PST | PEDIATRIC SURGERY IN TROPICS

TOTAL TUBULAR COLONIC AND APPENDICEAL TRIPLICATION CYSTS WITH AN ILEAL DUPLICTION AND RECTOVAGINAL FISTULA: A CASE REPORT AND LITERATURE REVIEW

Teboho Shabalala, Hansraj Mangray, Sanele Madziba, Amanda Ngobese, Yashlin Govender, Tafadzwa Dube-Zhiruvik, Zizodwa Nginase

[1]Department of Paediatric Surgery, Grey's Hospital in Pietermaritzburg, and [2]College of Health Sciences - Nelson R Mandela School of Medicine, University of KwaZulu-Natal, Durban, South Africa

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Keywords	**Abstract**
Colonic triplication, Duplication cyst, Rectovaginal fistula, Appendix	**Introduction:** There are few cases reported with total colonic triplication in the literature, and none have been reported with the association of ileal duplication, triplicated appendix, and rectovaginal fistula. Our case is the first in the literature where total colonic triplication is associated with all the above-mentioned abnormalities. **Case report:** A unique case of a total tubular colonic triplication, ileal duplication, triplicated appendix associated with a rectovaginal fistula in a 5-month-old female who presented with a history of stool draining from the vagina and anus, as well as a mass prolapsing from the anus when straining. The digital rectal exam could not advance more than a centimeter (cm). Diagnostic laparoscopy showed a dilated colon, and a divided colostomy showed three lumens proximally and distally. Stool is draining through two lumens postoperatively. CT imaging confirmed total colonic triplication. Laparotomy showed duplication of the terminal ileum 25cm, both communicating with a triplicated caecum, triplication of the appendix, and total colonic triplication. There was no evidence of bowel obstruction, but a rectovaginal fistula was confirmed. A colostomy was brought down, appendicectomy was performed, and the colon was shaved off the vagina. The vaginal defect was repaired, then enterotomy was extended, and a common rectal channel was created. The rectal wall was repaired, and the divided

	colostomy was redone with each stoma having three lumens. Albeit a coarse recovery in PICU, our patient has been discharged home. On follow-up, she is growing, and the stoma is functioning well. **Conclusion:** Colonic triplication is rare with complex anatomy; management should be multidisciplinary, and surgical intervention should be reserved for patients who are symptomatic or at risk of complications.
Abbreviations	PICU: Pediatric intensive care unit GIT: Gastrointestinal tract EDC: Enteric duplication cysts U/S: Ultrasonography PSARP: Posterior Sagittal Anorectoplasty

INTRODUCTION

Intestinal triplication cysts are a rare congenital abnormality of the gastrointestinal tract (GIT). [(1)] They may occur anywhere in the digestive tract. To date, there have been only 11 cases reported in the literature, with our case being the first reported in Africa. [(2)] There are a few cases reported with total colonic triplication in literature. [(1-4)] Nonetheless, our case is unique in that it is the first case to have associated ileal duplication, triplicated appendix, and rectovaginal fistula in addition to the total colonic triplication.

CASE REPORT

We present a unique case of an ileal duplication, triplicated appendix, and a complete colonic triplication cyst associated with a rectovaginal fistula in a 5-month-old female. She was born via normal vaginal delivery at term gestation and was unexposed to retroviral disease. Her mother had an uneventful antenatal history; however, no antenatal ultrasound scan was performed. The patient presented with a history of a mass protruding from the anus when straining and stools draining via the vagina and anus. He was feeding well and thriving. Clinical examination revealed no dysmorphic features, a non-distended, soft abdomen, and no palpable masses. Normal female external genitalia were noted, and the anus was in a normal position. On rectal examination, the finger could only be advanced up to 1 cm, and a mass was palpable and observed on straining.

The babygram revealed scoliosis, a hemivertebra of T12, asymmetrical ribcage with 11 ribs on the left and 12 on the right, and a normal bowel gas pattern. The abdominal ultrasound

was normal. A provisional report on CT scan of the abdomen and pelvis suggested anorectal duplication. (Fig 1-3) A vaginogram confirmed a fistula communicating with the colon with the contrast extending up to the transverse colon. Given the above findings, the patient was taken to the theatre for further evaluation. At laparoscopy, a large, dilated colon was noted. The decision to bring out a stoma was made during which features of colonic triplication were noted. On postoperative review, the mother reported stools draining from the two openings on the proximal stoma.

