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Case Report

## A low Anorectal Malformation Exposing a Complete Colorectal Duplication: A Case Report

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### Abstract

The gastrointestinal duplication is a rare congenital malformations and total recto-colonic duplication represents only 12% of them.

We describe an association of a total recto-colonic duplication with a low ano-rectal malformation in a one month girl. The duplication suspected during the first surgery for the ano-rectal malformation, and we realized after this surgery a barium enema confirmed the diagnosis of total colonic duplication with vestibular fistula. Barium enema is the best exam to objective a recto-colonic duplication. The fenestration is an interesting alternative treatment allowing to save the colon. However, they involve a long term follow up of this patient.

This case proves the importance to take time to realize some radiologic examination when we are in abnormal situation to make the good choice and so the adequate surgery.

**Keywords:** Colorectal Duplication; Fistulogram; Fenestration

### Introduction

Gastrointestinal duplications are rare congenital malformations with an incidence of 1: 4500 [1,2]. Among these, recto-colonic duplications represent only about 12% of the pediatric cases [1,2]. We herein report a total recto-colonic duplication case diagnosed during a low anorectal malformation surgical repair procedure.

### Case Report

A one-month-old baby girl was referred at our institution because her mother had observed she was passing stools from her vagina since birth, with recent discomfort.

On clinical examination, the vulva, vagina and urethra were normal, but the anus appeared narrow and located immediately behind the vulva. Catheterization of the anus indeed induced a bowel motion through both the anus and the vaginal vestibule. We supposed a diagnosis of anterior anus with a vestibular fistula. Pre-operative assessment included abdominal and medullar ultrasound as well as X rays of sacrum and coccyx, all normal.

Anoplasty with fistula resection was then scheduled. During the procedure, attempts were repeatedly made to reach the rectal lumen through the genital fistula, but despite the catheter was easily introduced and pushed at more than 10cm length, no connection of the fistula with the rectum could be identified.

We thus performed the anal enlargement and only moved the fistula from the vestibular wall to the perineum. Further assessment of the fistula included a barium enema and a fistulogram which revealed a total colonic duplication (Figure 1, 2).

Subsequent MRI confirmed the diagnosis of total colonic duplication joining a single sub-hepatic caecum (Figure 3).

Two months later, we performed resection of the duplication

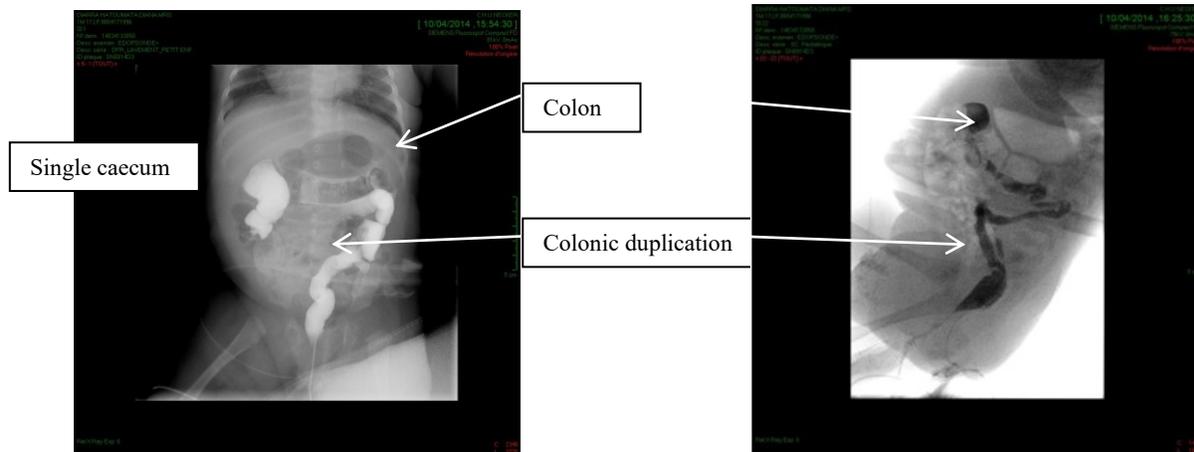


Figure 1, 2: fistulogram (face and profil): barium enema by fistula.

