



Rare anorectal malformation with a non-terminal colovesical fistula

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ABSTRACT

We describe a unique case of anorectal malformation (ARM) with a non-terminal colovesical fistula. While some aspects are similar to the congenital pouch colon (CPC), the differences make it a distinct form.

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Key words:

Anorectal malformation
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1. Case report

During a pregnancy made possible by reproductive technology, prenatal echography performed at 22 weeks revealed a tetralogy of Fallot. Amniocentesis at 16 weeks had shown a normal 46, XY karyotype. The child was born by caesarean section at 34 weeks following premature rupture of membranes. Birth weight was 1700 g. At birth, clinical examination showed an imperforate anus with meconium stained urine, a penile distal hypospadias and an empty left scrotum. Tetralogy of Fallot with pulmonary stenosis was confirmed by the work-up and corrected surgically at 5 months of age. Associated diagnoses at the work-up included a left anterior costal fusion; L2-L4 vertebral fusions without myelodysplasia seen on MRI; and bladder-sphincter dyssynergia with important post-void residual volume was confirmed by urodynamic study.

Based on the high form of ARM with an entero-urinary fistula, a laparotomy was performed on day 1. During this procedure we found a permeable colovesical fistula 10 cm distal to a normal ileocecal valve and a blind-ending mildly dilated distal colon. The colonic distension was localized to the middle portion of the colon, on either side of the fistula. The rectosigmoid colon was considered absent. The blind end of the colon was identified above the peritoneal reflection in the left upper quadrant and marked with a non-absorbable suture (Fig. 1). The vasculature was considered

abnormal: the mesentery was short and poorly developed, containing a short vascular pedicle. The vascular arcade was continuous between the ileocecal valve and the blind segment of the distal

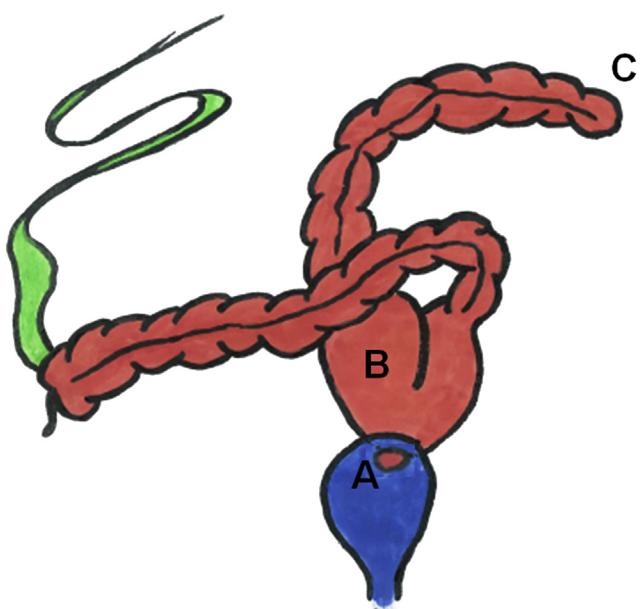


Fig. 1. Diagram of the malformation. Blue: bladder. Red: Colon. Green: small intestine. A: colovesical fistula. B: distension of the colon. C: blind segment of the distal colon.

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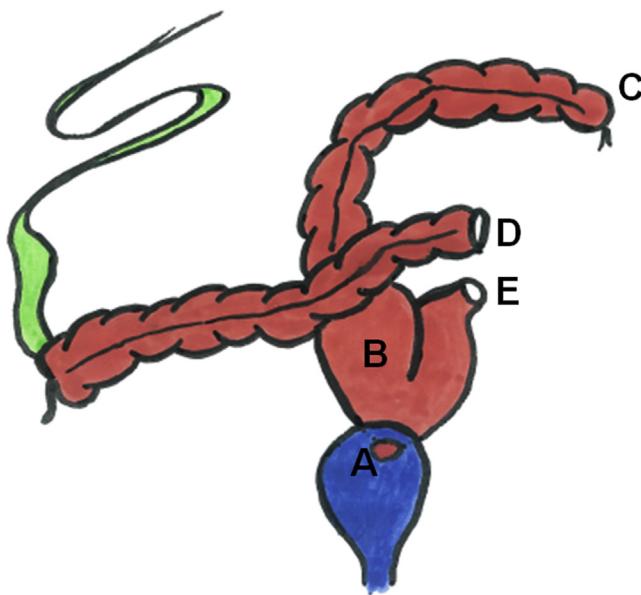


Fig. 2. Postoperative status. Blue: bladder. Red: colon. Green: small intestine. A: colovesical fistula. B: distension of the colon. C: blind segment of the distal colon. D: proximal colostomy. E: distal colostomy.

colon. Moreover, exploration of the abdominal cavity found a hypoplastic left intra-abdominal testis and a Meckel's diverticulum. A divided colostomy was placed between the ileocecal valve and the colovesical fistula (Fig. 2) and the Meckel's diverticulum was resected at that time. A postoperative loopogram of the excluded part of the colon through the distal colostomy confirmed the colovesical fistula (Fig. 3), which had not been visualized on cystography (Fig. 4).

