

AN UNUSUAL COMPLETE DUPLICATION OF THE HINDGUT AND UROGENITAL TRACT: CASE REPORT AND LITERATURE REVIEW

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Abstract

Complete duplication of the hindgut associated with duplication of the genitourinary tract and double termination is an extremely rare condition. The presence of complex duplications have fascinated most lay clinicians and parents. The present study describes a case of duplication of the hindgut associated with duplication of the genitourinary systems. A review of the literature revealed six reported patients with the same condition.

Key words: Complete duplication, hindgut, newborn, genitourinary tract

Introduction

Duplication commonly refers to the alimentary tract from the anus to the oropharynx. The earliest description of duplication may have been reported by Calder in 1733 [1]. Duplication may occur in the sagittal or coronal plane, and there is slight predominance for the male sex and the sagittal plane [1,2]. This anomaly is characterized by different presentations depending on the anatomy of the duplication.

A stillborn newborn with duplication of the bladder, urethra, vagina, uterus, and anus was first reported by Schatz in 1871 [2]. This condition was subsequently reported by Suppiger in 1876. Since that time, there have

been few reports of this condition [3]. However, treatment remains a dilemma. Beach et al. reported that duplication of the hindgut and genitourinary can be separated because they are usually not fused and have a separate blood supply, and thus resection of one side of the duplication is possible [4]. When reconstructive surgery is not planned, the possibility of neoplastic changes in alimentary tract duplications should be kept in mind [5]. We herein report a case of total complete duplication of the colon, anus, vagina, vulva, urethra, and bladder in the coronal plane with no additional congenital anomalies. In our patient, both fecal and urinary functions were normal.

Case

A 10-day-old term female infant was admitted with fecal and urine material passing from two openings in the perineum (fig. 1a, 1b, 2). There were no additional congenital anomalies. The physical examination revealed a separated double vagina, double vulva, double clitoris, double urethra, and two well-formed ani. One anus was in the right hip, and the other anus was in the left left hip. The two anal openings were widely separated with no communication between them.

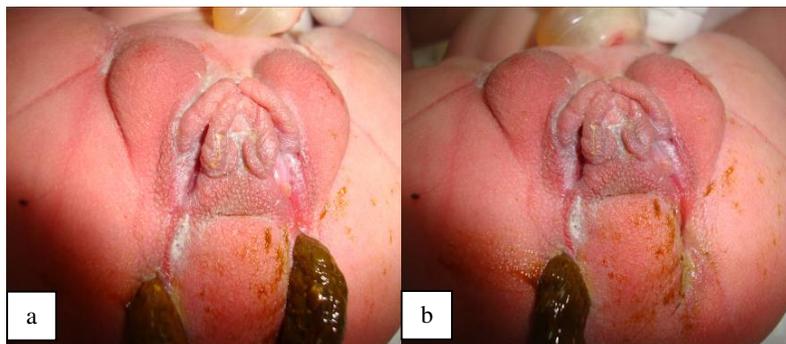


Figure 1a, 1b: The figures showing the duplication of extraurogenital strictures and ani.



Figure 2: The figures showing the extraurogenital strictures at 6 months old of the case.

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Due to the complexity of these abnormalities, the patient underwent plain X-ray, ultrasonography, contrast X-ray, magnetic resonance imaging, and videocystourethrography studies for investigation of these anatomical associations. Imaging of the upper digestive tract showed a normal esophagus and stomach. Contrast X-rays were obtained with installation of dye through the two anal openings, two urethral openings, and the upper digestive tract. Two colons were visible up to the cecum, two bladders were also visible, and the left bladder demonstrated reflux. The patient had no tailbone or pelvic anomalies. Ultrasonography revealed a horseshoe kidney at the pelvis. At the 6-month follow-up, the defecation and urinary functions of the child were normal.

Discussion

Complete duplication describes the presence of two of the same organ with separate walls of mucosal and muscular

layers; each organ empties through its own tract. Complete duplication of the hindgut is rare and is usually associated with genitourinary tract duplications and neural tube defects [6,7]. In patients with this kind of complex abnormality, both openings are usually blind or inadequate [8]. The present patient had duplication of the hindgut and genitourinary systems, but both anal and urethral openings were adequate and functional. This anomaly has a widely variable anatomy and presents in different ways [6]. There is a female predominance [6]. The present patient was female; however, when we reviewed the literature, we found more male than female patients. In our review, we found seven similar cases of a duplicated hindgut and genitourinary tract (table 1). Most cases have been reported both of ani or one of ani had any type of anal atresia, and mostly included sacrum and vertebral column anomalies [8]. The present case showed normal function of both ani; however, both were malpositioned and without vertebral anomalies.

