Surgical management of complete penile duplication accompanied by multiple anomalies

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Abstract

Diphallus (penile duplication) is very rare and seen once every 5.5 million births. It can be isolated, but is usually accompanied by other congenital anomalies. Previous studies have reported many concurrent anomalies, such as bladder extrophy, cloacal extrophy, duplicated bladder, scrotal abnormalities, hypospadias, separated symphysis pubis, intestinal anomalies and imperforate anus; no penile duplication case accompanied by omphalocele has been reported. We present the surgical management of a patient with multiple anomalies, including complete penile duplication, hypogastric omphalocele and extrophic rectal duplication.

Introduction

Penile duplication (diphallus) is rare and seen in about 1 in of every 5.5 million live births; there are 100 reported cases.1 It is mainly divided into 4 classes: complete, incomplete, bifida, and pseudo diphallus.2,3 Previous studies have reported many concurrent anomalies, such as bladder extrophy, cloacal extrophy, duplicated bladder, scrotal abnormalities, hypospadias, separated symphysis pubis, intestinal anomalies and imperforate anus; no penile duplication case accompanied by omphalocele has been reported.4 The large variability in the degree of duplication and the number and type of additional anomalies in such patients leads to different surgical approaches for each patient.5

In this study, we present the surgical method used in a patient with complete penile duplication, hypogastric omphalocele, rectal duplication, extrophy variant, bladder duplication, proximal hypospadias, anal stenosis, undescended testes and bifida scrotum.

Case report

The infant was born at 37 weeks with a birthweight of 1800 g; the birth was a Cesarean section with a 30-year-old mother with a history of smoking and 4 previous stillbirths. The male infant was referred to our hospital with omphalocele and suspect genital structure. The physical examination revealed hypogastric omphalocele, and suspect genital structure. The physical examination revealed hypogastric omphalocele, and suspect genital structure. The physical examination revealed hypogastric omphalocele, and suspect genital structure. The physical examination revealed hypogastric omphalocele, and suspect genital structure.

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pexy for the intra-abdominal testes, the anterior abdominal wall was primarily closed (Fig. 2, parts A, B). The pathology result of the blunt-ended intestinal segment was tubular rectal duplication. Tubularized insert plate urethroplasty was performed for the penoscrotal hypospadias when the patient was 8 years old (Fig. 3, parts A, B, C). Scrotoplasty is planned for the patient when he becomes 10 years old with full anal continence. However, there is intermittent urinary incontinence and a simultaneous bladder neck injection is planned.

**Discussion**

The most common additional anomalies are intestinal and genitourinary tract anomalies in cases of complete diphallus. The most common intestinal tract abnormality is imperforate anus, and the least common is rectal duplication that may be tubular or cystic in structure.\(^4\),\(^6\) No omphalocele association has been reported. Our case is the first reported case with the complete penile duplication and omphalocele association and the presence of rectal duplication in tubular structure. Treatment of diphallus usually includes excision of the duplicated penile structure and its urethra. Associated anomalies can also be repaired surgically.\(^5\),\(^6\) The prognosis usually correlates with the general condition of the patient and the severity of the anomalies.\(^5\),\(^7\) We believe that the lack of involvement in vital organs, such as the heart and lungs by the other anomalies seen in our patient, had a positive impact on the prognosis.

The karyotype of cases with a diphallus anomaly is usually normal, and the only exception is the balanced translocation t (1;14) (p36.3; q24.3) case.\(^8\) Karyotype analysis performed on our patient was normal. Chemical or genetic effects may occur in the Homeobox (HOX) family of transcription factors; these are important in early embryonic development and regulate several target genes for the formation of many body structures. These genes include BMP-4,
FOXC-1, WT-1, SOX-9 and SF-1. When HOX genes cannot provide the required control, development is impaired and problems may occur when the caudal cell mass in the mesoderm is divided from the genital tubercle and rectum in the urogenital sinus to form the penis with the influence of the affected genes.9,10 Mutations in these genes can cause related syndromes associated with diphallus.10,11

Conclusion

The surgical treatment of penile duplication patients planned should be specific for each patient according to the degree of penile duplication and additional anomalies so as to preserve continence and erectile functions.

Competing interests: Dr. Karaca, Dr. Turk, Dr. Ucan, Dr. Yayla, Dr. Itirli and Dr. Erkal all declare no competing financial or personal interests.

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References


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Fig. 3. The patient before and after the third operation – primary repair of the proximal hypospadias with the tubularized insert plate urethroplasty method (after 8 years).