

Case report

Atypical presentations of alimentary tract duplications in children: two case reports

Nimmita Srisan^a, Palittiya Sintusek^{b,c}, Teerasak Phewplung^{c,d}, Paisarn Vejchapipat^{a,*}

^aDepartment of Surgery, Faculty of Medicine, Chulalongkorn University and King Chulalongkorn Memorial Hospital, Thai Red Cross Society, Bangkok, Thailand

^bDivision of Gastroenterology, Department of Pediatrics, King Chulalongkorn Memorial Hospital and Faculty of Medicine, Chulalongkorn University, Bangkok, Thailand, and Pediatric Gastroenterology and Hepatology STAR (Special Task Force for Activating Research), Department of Pediatrics, King Chulalongkorn Memorial Hospital and Faculty of Medicine, Chulalongkorn University, Bangkok, Thailand

^cDepartment of Radiology, King Chulalongkorn Memorial Hospital and Faculty of Medicine, Chulalongkorn University, Bangkok, Thailand, and Pediatric

^dGastroenterology and Hepatology STAR (Special Task Force for Activating Research), Department of Pediatrics, King Chulalongkorn Memorial Hospital and Faculty of Medicine, Chulalongkorn University, Bangkok, Thailand

Alimentary tract duplication is a rare congenital anomaly. We present herein, two cases of different types of alimentary tract duplications that have entirely different clinical presentations. The first case was a girl presented with chronic constipation was finally diagnosed with tubular duplications of the entire colon. The other was a toddler with acute intestinal obstruction diagnosed with cystic duplication at terminal ileum. These two cases remind us of the difficulty in preoperative diagnosis. High suspicion of the disease and multimodality approaches were beneficial for accurate preoperative diagnosis. Surgery was the mainstay of the treatment.

Keywords: Case report, alimentary tract duplication, children, constipation.

Alimentary tract duplication is a rare congenital anomaly with an incidence of 1: 4,000 - 5,000 live birth and could occur anywhere along the gastrointestinal tract.⁽¹⁾ The duplication was classified into 2 types, namely: cystic (80.0%) and tubular (20.0%). The former does not communicate with the adjacent lumen while the later might communicate and share the common adjacent lumen with the main alimentary tract. Ileum is the most common site (33.0%) followed by esophagus (20.0%) and colon (13.0%).⁽²⁾ Clinical presentations are varied and determined by the type, site and size of the duplications.⁽³⁻⁵⁾ The preoperative diagnosis is usually difficult to establish especially in atypical presentations,

due to the rarity of the anomaly. We herein report two cases of alimentary tract duplications in which clinical manifestations occurred beyond infant period, i.e., a girl with chronic constipation and a boy with acute intestinal obstruction.

Case presentation

Case report 1

Chief complaints

An 8-year-old girl with double inlet left ventricle post pulmonary artery banding and Blalock Hanlon procedure presented with acute abdominal pain and vomiting for 2 days during the regular visit at our hospital.

History of present illness

She had recurrent abdominal pain and chronic constipation for 4 years. The colicky pain had been worsened and she developed vomiting during the last 48 hours.

*Correspondence to: Paisarn Vejchapipat, Department of Surgery, King Chulalongkorn Memorial Hospital and Faculty of Medicine, Chulalongkorn University, Bangkok 10330, Thailand. E-mail: paisarnv@gmail.com

Received: September 28, 2020

Revised: October 30, 2020

Accepted: November 4, 2020

History of past illness

The patient had double inlet left ventricle which underwent pulmonary artery banding and Blalock Hanlon procedure.

Physical examination

The examination reveals mild abdominal distension, normo-active bowel sound with palpable, movable and soft-consistency mass size 6 x 8 cm² at the lower mid abdomen. Per rectal examination reveals empty rectum and minimal non-bursting soft yellowish feces without glove-like sensation implying that neither fecal impaction from functional constipation nor Hirschsprung's disease was likely.