Table 1: Demographic and clinical characteristics of the present case with cases in the literature duplication of the hindgut and urogenital system

No	Cases	Age / Gender	Anomaly	Additional anomalies	Operation	Outcomes
1	Banu T et al.[7]	42-day/ Female	2 ani,2 vagina, 1 vulva, 1 clitoris, 1 urethral orifice	Total colon duplication, vestibule fistula ani, polidaktili	No	Follow up for 17 months growing well
2	Azmy AF[8]	1-day/ male	2 penis, 2 ani,2urethra, 2 bladder, total colon duplication	Adbominal hernia, omphalocel, pes ecinovarus, horse-shoe kidney, vertebral column abnomaly, undescending testises, thoracic scoliosis	Neorecnum and anal canal, colon divided, bladders are jonied, testis was descended, and excision left phallus, urethras were corresponding	Follow up for 2 years, good stream urine, has regular bowel movement growing well
3	Okur Het al. [5]	1-day/ female	2 ani,2 vagina, 2 colon,1vulva, 1 clitoris, extrophy of the bladder	malrotation	no	Un clear
4	Smith ED[11]	Not clear	2 ani and 2 genitouriner tract	Not clear		
5	Cohen SJ[12]	1-day/ male	2 ani,2 fallus, 2 urethra 2 bladder, 2 colon	Anal atresia fistula with bladder	Staged procedure	Not clear
6	Liu et al. [13]	13 year/ male	Incomplet 2 phallus,2 colon 2 urethra,2 bladder, 2 ani	Hemicorpus vertebra fusion, accessoir ear	Remeved colon, septum of bladder, and a fallus	Follow-up for2 years
7	The present case	10-day/famale	2ani, 2colon, 2 urethra, 2 bladder, 2 clitoris, 2 vagen	Horse-shoe kidney	No	6 months follow-up

Most previous studies are case reports. Therefore, their descriptions were not sufficient to explain the etiopathogenesis of this complex anomaly. Several theories have been proposed regarding the etiology of this duplication. However, the embryologic basis of it is speculative and unknown. One hypothesis is that duplication of the hindgut and geniturinay structures is a result of partial or abortive splitting or twinning of embryonic structures

during the early stages of development [9-11]. The separation may start with a divided notochord that fuses again cranial to the separation, beneath the paired notochord where the endoderm forms two hindguts, each of which gives rise to the allantoic stalk and a cloaca [8]. This explanation does not apply to the present case because no vertebral column abnormality was present. In patients with these associated abnormalities, one or both ends of the

urinary tract or intestine is usually either blind or has an inadequate opening. In the present case, all openings were adequate and at the coronal plane. Previous studies have shown a slight male predominance, and the sagittal variant appears to be more common than the coronal plane. However, the coronal is more common than the sagittal plane in females.

Duplications present in a variety of ways depending on their size, location, and associations. Most duplications have few or no symptoms and are therefore diagnosed incidentally or late in life. The present patient had a pair of extra urogenital structures and ani; therefore, she was diagnosed early in life.

Antenatal diagnosis of duplication has become common with the availability of prenatal ultrasonography [1].

Not only plain radiography, but also postnatal ultrasonography, contrast studies, computed tomography, and magnetic resonance imaging may help to localize the duplication.

Previous studies have identified malignant changes of the duplicated segment and removal of the segment was thus performed. The operative technique depends on the duplicated segment. The present patient was female, so the two anuses and urethras were not apparent. Therefore, her

condition may not socially affect the patient later in life. In our case, both urethras and anuses were well functional. Poor postoperative activity of the rectal muscles was anticipated; therefore, surgical reconstruction was extended. In terms of operative timing, we suggest waiting until it is apparent which duplicated segment has impaired function. The possibility of neoplastic changes must be kept in mind during follow-up. Banu et al. did not treat their patient, and no problems occurred during the 17-month follow-up. Another previous study described two female patients who were 22 and 25 years old, respectively. They had normal lives, and the 25-year-old gave live birth [7]. The other had a miscarriage and was subsequently diagnosed with this condition [6]. Therefore we suggest that may be best to accept the permanence of the two ani and urethras.

Three of seven patients who underwent surgery were male, and their operations were long and comprised many stages [8,12,13]. Therefore, the goal of treating this kind of complex anomaly is to relieve symptoms, not to restore normal anatomy.

In conclusion, the goal of treatment, case with many complete duplication should be monitored closely without surgery. Duplicate segments in which is insufficient according to plan surgical treatment.

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