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Colonic duplication with recto-urethral fistula: Elusive diagnosis and successful treatment in a resource-limited setting☆

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ABSTRACT

A 3-year-old Ugandan boy presented with 18 months of constipation and 12 months of pneumaturia and fecaluria. Physical exam revealed abdominal distension and a palpable mass anterior to the rectum. Previous contrast enema had been non-diagnostic, and a voiding cystourethrogram (VCUG) had confirmed a recto-urethral fistula. After surgical evaluation, a computed tomography (CT) scan suggested colonic duplication, and a laparotomy was performed for rapidly progressive bowel obstruction. A tubular colonic duplication with fecal impaction was found, necessitating fecal disimpaction and a double divided colostomy. Two months later at re-laparotomy, the septum between the duplicated colonic lumens was found to extend proximally to the mid transverse colon and distally to the upper rectum, and was divided. No urethral fistula was identified, and the colostomy was recreated. At a third operation, the colostomy was reversed. The patient is well at one-year follow up, without evidence of recurrent fistula or stricture. This case shows that colonic duplication in children can be an elusive diagnosis. Often, a variety of radiographic studies may be needed and may be difficult to interpret. In cases with colo-urinary fistula, the fistula may respond to fecal diversion without requiring operative repair. Management in a resource-limited setting can still yield positive outcome.

Duplication of the colon is rare with a variable and non-specific clinical presentation, often creating a diagnostic and therapeutic challenge. This case illustrates these features in a resource-limited setting and demonstrates successful treatment in three surgical stages.

1. Case report

A 3-year-old Ugandan boy presented with 18 months of constipation and 12 months of pneumaturia and faecaluria. He was well until reaching a year of age when he developed intermittent constipation after starting solid foods in addition to breastfeeding. He had about 2–3 painful, hard, pellet-like bowel movements per week. There was no melena, but he did have reducible rectal prolapse on straining. His constipation and rectal prolapse were managed in primary care with laxatives and recurrent enemas but symptoms were refractory.

At 2 years of age, his mother noted that he passed gas and stool per urethra. He was treated for recurrent urinary tract infections and had abdominal distension but no vomiting or diarrhea. He had weight loss but no fevers or anorexia.

The mother's pregnancy was uneventful and she had a term delivery by caesarean section due to polyhydramnios with a birth weight of 2.8 kg. Meconium was passed within 24 h of delivery and no gross anatomical anomalies were noted. He grew well with timely achievement of developmental milestones.

At his initial presentation to a general doctor, plain abdominal films suggested constipation without evidence of obstruction. An abdominal ultrasound showed no masses or ascites. A barium enema showed fecal loading in the colon and rectum but was otherwise non-diagnostic. He was referred for surgical evaluation at that point.

On our initial examination, he had moderate abdominal distension with a palpable non-tender mass in the right lower quadrant. He had normal appearing genitalia with bilateral descended testicles. He had a single anal opening with no fissures or anal tags. On rectal exam, there was no anal stenosis and the

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mucosa was smooth but there was a firm non-tender mass anterior to the rectum and the superior extent of this mass was not palpable.

A repeat contrast enema done soon thereafter was difficult to interpret due to retained contrast from the initial examination (Fig. 1). A VCUG suggested a fistula to the rectum at the approximate level of the prostatic urethra with an associated stricture (Fig. 2). An abdominal CT scan was subsequently obtained which showed fecal impaction without any visible extraluminal mass. On the CT scan, there was retained contrast from the enema studies, and the appearance of a possible septum of the sigmoid colon to suggest colonic duplication (Fig. 3).

Laboratory studies (complete blood count, electrolytes, liver function tests) were within normal limits.

The patient was then admitted with acute abdominal distension, nausea, vomiting, and failure to pass stool. At emergency laparotomy, grossly dilated bowel was found with a tubular colonic duplication appearing to extend from the hepatic flexure to the rectum. The duplicated colons shared a mesenteric blood supply (Fig. 4). A double divided sigmoid colostomy was performed to decompress the bowel and to divert stool from the urinary tract (Fig. 5).