Imaging examinations

Abdominal radiography showed the localized dilatation of the descending colon with fecal impaction. But there was no proximal colonic nor small bowel dilatation. (Figure 1A). Disimpaction with polyethylene glycol and rectal enema initially failed but was finally successful by high rectal irrigation with glycerin-containing saline. Barium enema was done and revealed two colonic lumina from the ascending colon down to the distal descending colon, separated by the radiolucent line (white arrows) (Figure 1B). There was poor emptying function of the dilated part of descending colon and fairly good emptying function of the rectosigmoid colon after evacuation. The colonic transit study was performed with prolonged segmental and total colonic transit time (58 hours) (Figure 1C). To identify problematic segment, colonic manometry was studied. A colonoscopy with colonic manometry catheter insertion was performed. Colonoscopy showed two opening lumens of the colon at 38 cm from anal verge likely true diverticulum (Figure 1D). Colonic manometry study showed the normal colonic function with frequent high amplitude propagation contractions (HAPCs) after bisacodyl stimulation (Figure 1E). Further imaging with abdominal and pelvic computed tomography (CT) to evaluate this anatomical abnormality confirmed two laminae of the descending colon which one was dilated whereas the other was not. Moreover, there were two appendices at the right lower abdomen (Figure 1F).

Final diagnosis

The final diagnosis of this case was tubular duplication of nearly the entire colon.

Treatment

Exploratory laparotomy was performed to find a tubular duplication from descending colon to terminal ileum (Figure 1G). The descending colon was dilated and the involved ileum was 34 cm from ileocecal valve with normal caliber. There were two appendices and two Meckel diverticula. Left hemicolectomy was performed followed by dividing common wall of transverse colon with surgical staplers. After spatulating the transverse colon's distal end, primary end to end anastomosis with native sigmoid colon was done.

Outcome and follow-up

The postoperative course was uneventful. The patient started enteral feeding on postoperative day 7 and was discharged on day 10. At 18-months post-operation, the patient was very well without the symptoms of recurrent abdominal pain and constipation anymore.

Case report 2

Chief complaints

A previously healthy 13-month-old boy was referred from a private hospital due to suspected small intestinal obstruction.

History of present illness

He was a full-term baby of an uncomplicated pregnancy and well-grown since birth.

History of past illness

He was hospitalized due to 4 times of non-bilious vomiting and poor intake. During admission his abdomen became distended and he developed bilious vomiting. There was no history of constipation, diarrhea, or bloody stool.

Physical examination

On examination, the child looked sick without a fever. His abdomen was soft and distended. Bowel sounds were present and hyperactive. Per rectal examination showed yellowish stool without blood.