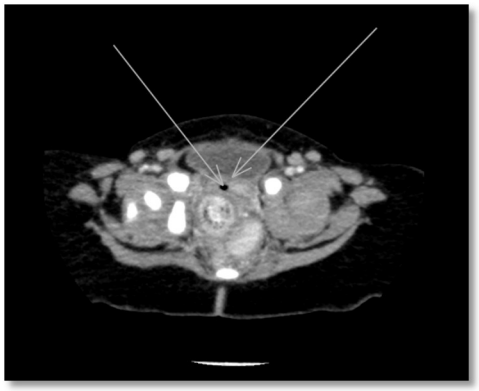


Fig 1. An axial enhanced CT scan imaging demonstrates a rectovaginal fistula as shown by the arrows

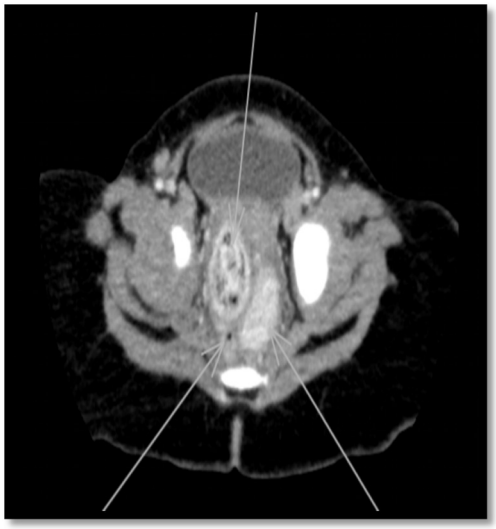


Fig 2. Axial projection CT scan images demonstrate 3 luminal large bowel openings in the pelvis, indicated by the arrows. Of note, no oral contrast was administered during this examination. High-density material within the bowel is thought to be due to proteinaceous fecal matter

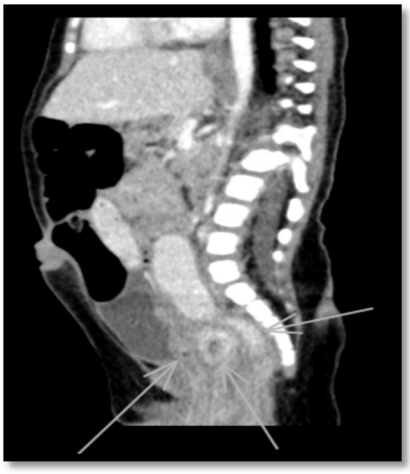


Fig 3. Sagittal projection imaging of the contrasted CT scan demonstrating the suspected large colonic lumens in keeping with colonic triplication

An exploratory laparotomy was done; intraoperative findings included duplication of 25 cm of the terminal ileum, (Fig. 4) both communicating with the triplicated cecum, (Fig. 5) which drained into a triplicated colon. (Fig 6 and 7) The appendix was triplicated. (Fig 5 and 6) Figure 8 demonstrates the schematic intraoperative findings of the distal colon in our case.

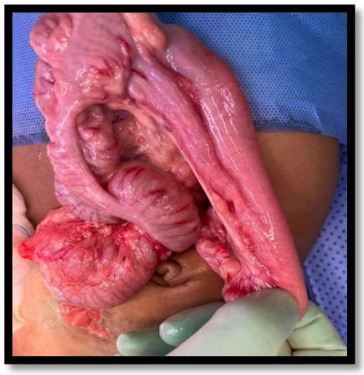


Fig 4. Duplication of Ileum

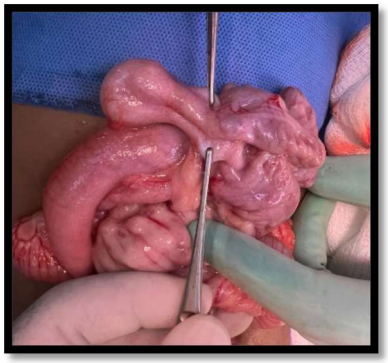
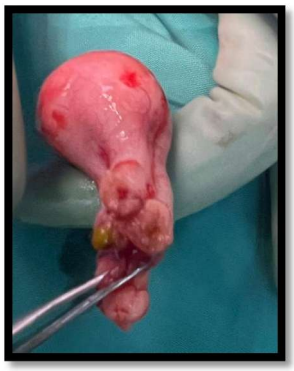


Fig 5. Triplication of appendix and cecum



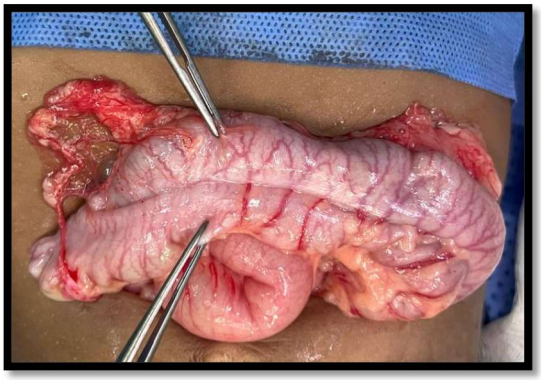


Fig 6 & 7. Triplication of the appendix post appendicectomy, triplication of the colon and reconstruction of the common channel of the rectum