The patient recovered well and his urinary symptoms subsided completely. A repeat enema was obtained retrograde simultaneously through the two lumens of the end colostomy and showed a septum extending toward the proximal transverse colon. Reassessing the child's previous CT scan, it was apparent that this was the visible septum on the CT scan, with impacted stool in a "false lumen" (or duplicated colon) ending in the upper rectum (Fig. 3). In addition, the second enema study that had been performed also appeared to delineate air and contrast in the non-opacified duplicated colon adjacent to the "true" colon (Fig. 1).

Two months after the child's first operation, a laparotomy was performed and the colostomy and mucous fistula were taken down. Our pre-operative plan was to search for a fistula to the urinary tract, and to attempt division through a posterior sagittal approach as for congenital recto-urethral fistulae associated with anorectal malformations.

On the distal side, the septum was divided with a donated stapling device. By looking into the distal colon, we could confirm that the septum (and "false lumen") ended in the upper rectum. There was no visible opening to the urinary tract or visible or palpable inflammation to suggest a colo-urinary fistula. Methylene blue was injected retrograde through a urethral catheter and did not demonstrate a fistula. Cystoscopy and fluoroscopy were not available and could not be utilized. On the proximal side, the septum was divided with electrocautery. This was done by mobilizing the splenic flexure and serially intussuscepting the colon on itself. The

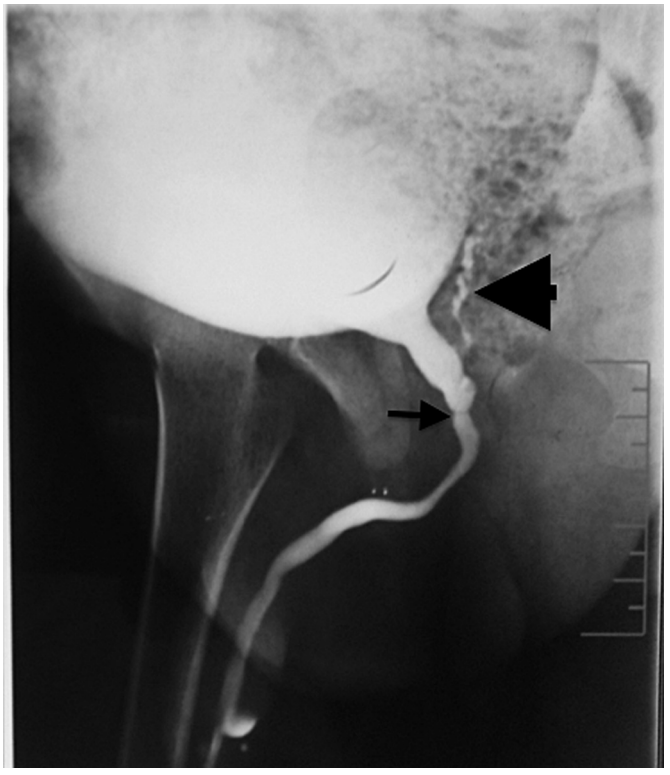

Fig. 2. VCUG showing a colo-urethral fistula with a stricture of prostatic (approximate location) urethra (small black arrow) with contrast pooling in colon (large black arrow) confirming colo-urethral fistula.