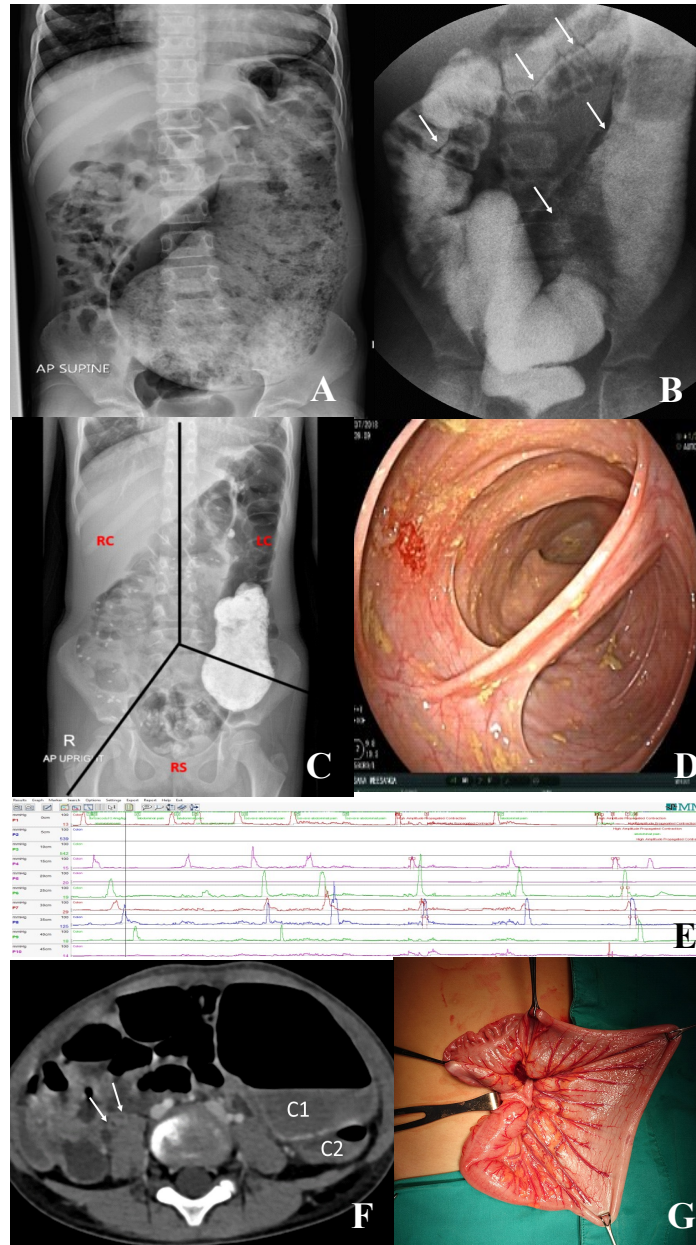


Figure 1. An 8-year-old girl with history of recurrent abdominal pain and chronic constipation for 4 years. **(A)** Anteroposterior supine abdominal radiography showed the localized dilatation of the descending colon with fecal impaction. But there was no proximal colonic nor small bowel dilatation. **(B)** Single contrast barium enema revealed two colonic lumina from the ascending colon down to the distal descending colon, separating by the radiolucent line (white arrows). **(C)** The distribution of sitz marks after 3-day ingestion. The segmental colonic transit time of right, left, rectosigmoid colon were calculated to 30, 16, 12 and 58 hours, respectively. **(D)** Colonoscopic view demonstrated the two colonic lumens of native colon and the true colonic diverticulum that shared the one-side colonic wall with the native colon. **(E)** Colonic manometry tracing showed high amplitude propagation contractions after bisacodyl stimulation (0.2 mg/kg/dose). **(F)** Axial post contrast enhanced CT of lower abdomen demonstrated two lumina of the descending colon, one was dilated (C1) and another was not dilated (C2). Moreover, there were two appendices at the right lower abdomen (white arrows). **(G)** Tubular duplication from descending colon to terminal ileum.

Imaging examinations

Abdominal radiograph revealed gasless abdomen pattern. There are a few abnormal small bowel loops were dilated at the mid and left upper abdomen (white arrows) (Figure 2A). Ultrasound abdomen showed a cystic lesion with the gut signature wall in mid-abdomen (Figure 2B), measured about 4.7 x 4.3 cm². The later abdominal and pelvic CT demonstrated the cystic lesion in mid-abdomen (Figure 2C) which compressed on the adjacent small bowel loop and caused abnormal dilatation of the multiple proximal small bowel loops with collapsed terminal ileum, entire colon, and rectum (Figure 2C).

Final diagnosis

Distal ileal obstruction due to cystic lesion was diagnosed in this patient.

Treatment

Emergent exploratory laparotomy was performed and revealed cystic duplication in the mesentery of the terminal ileum (10 cm from ileocecal valve). The duplication cyst was about 4 cm in diameter compressing the adjacent ileal lumen (Figure 2D). The residual bowel was in good condition. Segmental ileal resection with primary end to end anastomosis was done.

Outcome and follow-up

The patient improved and started enteral feeding on postoperative day 4. He was discharged on postoperative day 6. At 13-month follow-up, the patient was well and active.