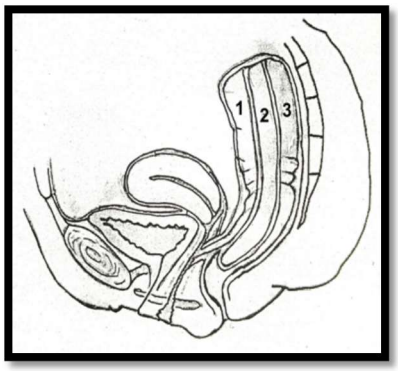


Fig 8. Lateral diagram of colonic triplication with a rectovaginal fistula (1), a blind end (2), and a patent component (3)

Furthermore, there was no evidence of bowel obstruction, and a rectovaginal fistula was confirmed. A colostomy was brought down, an appendicectomy was performed, and the colon was shaved off the vagina. The vaginal defect was repaired, the enterotomy extended, and a common rectal channel created as demonstrated in Figure 9. Lastly, we repaired the rectal wall and redid the divided colostomy with each stoma having three lumens. The schematic diagram in Figure 10 gives clear details of the image in Figure 9.

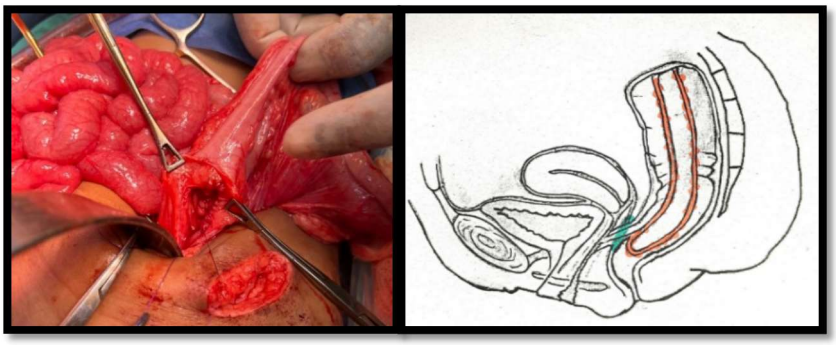


Fig 9. Reconstruction of the common channel of the rectum Fig 10. Lateral diagram highlighting the ligated rectovaginal fistula (green) and resection of the intraluminal walls (red)

Postoperatively, the patient was transferred to the PICU, where she developed hospital-acquired pneumonia. Unfortunately, she required escalation of ventilation to high-frequency oscillator ventilation. Nonetheless, she made a wonderful recovery.

Once she fully recovered our patient was discharged home with a divided colostomy. Her contrast enema three months post-operatively showed a patent rectal channel without a stricture or recurrence of the rectovaginal fistula. (Fig 11) She has had no complications thus far, is growing well, and her stoma is functioning well. She is planned for closure of the colostomy in the future, with long-term follow-up in our department.

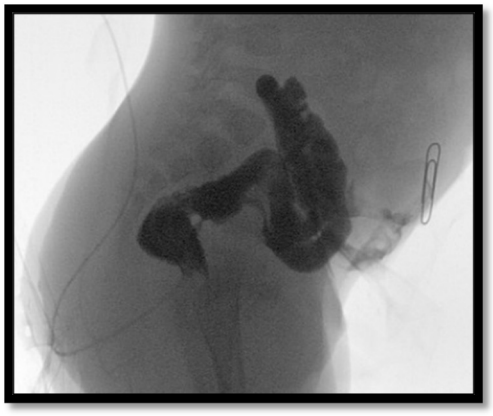


Fig 11. Post-operative Contrast enema – single rectal channel demonstrated

DISCUSSION

Histologically, smooth muscle and gastrointestinal epithelium are well-developed in GIT triplications, even though the etiology is unknown. [1,5] Postulated theories on duplication cysts may also explain how triplication cysts occur. [6] The split notochord hypothesis states that embryonic notochord doubling causes cysts. Thus, it is possible that gastrointestinal tract and urogenital tract duplications happen during the solid phase of embryonic development because of luminal recanalization and flawed or partial twinning. [5,6] These theories on duplication cysts may also explain how the triplication cysts occur. Dealing with patients with colonic triplication cysts is challenging. There are no specific clinical features or association with specific syndromes, but it is important to evaluate patients for possible associated lesions. [7,8] Currently, triplication cysts pose a diagnostic and treatment challenge.