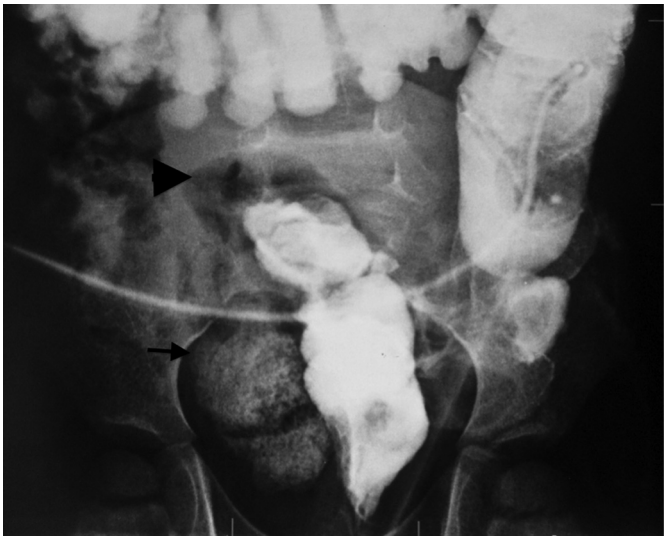

Fig. 1. Second contrast enema, this one obtained after surgical referral, which was initially difficult to interpret. After CT scan and surgery, dual colonic lumen was appreciated, with air column visible (large black arrow) and retained contrast from first enema in "false lumen" (small black arrow) with residual contrast from first enema.

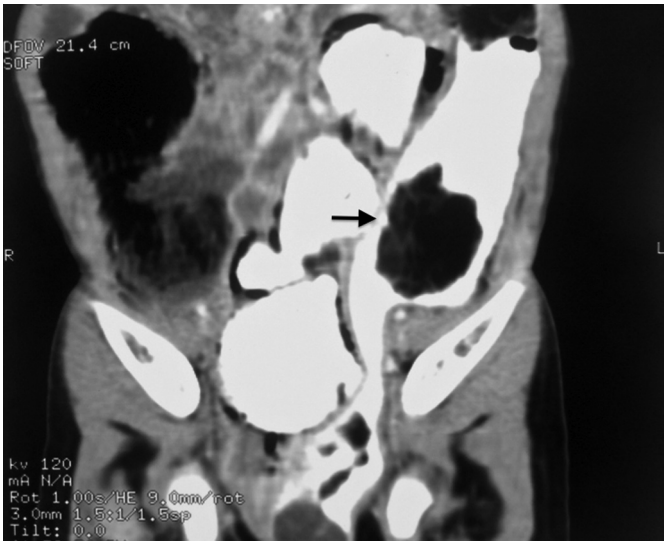

Fig. 3. CT scan (coronal image) showing high degree of fecal loading and possible septum (black arrow) suggesting colonic duplication with stool and contrast in both lumens.

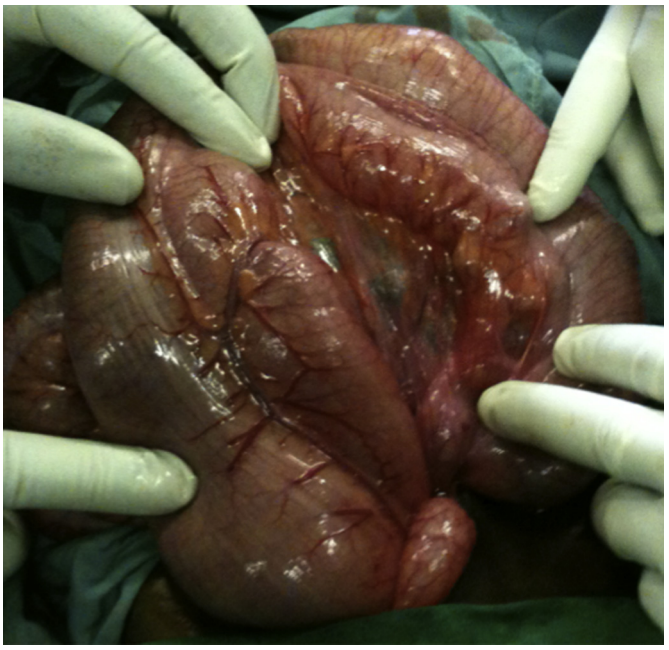

Fig. 4. Duplicated colon (sigmoid) discovered at laparotomy, sharing a common blood supply.

septum extended to the mid transverse colon, where it ended. The colon appeared mildly dilated but viable and no resection was performed. The sigmoid colostomy was then re-created at this time to allow any residual inflammation at the previous urethral fistula site to resolve.