Discussion

Alimentary tract duplications are rare congenital abnormalities that could occur in any area from the mouth to the anus. The pathogenesis of enteric duplication is unclear. Several theories propose the arising of the duplications. However, no single hypothesis can explain all sites and types of the anomaly.^(2, 6) Duplications must be composed of 3 components, including continuity or adherence to a part of the alimentary tract, having a smooth muscle wall, and containing a mucosal lining of one or more cell types found in the alimentary canal.⁽²⁾

Our first case was a girl who presented with recurrent abdominal pain and chronic constipation that finally diagnosed long tubular colonic duplication. Duplication of the colon is extremely rare and first described in 1876.⁽⁷⁾ The studies that indicated the

high association of tubular colonic duplication with other abnormalities especially lower genital, urinary tracts, and lumbosacral vertebra. To the best of our knowledge, only 2 previous cases of the tubular colonic duplications associated with congenital heart diseases were reported. One was a girl with Tetralogy of Fallot, agenesis of the left kidney and ureter, and S-1 spina bifida occulta.⁽⁸⁾ Another was a male infant with extensive vertebral defects, anal atresia, cardiac defects, tracheo-esophageal fistula, renal anomalies, and limb abnormalities.(VACTERL)-associated anomalies, namely, hemi-vertebrae, anal atresia with a recto-urethral fistula, hypospadias and common AV canal defect.⁽⁹⁾ Unlike the present case, these two cases had entire colon duplication but they also had other obvious abnormalities as clues for the early investigations. As functional constipation and fecal impaction might be explained, the chief complaint of recurrent abdominal pain in the general population. Per rectal examination that showed "empty rectum" in the present case could not explain the functional cause of fecal impaction, hence mechanical obstruction from other causes should be sought out. However, it is difficult to diagnose colonic duplication before surgery, and misdiagnosis is common.^(10, 11) Despite of imaging studies, some duplication cysts are diagnosed intraoperatively (38.0%).⁽¹²⁾ Barium enema could be challenging to confirm the diverticulum if two colons' opacification is overlap or in the same plane. Like our present case, the two colonic lumina from the ascending colon down to the distal descending colon, separating by the radiolucent line from barium enema study was retrospectively identified by an experienced pediatric radiologist later. Yousefzadeh DK, *et al.* suggested using contrast media of two different densities during simultaneous evaluation of both colons in case suspected of colonic duplication.^(7, 9) A colonoscopy might be valuable if there is the communication between the anomalous, and actual enteric lumens and CT whole abdomen is considered the most useful for the diagnosis preoperatively.⁽¹³⁾ In the present case, the enlarged descending colon without any obvious mechanical obstruction from barium enema and the prolonged colonic transit time from colonic transit study misleading us to investigate more for colonic motility, and we accidentally diagnosed colonic diverticulum from colonoscopy. Again, to the best of our knowledge, this is the first case of an entire colonic diverticulum who received colonic physiologic study. The result showed normality of the entire colonic function, even