Most enteric duplication cysts (EDC) are diagnosed with ultrasonography (U/S), which provides distinct features such as the "double wall" sign, which is made up of a hyperechoic view representing mucosa and hypoechoic muscle. [9] In other cases, even though U/S may detect the cystic mass, it may not be easy to determine the organ of origin, as was found in a case reported by Chatta et al. [8] AE Joda et al implied that U/S seems to have good

diagnostic efficacy in cystic type of duplication as opposed to tubular duplication. [6] Computed tomography (CT) and Magnetic resonance imaging (MRI) scans are also useful in difficult circumstances or when precise body component images are needed for surgical planning. By using high-resolution and three-dimensional contrast imaging, McKenna et al. found it helpful when planning surgery for a newborn with intestinal triplication and

associated congenital anomalies. [4] It is important to rule out associated abnormalities of the genitourinary system, such as vaginal atresia, prior to reconstructive surgery, as the excess triplication cysts may be used for vaginal reconstruction. [1-4]

Colonic triplication cysts bear a complex anatomy that leads to a challenging management strategy. [1] Table 1 summarizes cases that have been reported in pediatric literature. (Table 1) Surgical management is not standardized due to the rarity of this condition, and it differs from case to case depending on clinical presentation and associated anomalies. Some cases may need no intervention. [6,10] It becomes more challenging when there is total colonic triplication; some surgeons believe surgery should be done to prevent complications, while other surgeons suggest intervention should be reserved for symptomatic patients. [1,4] In our case, indications for surgical intervention were a mass protrusion from the anus, rectovaginal fistula, and suspected anal atresia, hence a divided colostomy.

Nonetheless, surgical intervention is essential to prevent bowel obstruction, GIT bleeding, and infections. The surgical procedure will depend on the side and length of the triplication cyst. Nebot et al. recommend meticulous surgical planning to preserve as much normal gut as possible. [5] Other pediatric surgeons found it important that a multidisciplinary approach be implemented in the management of these children for the best outcome. [2,11]

The surgical management principles are extrapolated from the management of enteric duplication and should be tailored per case. [1,10] The main surgical goal in this condition is to create a single lumen for defecation. [4,11] This can be achieved in one sitting or by following a multi-staged procedure. [1] We followed a three-stage procedure for our patient. First a divided colostomy, then we created a single rectal channel and performed an appendicectomy, and the last stage of the procedure was closure of the colostomy. Other surgical options include enucleation of an enteric cyst without a bowel resection (resection of non-communicating cyst), enterostomy, then GIT reconstruction with or without bowel resection once anatomy has been delineated. Other surgeons suggest that only symptomatic duplications should undergo surgical intervention. [3,12] Long-term follow-up is necessary to identify complications and ensure that growth and development are not

negatively impacted. Some patients may have complications of stool or urinary incontinence later in life. [4]

Year	Author	Age/sex	Symptoms	Location	Associated anomaly	Management
1985	J Milsom et al	2.5yrs/ F	Abdominal mass	Esophagus	Gastric duplication	Resection
1990	Klumpp et al	3yrs/ M	Per-rectal bleeding	Ileum	None	Resection of the entire involved intestine
1997	Luis De la Torre et al	6m/ M	Abdominal mass	Stomach	Peritoneal melanosis	Resection of gastric cystic mass
2006	YK Sarin et al	8m/ M	Meconium-stained urine	Colon	Bifid glans penis, rectovesical fistula H-type, duplicated bladder, umbilical hernia	Colostomy, Resection, Anastomosis
2006	Heloise Gisquet et al	0d/ M	Imperforate anus	Transverse colon	Facial dysmorphism, Anorectal malformation	Colostomy, Radical resection of triplication
2020	M Gaffley et al	0d/ M	Imperforate anus, perineal fistula	Total colon	Anorectal malformation - perineal fistula	Colostomy, demised
2021	AE Joda et al	3yr/ F	Acute abdomen – perforation	Ileum	None	Resection, Anastomosis
2022	McKenna et al	0d/ M	Imperforate anus	Colon	Anorectal malformation - fistula distal to bladder neck	Colostomy, PSARP, Resection, Anastomosis
2023	A Chattha et al	1.5yr/ F	Abdominal distension	Stomach	Peritoneal melanosis	Partial excision, Mucosal stripping
2024	E Rasouli et al	2d/ F	Protrusion from vestibule, failure to pass meconium, despite normal anus, vestibule protrusion opened and drained meconium	Total colon	Anorectal malformation - rectovestibular fistula	Colostomy, Resection, Anastomosis
2024	Z Zain et al	9m/ M	None	Sigmoid colon	Anorectal malformation - prostatic fistula	Stoma, PSARP, Resection, Anastomosis

Table 1. Summary of published case reports of enteric triplication cysts

CONCLUSION

Colonic triplications are less prevalent . Clinical presentation is very nonspecific, and many cases are diagnosed intraoperatively. Surgical intervention should be aimed at creating a single rectal channel without fistula to the genitourinary system. Multidisciplinary team approach is best to treat these unique and rare cases.

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