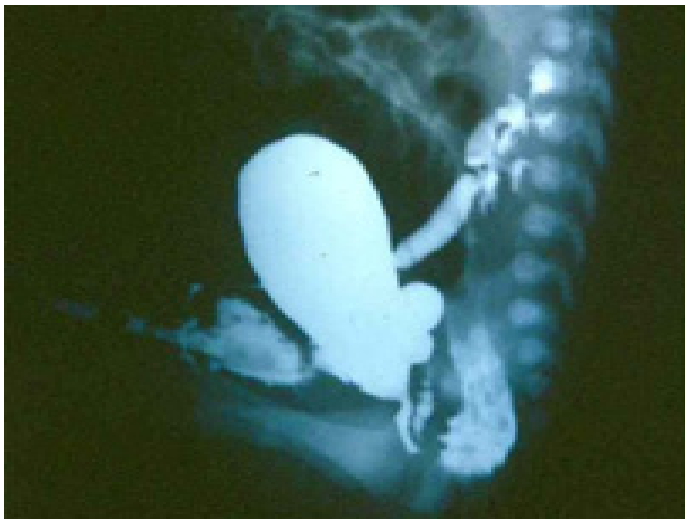

Figure 3: Cystography showing a urethro-anal fistula and vesicourethral reflux in one of our cases.

Figure 4: 2-day-old boy with aphallia after labioplasty

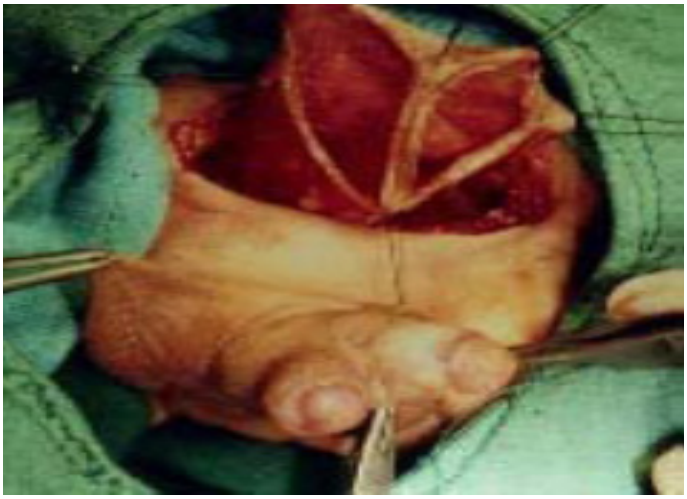

Figure 5: Diphallus and duplicated colon and bladder in a 2-day neonate

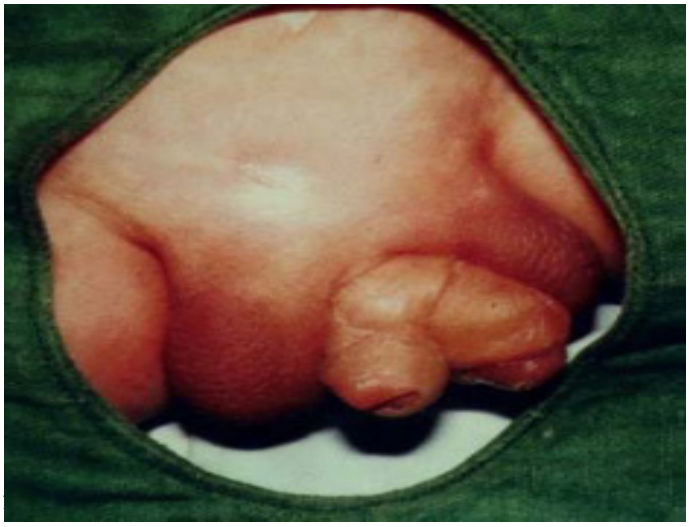

Figure 6: Diphallus in a one year old male

Discussion

Congenital PA is an unusual form of ambiguous genitalia, and penile agenesis is believed to be due to deficient development of the genital tubercle or its non-development in the fourth week of embryogenesis.[20, 21]This is true in the usual case of PA where all other anatomical structures of the caudal axis are normal except for the penis itself.[22,23] As for treatment, if the patient is brought in infancy, feminizing operations are indicated.[24,25] For the cases brought after the second year of life, since sexual identification has took place it is advised to perform masculinizing operations, so as not to disturb the patient psychologically, [4, 7□9] as we have done in one of our cases. Reconstruction of penis in children has been a controversial subject and is a challenge in pediatric surgery. [9, 25] Gilbert et al. [9] performed penile reconstruction in seven children, Perovic [26] in 5 children, and Akozet al. [6] in two children.

In children penis reconstruction depends on patients

age at the time of reconstruction and dimensions of the neophallus and is still a matter of debate.[6, 9] The present consensus is that patients with aphallia should be raised as males .[27] Recently, successful phallic reconstruction in two patients with aphelia were presented from Bologna (Italy) using the lower abdominal wall skin flap as the shaft and the bladder/labial mucosa free graft as the urethra. The procedures were completed at 9 and 17 months of age.

In such procedures (male genitoplasty) one needs to create a long urethroplasty which is not supported by corpus spongiosum. Due to lack of urethral resistance, the long and short term results of this procedure (long urethroplasty) are not pleasing. Patients cannot create a forceful stream of urine. This complication also occured in one of the two cases from Bologna and a scrotal urethrostomy was created in order to manage the problem. We experienced the same complication in one of our own cases in which male genitoplasty was performed. The first step in these cases should be to dissect the urethra from the anorectum and place it in the perineum (as preliminary urethrostomy).[28] This may be done even during the newborn period inorder to escape urinary complications in the future. The child should not be discharged too early without an urethrostomy since there have been reports of death due to chronic renal failure. [28] If the parents have not made up their mind about the gender, they may be given time to think it over.[28]

We had reviewed 8 cases of penile agenesis from 3 different specialized pediatric surgical centers in Tehran. In two of our cases parents refused our medical advice and left the hospital with our receiving further treatment, but in the remaining 6 patients we were able to perform different surgical interventions. Only one of our patients had mid-scrotal type urethral opening.

Duplication of the penis or diphallus is very rare. Within 20 years we could only find six patients with this type of anomaly. Diphallus may appear in clitoris which is also very rare, as reported by Jeffcoate [29], but we did not have any in our series. There are 3 different types of diphallus: diphallus of glans, bifid diphallus, and complete diphallus. Also a fourth category called pseudodiphallia exists.[30]

In our study 5 patients had a single corpus cavernosum. The scrotum may be bifid or normal; in our cases we had 5 bifid scrotums and one normal type. Priyadarshi reported one case of bifid scrotum.[18] Associated congenital anomalies are present in most of diphallus cases. We also had many of them in our study as stated earlier. This is different in other reported cases.[31, 32] Intestinal anomalies are mostly seen along with complete diphallia, and imperforated anus.[33] Only one of our cases had this combination.

Conclusion

In the newborn period, feminizing operations should be carried out for treatment of

PA, but after two years of age, it is advised to perform masculinizing operations. Finally, no surgical intervention should take place without counseling the parents. All the patients with diphallus should undergo careful evaluation because of the high incidence of associated anomalies.

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