In the early post-operative period, the child had urinary retention managed with re-catheterization and antibiotics. After the catheter was removed, he had several episodes of urinary retention requiring re-catheterization. We attributed this to inflammation and edema at the prior fistula site. His symptoms improved over the next eight weeks and a repeat VCUG ten weeks after the second operation showed a mild stricture with no visible fistula. We surmised that there was no more communication with bowel lumen and decided to close the colostomy. The colostomy was subsequently closed and the child has recovered well after a year of follow up.

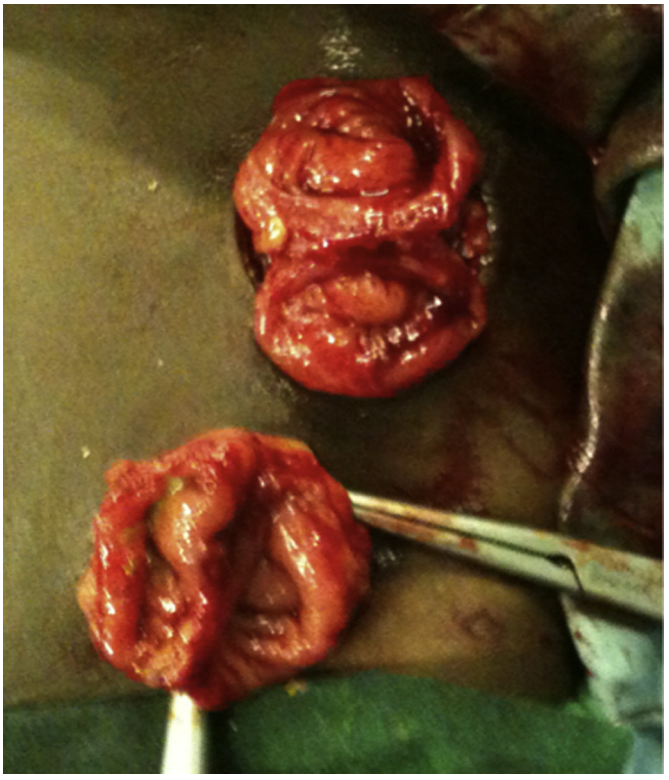

Fig. 5. Double divided colostomy with two lumens visible in each barrel.

2. Discussion

This case once again illustrates that pediatric health care providers and surgeons should consider congenital causes of constipation. In addition to anorectal malformations and Hirschsprung's disease, colonic duplications should also be considered. Colonic duplications are rare, with an incidence of approximately 1 in 4500–5000 autopsies, and they have a heterogenous etiology as reflected in existing case series and reports [1–3]. They are associated with bowel atresias and genito-urinary anomalies such as recto-urethral fistula in up to 50% cases and are classified as Type I (limited to alimentary tract; cystic or tubular) or Type II (associated duplication of genito-urinary tract with, fistula or imperforate anus). While cystic duplications predominate in the rest of the alimentary tract, tubular duplications are more common in the colon. The presentation varies with the patient's age, the location of duplication, type of mucosal lining, and the duration of disease. Patients may present with a wide variety of symptoms: intestinal obstruction, gastro-intestinal bleeding (especially secondary to ectopic gastric mucosa), perforation, abscess, neurologic symptoms, chronic constipation, rectal prolapse, pneumaturia and fecaluria, and asymptomatic masses or foreign bodies on ultrasound or plain x-rays [4–10]. Diagnosis may be more likely to be made in the newborn period when associated with a visible anomaly such as a duplicated anal opening, but even in such cases, the duplication may be difficult to identify [11–13].

We believe our case is additionally unique due to the presentation in a resource-constrained environment, which broadened our differential diagnosis and presented additional challenges. Previous work has highlighted the challenges of caring for children with colorectal disease in a resource-limited environment, though anorectal malformations and Hirschsprung's disease were discussed [14]. However, similarly in our case, there was difficulty in obtaining adequate pre-operative and intra-operative imaging studies and this challenged our surgical decision-making. In addition, like many other medical specialties in Uganda, there is a shortage of radiologists, and specifically, there is no specialty-trained pediatric radiologist. This case underscores the difficulty of performing and interpreting studies for rare congenital anomalies in the resource-limited setting. Nonetheless, even with the best available imaging, diagnosis can be difficult, as evidenced by prior case reports."