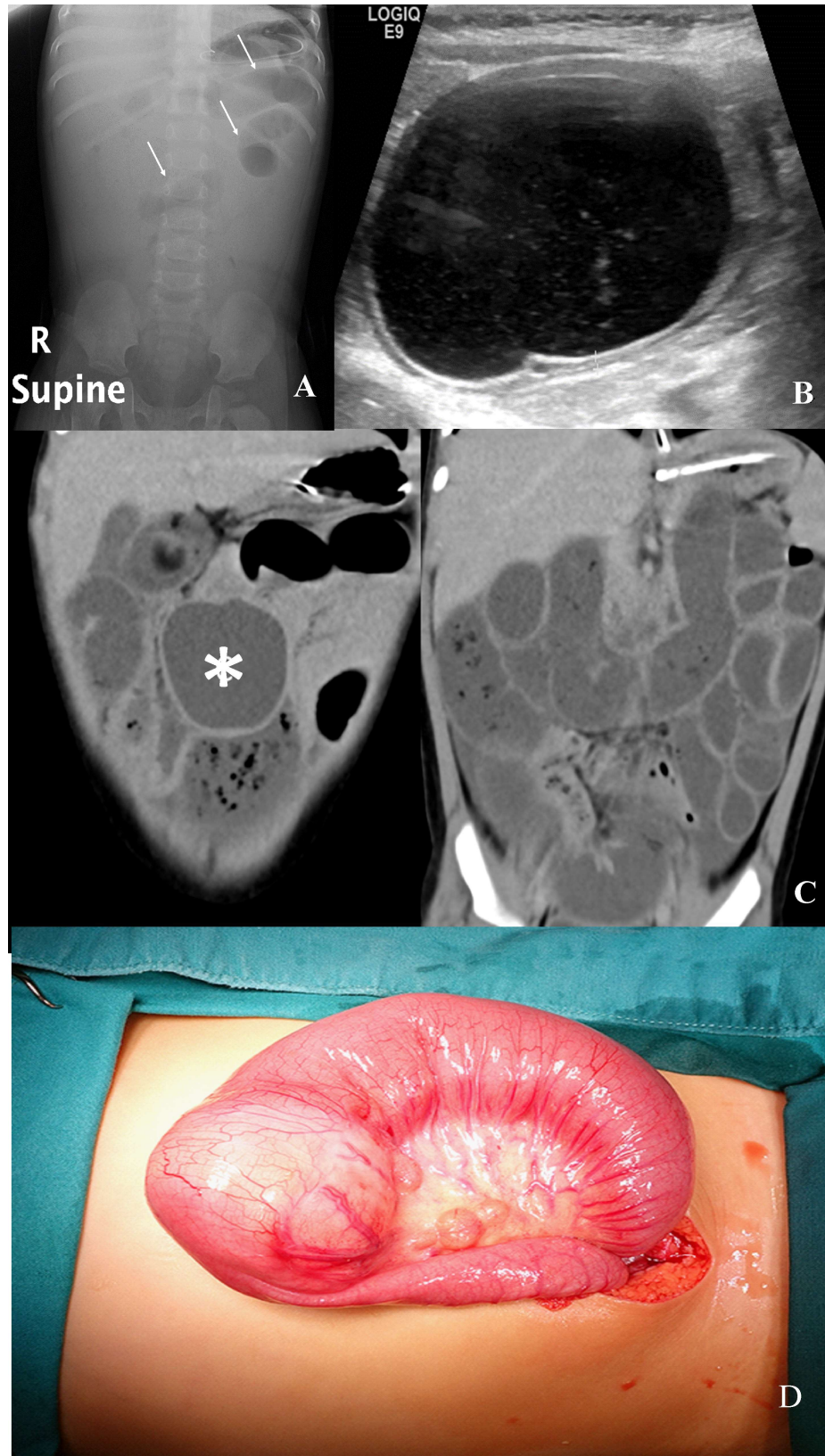


Figure 2. A previously healthy 13-month-old boy with suspected small intestinal obstruction. **(A)** Anteroposterior supine abdominal radiography revealed gasless abdomen pattern with a few abnormal small bowel loops dilatation at the mid and left upper abdomen (white arrows). **(B)** Abdominal ultrasonography showed a cystic lesion with the gut signature wall lesion in mid-abdomen. **(C)** Two coronal images of post contrast enhanced abdominal and pelvic CT demonstrated the cystic lesion in mid abdomen (*) with abnormal dilatation of the multiple proximal small bowel loops and collapsed terminal ileum down to rectum. **(D)** Cystic duplication compressed adjacent ileal lumen.