The definitive repair of the ARM was performed at the age of 13 months. The colovesical fistula was closed through a Pfannenstiel incision (Fig. 5). We measured 15 cm of colon left beyond the fistula down to the cul-de-sac. Vascularization of the distal intestine was

unusual with a very short vascular pedicle. For this reason, 4 cm of distal colon was resected to allow a pull-through without tension. A plastic catheter fixed on the distal end of the colon and positioned at the colonic cul-de-sac helped us find the colonic distal end during the pull-through. The procedure was performed in the prone position by the postero-sagittal approach described by Peña [1].

The postoperative course was marked by a mechanical intestinal obstruction responding to medical treatment. Anal dilatations were started 2 weeks after operation and the colostomy was closed two months after the pull-through. The patient has normal bowel movements 1 to 3 times a day and fecal continence seems to be good 7 months after the pull-through (at 2 years of age).

2. Discussion

The most common and recent international classification of ARM, a modification of Peña's classification [2], was elaborated during the Krickenbeck conference [3]. The major clinical groups are classified according to the presence and the position of the fistula (perineal fistulas, rectourethral fistulas, rectovesical fistulas, vestibular fistulas, cloacal malformations, patients with no fistula and anal stenosis). Rare or regional variants were described but none matched the defect described above. Our case has certain similarities with a congenital pouch colon (CPC) first described by Spriggs in 1912 [4]. CPC is an extremely rare variant of ARM with most of the cases being reported in northern India where this malformation represents 7–17% of ARM [5,6]. Only a few reports have originated from other parts of the world.

In CPC, part or whole of the colon is replaced by a distended pouch ending in a large fistula communicating with the genitourinary tract, more commonly with the bladder [6]. In our patient the distal colon ended blindly; a fistula connected the middle part of the colon to the bladder. The opacification by the distal stoma allowed understanding and confirmation of the unusual type of fistula. The colon proximal to the fistula was slightly dilated, but had a normal caliber over 15 cm of corresponding transverse and descending colon. As described in the literature [6], the blood supply to the colon was abnormal as well. The distal colon, the

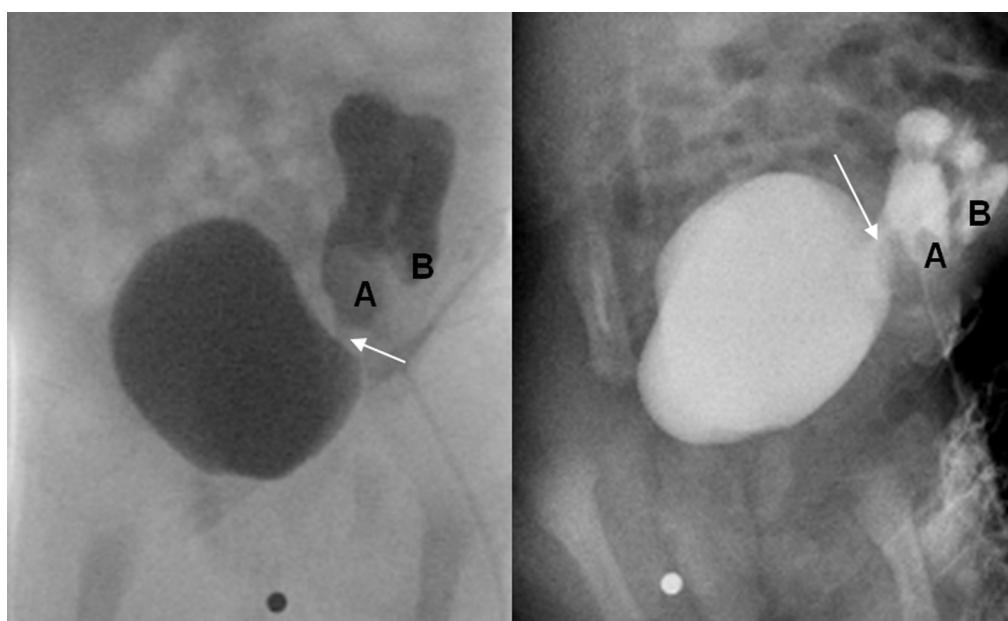


Fig. 3. Opacification of the excluded part of the colon through the distal colostomy. White arrow: colovesical fistula. A: distal colostomy. B: blind segment of the distal colon.