Our patient turned out to have a Type II fistula and presented with constipation and rectal prolapse as primary symptoms and later developed pneumaturia and fecaluria. Likely, there may have been a small connection present to the urinary tract at birth but this may have only become clinically significant after a large degree of stool loading. Once a surgical referral was made, the presence of a pelvic mass on examination raised concerns for malignancy. In addition, many patients in our environment present with chronic constipation and a delayed diagnosis of Hirschsprung's disease; however, this did not explain the urinary symptoms. Similarly, rectal prolapse is very common in our environment due to high incidence of infectious diarrhea and malnutrition but this patient's symptoms were not linked to loose stool; in fact, the primary features were constipation.

Plain abdominal films, abdominal ultrasound, and several enemas did not reveal the diagnosis on reviewing the studies. While a VCUG did demonstrate a fistula, the origin was unclear. In our case the diagnosis was suspected on CT scan and made at laparotomy performed urgently due to the patient's rapidly evolving obstructive symptoms. Complete assessment of the extent of duplication could not be made at the initial laparotomy due to the patient's severity of illness and fecal impaction. The urinary symptoms responded to fecal diversion and disimpaction. As reported in other studies, in hindsight, signs of the duplication were apparent on previously obtained studies.

Tubular duplications can be treated with resection if needed. As there is a shared blood supply, the duplicated colon cannot be isolated and resected. In our case, as the duplication involved most of the colon, and due to the child's age, the septum was simply divided. There have been isolated reports of adult malignancy arising from tubular colonic duplication, but this is thought to be rare as it is not associated with ectopic gastric mucosa [15].

Prior to the second operation, we did not know the proximal or distal extent of the duplication despite attempts to determine this radiographically. We assumed that the urinary fistula marked the distal end of the duplication, which turned out to be the case (upper rectum). Proximally, we had planned to intussuscept the bowel on itself as far as possible, then perform additional colotomies as necessary divide the septum as extensively as possible, if necessary, all the way to the cecum. If the rectum had been involved, we would have divided the septum from the abdominal side as distally as possible, then transanally proceeding proximally. While numerous septal fenestrations have been described for total tubular colonic duplication, recurrent obstruction has been noted in cases where the septum was not completely divided to its most distal extent [16]. We had several donated stapler reloads to perform the distal portion and completed the remaining portion with diathermy and scissors.

Up to 50% of duplications are associated with genito-urinary malformations, most commonly entero-urinary fistulae, as in our case. Our patient had recto-urethral fistula at the approximate level of the prostatic urethra with a stricture in the membrano-prostatic area. Only two other such cases, one very recently, have been reported [17,18]. This most recent case is also the only other reported case where the fistula has been able to be identified on enema or VCUG. In that case, fistulae to the duplicated colon were identified intraoperatively at cystoscopy and separation performed through posterior sagittal approach. In our case, cystoscopy and fluoroscopy were not available and we were unable to identify any fistula through intra-operative methylene blue instillation and visual inspection and probing. The patient had initial intermittent urinary obstructive symptoms post-operatively that cleared over several months and is now symptom-free a year after colostomy closure. This suggests that some patients with urinary fistula may respond fecal diversion.

3. Conclusion

Pediatric surgeons must consider colonic duplications, though rare, in the differential for chronic constipation in children. The diagnosis of colonic duplication is elusive due to the breadth of possible presenting symptoms and the associated differential diagnoses. Multiple imaging modalities are often required, as reported previously. While we had the advantage of working in a tertiary referral center in a low-income country with a fairly wide possibility of diagnostic options (ultrasound, contrast studies, CT scan), there are no specialty-trained pediatric radiologists and intra-operative diagnostic studies are limited (no cystoscopy and fluoroscopy). The fistula responded to fecal diversion, without symptoms suggestive of stricture, suggesting all fistulae may not require operative treatment. As stressed in prior reports, the septum can be divided without requiring a colonic resection. Outcomes can be good in a limited-resource setting.

Conflict of interest

The authors have no conflict of interest to disclose.

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