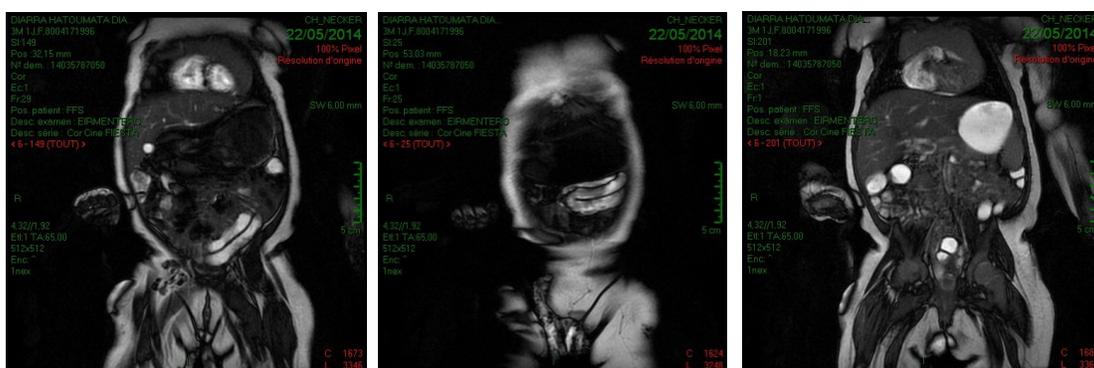


Figure 3: MRI: total rectocolonic duplication with a single caecum.

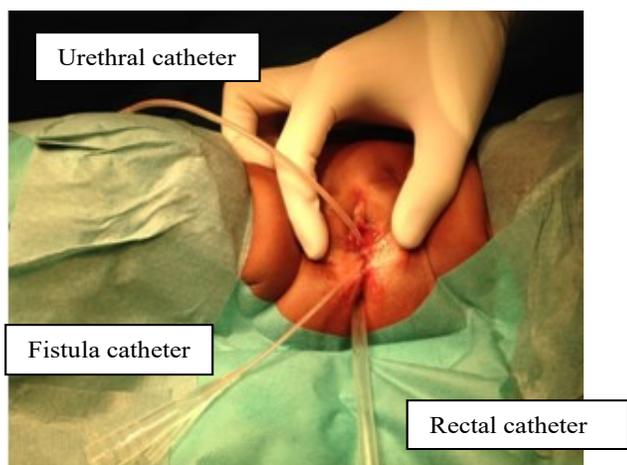


Figure 4. catheterism.

limited to its rectal part (including the fistula), with anastomosis of the two colonic segments above (Figures 4-7).

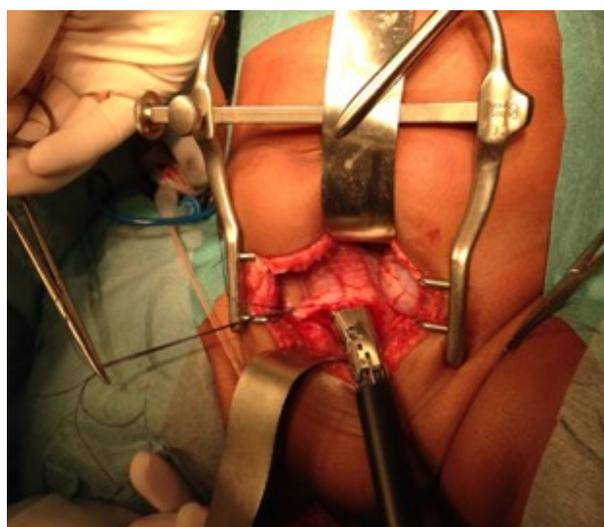
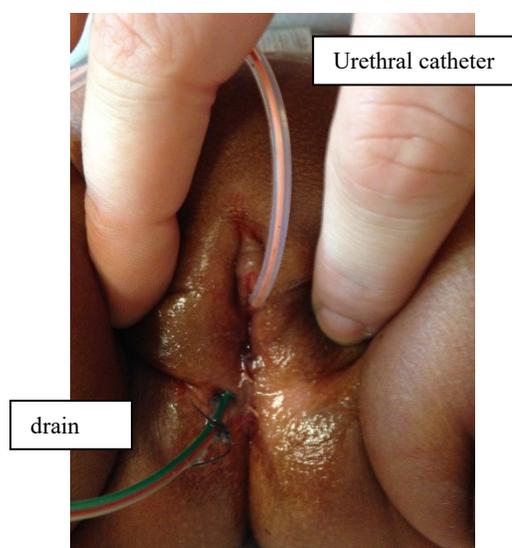


Figure 5. mechanic anastomosis between sigmoid duplication to the right and sigmoid to the left.



