Journal of Pediatric Surgery (2012) 47, E19–E23

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Journal of Pediatric Surgery

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Perinatally discovered complete tubular colonic duplication associated with anal atresia☆

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Received 18 May 2011; revised 17 January 2012; accepted 24 January 2012

Key words:
Duplication;
Colon;
Anal atresia;
Children;
Malformation

Abstract Complete tubular colonic duplication (CTCD) is exceedingly rare. The association of CTCD with an anorectal malformation is unusual. This malformation may be found unexpectedly at laparotomy. We present 3 cases of surgically proven neonate CTCD discovered at laparotomy for anal atresia. We reviewed the mode of clinical presentation, the imaging, and laparotomy findings. Our series illustrates that this rare disease presents perinatally in association with anal atresia, with or without other associated anomalies.

Complete tubular colonic duplication (CTCD) is exceedingly rare [1]. The association of CTCD with an anorectal malformation is unusual. This malformation may be found unexpectedly at laparotomy [2]. We present 3 cases of surgically proven neonatal CTCD discovered at laparotomy for anal atresia. The aims of this study were to illustrate the clinical manifestations and to describe and discuss the imaging features and treatment options.

1. Patients and method

This is a retrospective study done in the pediatric surgery department of Monastir between 1984 and 2000. We reviewed the mode of clinical presentation, imaging, and laparotomy findings in 3 neonatal cases with surgically proven CTCD. The mode of clinical presentation and means of definitive diagnosis were recorded.

1.1. Case 1

A female infant was referred to our institution at 2 days of age for anorectal malformation discovered at birth. Clinical examination revealed an anal atresia and split vaginal orifice. There was no evidence of rectovaginal or rectourethral fistula. An invertogram showed high type of anorectal malformation. A colostomy was performed as an emergency. During surgery, a CTCD was noted. A contrast enema confirmed the diagnosis of CTCD with a proximal communication and with both limbs ending blindly in the pelvis without either an enterourinary or enterogenital fistula (Fig. 1). An anorectal reconstruction was successfully performed at 5 months of age

☆ Conflict of Interest: None.

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doi:10.1016/j.jpedsurg.2012.01.082

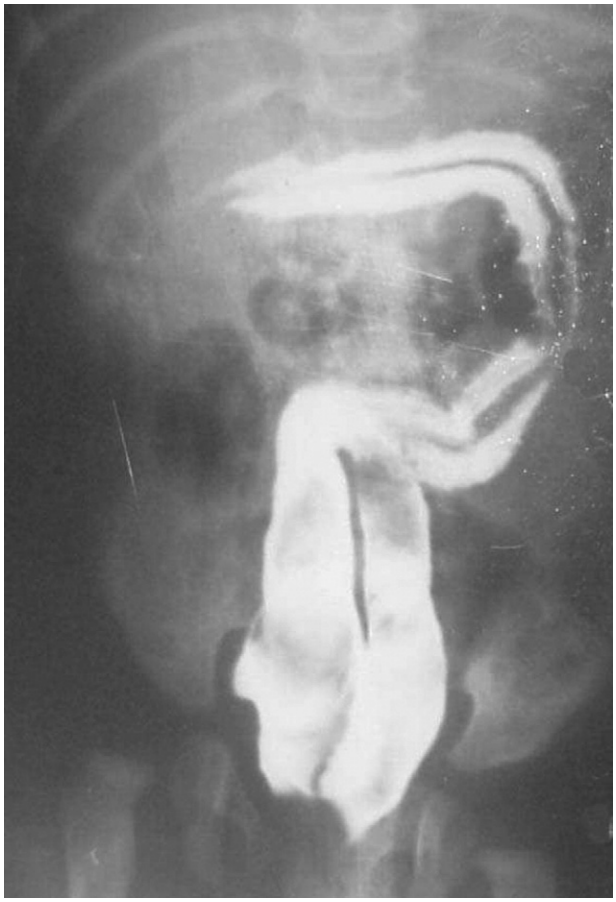

Fig. 1 Contrast enema through both colostomy orifices (case 1), showing CTCD with both limbs ending separately blindly in the pelvis without neither enterourinary nor enterogenital fistula.

with the "Stephens procedure." A distal communication was made surgically between the duplicated colons to enable emptying of the twin colon into the anal canal. During surgical exploration, a single uterus with 2 fallopian tubes were found. The colostomy was closed 3 months later. Microscopic examination showed a muscular coat in 2 layers and colonic mucosal lining that was indistinguishable from the normal colon, except the cul-de-sac of the duplication, which was covered with urinary-type epithelium. The patient is doing well 17 years after the surgery.

1.2. Case 2

A newborn baby boy was referred to our institution for an anorectal malformation discovered at birth. Physical examination revealed anal atresia with history of meconium passed in the urine. There was neither cutaneous fistula nor perineal bulge convexity at the supposed site of the anus. There was no anomaly of external genitalia. An invertogram showed high type of anorectal malformation. A right transverse colostomy was performed. At surgical exploration, the baby was found to have a complete side-to side colonic tubular duplication with presence of 2 ceca and 2 appendices. Contrast enema studies showed a CTCD that communicated proximally at the level of the ileocecal junction. Distally, both segments were not fused, resulting in a cul-de-sac twin limbs with both segments ending as fistulae to the urethra (Fig. 2A and B). An anorectal reconstruction was successfully performed at 5 months of age. A distal communication was made surgically by creation of a communicating window of approximately 5 cm between the 2 sigmoid colons and rectums as low down as possible. We excised the bilateral rectourethral fistulae through a posterior sagittal approach (Penã and Devries procedure) [3]. The colostomy was closed 2 months later. Twelve years after surgery, the patient has only intermittent gastrointestinal symptoms comprising alternating diarrhea and constipation.

1.3. Case 3

A newborn baby girl was transferred 7 hours after birth for an anorectal malformation discovered at birth. Physical examination revealed anal and vaginal atresia, without evidence of a fistula. Radiographs of the spine showed lumbosacral scoliosis with sacral hemivertebra. An invertogram done at 24 hours of age showed high type of anorectal malformation. A colostomy was performed at 2 days of age. During surgery, a CTCD was noted, communicating proximally with presence of 1 cecum and 1 appendix. Contrast study via the distal colostomy orifices confirmed the diagnosis and revealed the presence of 2 rectourinary fistulae allowing opacification of 2 bladders (Fig. 3). Ultrasonographic examination showed a bilateral ureteral dilatation and 2 bladders with no presacral mass. The intravenous urography showed duplicated bladder and urethra and confirmed bilateral ureteral dilatation. These findings were confirmed by an endoscopic examination, which revealed 2 rectourethral fistulae. There was only 1 vagina, which was atretic at its distal portion.

Surgical correction was planned, but unfortunately, the patient died before this could be implemented.

2. Discussion

Gastrointestinal duplication is an infrequent congenital abnormality. The reported incidence is 1 in 5000 live births [2,4]. It represents only 0.1 % to 0.3 % of all malformations [5,6]. Colonic localization accounts only for 4% to 18 % of all gastrointestinal duplication [7]. Duplications of the colon are most commonly isolated cystic structures, but rarely are they complete, side-by-side duplications of the entire colon, including the rectum. They can occur at any age, but most are discovered during the neonatal period or infancy particularly when they are associated with anorectal or urogenital malformation [6,8,9]. In our series, no case of CTCD was