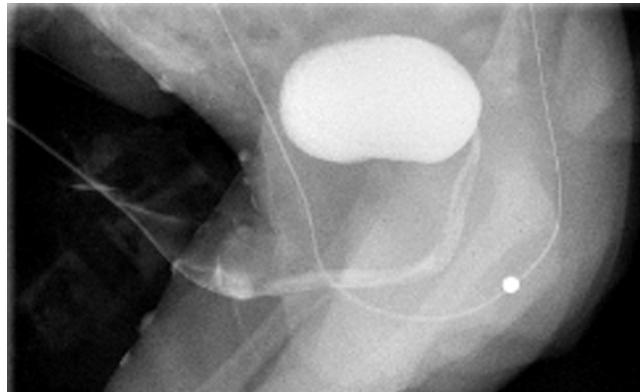


Fig. 4. Cystography: no demonstrated fistula.

rectum and the inferior mesenteric artery were considered absent. These features, together with a Meckel's diverticulum, are frequent in the CPC. However, based on the unusual position of the colovesical fistula and the absence of important colonic distension, we kept the diagnosis of a rare type of ARM, rather than an incomplete CPC.

We decided on staged procedures according to Peña's technique [1] due to the high type of ARM and the associated heart disease. The main differences between ARM and CPC surgical treatments rely on the management of the fistula and the type of colostomy during the first surgical intervention: in CPC, Gupta et al. [7] suggest a primary division of the fistula and an ileostomy or a window colostomy. The length of colon available between the ileocecal valve and the fistula allowed us to perform a divided colostomy. As we chose not to divide the fistula during the first procedure, this solution offered us both the possibility to carry out a loopogram and to isolate the distal colon and the urinary tract from fecal material. The division of the fistula was done during the second procedure, after confirmation of the diagnosis. An abdomino-perineal pull-through is recommended in both management of rare type of ARM and incomplete CPC [7,8]. Laparoscopy was not considered at the time of the abdomino-perineal pull-through.

The presence of a sufficient length of colon in CPC enables total resection of the pouch and an abdomino-perineal pull-through of normal colon [7]. In our patient no distension of the colon was found, therefore no resection was needed. However the vascularization of the colon was abnormal, as described above,

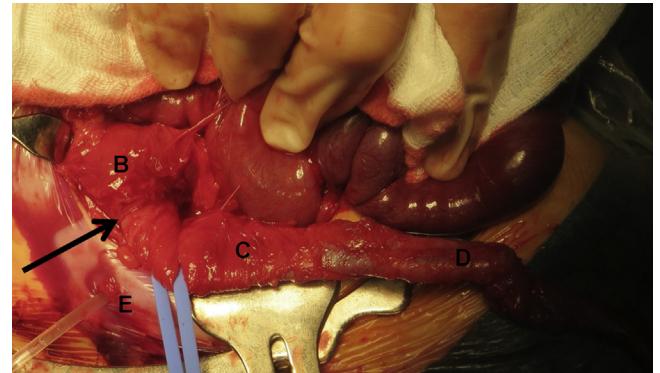


Fig. 5. Intraoperative view: Black arrow: colovesical fistula. B: bladder. C: distal end of the colon which was pulled-through. D: colon (distal part). E: distal stoma.

and so the distal part of the colon, at the cul-de-sac, had to be resected to allow a pull-through without compromising the distal vascularization.

3. Conclusion

Although similar to a CPC in some respects, this case differed because the colovesical fistula was not at the distal aspect of the colon and a colonic distension was notably absent. To our knowledge these differences made this a unique case of rare high ARM, which required a staged surgical repair.

References

- [1] Peña A. *Atlas of surgical management of anorectal malformations*. New York: Springer-Verlag; 1990.
- [2] Peña A. Anorectal malformations. *Semin Pediatr Surg* 1995;4:35–47.
- [3] Holschneider A, Hutson J, Peña A, Beket E, Chatterjee S, Coran A, et al. Preliminary report on the International Conference for the Development of Standards for the Treatment of Anorectal Malformations. *J Pediatr Surg* 2005;40:1521–6.
- [4] Spriggs NJ. Congenital occlusion of the gastrointestinal tract. *Guy's Hosp Rep* 1912;76:143.
- [5] Wakhlu AK, Wakhlu A, Pandey A, Agarwal R, Tandon RK, Kureel SN. Congenital short colon. *World J Surg* 1996;20:107–14.
- [6] Saxena AK, Mathur P. Classification of congenital pouch colon based on anatomic morphology. *Int J Colorectal Dis* 2008;23:635–9.
- [7] Gupta D, Sharma S. Congenital pouch Colon. In: Holschneider A, Hutson J, editors. *Anorectal malformations in children*. Berlin, Heidelberg: Springer; 2006. p. 211–21.
- [8] Mathur P, Saxena AK, Simlot A. Management of congenital pouch colon based on the Saxena-Mathur classification. *J Pediatr Surg* 2009;44:962–6.