**Figure 6.** Perineal fistulectomy.



**Figure 7.** End of surgery: final aspect.

The post-operative course was uneventful.

The pathological examination confirmed a normal rectal wall without gastric heterotypic mucosa. At two years follow-up, the girl has a good looking perineum. She passes stools normally without any trouble of continence.

## Discussion

Duplication of the gastrointestinal tract is a rare pediatric malformation that can potentially affect any portion of the gastrointestinal tract. In 1937, William E Lass defined this defect by three criteria: 1) a smooth muscle layer; 2) a digestive tract epithelium; 3) a contiguity with some parts of digestive tract [4].

There are two kinds of duplication. The first and most frequent (80%) is cystic and has no communication with the bowel lumen. The second type is tubular and may communicate with intestinal lumen [2].

Hindgut duplications account for 7% to the colon and 5% to the rectum of all gastrointestinal duplications [5].

The association of hindgut duplication and anorectal malformation is not the rule [1, 6]. So far, less than a hundred patients have been reported with this condition, since Smith established the first hindgut duplication classification in 1969 [1]. He reported 32 children with hindgut duplication, out of whom only 9 (28%) also had an anorectal malformation, and described three groups regarding the status of duplication colon ending (blind ending, terminating as a fistula, or as a true anal duplication).

Yousefzadh and al. reported later 57 colonic duplications and enhanced Smith's classification with 5 categories: 1) perineal ani, 2) duplication in females with fistula, 3) duplication in males with fistula, 4) duplication without fistula with imperforate anus, and 5) single perineal anus with communicating duplication. In their series, 28 patients presented a perineal fistula [3]. A posterior perineal fistula was also described in 45% of colonic duplication by La Quaglia [7].

The clinical presentations were rectal prolapse, bleeding, constipation, or the persistence of a perineal abscess despite antibiotic treatment, drainage and/or excision [7, 8]. The finding may also be fortuitous, often during another surgery [9], such as colostomy for anorectal malformation with or without fistula, as recently reported [1, 6, 10]. In our study, Sarpel and al. recommend to search for hindgut duplication in any patient with normal anus and anterior perineum fistula (recto-vestibular or recto-vaginal fistula) [11].

Overall, 80% of duplication cases are diagnosed before 2 years of age [2].

When duplication is suspected, a radiologic evaluation has to be done to assess the duplication localization and extension. The barium contrast enema studies are the keys of the tubular colonic duplication diagnosis [12], and should be performed through the different pertaining orifices (anus, fistula and/or stoma) [3, 6].

Other imaging modalities such as computed tomography or magnetic resonance may also be helpful to investigate the duplication and associated anomalies [2, 12], considering that 60% of duplications are associated with genito-urinary anomalies [1].

The treatment in most cases is a complete duplication resection with primary anastomosis because of the risk of heterotopic gastric mucosa (35%) [3] and malignant change in adulthood [7, 10]. However, when complete resection is impossible with acceptable digestive loss, partial resection with complete mucosal excision can be done, reducing bleeding and malignancy risk to nothing [13].

In hindgut duplications, total resection is seldom a good op-

tion, as the common blood supply and length of colonic duplication increase the risk of a total colectomy [11, 13].

The identification of colon itself in comparison to the colonic duplication is difficult because they are not necessarily mesenteric [1, 3]. Alternative treatment option is then to perform an internal drainage of tubular colonic duplications, a “fenestration” between the two colons [11, 12, 14]. Furthermore, ectopic gastric mucosa in hindgut duplications has never been described in the literature, probably advocating for more conservative procedures in these cases than in other digestive locations. However, long-term follow-up seems to be mandatory to detect complications like constipation, bleeding, perforation or malignancy leading to a radical surgery [7, 10].

## Conclusion

A low anorectal malformation may hide another condition, such as colonic duplication in our case. When in unusual situations, we think one should assess the malformation with radiologic examinations (contrast enema and/or MRI) even if the association remains uncommon, in order to perform the adequate surgery.

The “fenestration” is an interesting alternative treatment enabling the preservation of the colon, but long term follow up of the patient is mandatory.

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