long standing chronic obstruction that implied the intact enteric nervous system and intestinal muscle in patients with chronic intractable constipation or obstruction. In aspect of surgical treatment, to preserve the normal colon and intestine, left hemicolectomy with dividing common wall at transverse colon and primary end-to-end anastomosis to native distal sigmoid colon were fashioned. As a result, there was a residual duplicated descending, transverse colon and small intestine which possibly contains ectopic gastric mucosa, especially at 2 Meckel's diverticula. Although the best option of treatment is the complete surgical removal of the duplicated part. Other considerations might be concerned, especially in this very young child with the long tubular duplication that involved the whole colon and terminal ileum. Complete surgical resection of the whole lesion as in one step might affect her nutrition status and the final long-term growth. Hence, we gave parental information of concerning symptoms, especially lower gastrointestinal bleeding and cancer surveillance as some evidence of a higher risk of adenocarcinoma of the residual duplication.^(14, 15) Furthermore, long-term follow-up and careful monitoring are needed.

The second case, extrinsic compression by cystic duplication located at the terminal ileum resulted in intestinal obstruction. The diagnosis was missed due to a low index of suspicion. After a retrospective review of the ultrasonography with an experienced pediatric radiologist, the picture demonstrated the double layered wall, the so-called "gut signature"^(2, 6, 16) This finding consistent with located adjacent to the bowel wall should prompt the suspicious of duplication cyst. CT typically is not required to evaluate the duplication cyst. As the diagnosis could not be determined, CT could help us exclude some common causes of obstruction, including intussusception, ruptured appendicitis, and Meckel's diverticulum with complications. CT may depict the location, extent of the cyst, and complications.⁽²⁾ Enteric duplications can present with gastrointestinal bleeding if the lesions contain gastric mucosa.⁽¹⁷⁻¹⁹⁾ Some patients are asymptomatic but the incidental finding was revealed during obtaining intra-abdominal workup for other purposes.⁽⁶⁾ Since the symptoms can overlap with other gastrointestinal abnormalities, it makes the diagnosis more difficult.

Conclusion

Alimentary tract duplications are rare and could present asymptomatic or with diverse manifestations. The high index of suspicion is needed, especially the clinical signs and symptoms that could not be served other more common diseases. Imaging studies are suggested as contrast study and CT can raise a clue for tubular duplications as well as ultrasound can help diagnose cystic duplications. A colonoscopy might be helpful, only the duplication with opening orifice to the main alimentary tract. Although the definite diagnosis could be made intraoperatively, clues from those investigations could help surgeons for the well-planned operation. Options of surgical procedure depend on the locations, types and involved structures. If possible, complete surgical resection is the treatment of choice.

References

1. Patino Mayer J, Bettolli M. Alimentary tract duplications in newborns and children: diagnostic aspects and the role of laparoscopic treatment. *World J Gastroenterol* 2014; 20:14263-71.
2. Sanguesa Nebot C, Llorens Salvador R, Carazo Palacios E, Pico Aliaga S, Ibanez Pradas V. Enteric duplication cysts in children: varied presentations, varied imaging findings. *Insights Imaging* 2018;9:1097-106.
3. Erginel B, Soysal FG, Ozbey H, Keskin E, Celik A, Karadag A, et al. Enteric Duplication Cysts in Children: A Single-Institution Series with Forty Patients in Twenty-Six Years. *World J Surg* 2017;41:620-4.
4. Herman AM, Msuya D, Kaino M, Shilaniman H, Magoma J, Issangya C, et al. Tubular Duplication of the Midgut Presenting with Acute Abdomen and Hematochezia: A Case Report from Northern Tanzania. *Case Rep Surg* 2018; 2018:2858723.
5. Rasool N, Safdar CA, Ahmad A, Kanwal S. Enteric duplication in children: clinical presentation and outcome. *Singapore Med J* 2013;54:343-6.
6. Jezioreczak PM, Warner BW. Enteric Duplication. *Clin Colon Rectal Surg* 2018;31: 127-31.
7. Yousefzadeh DK, Bickers GH, Jackson JH, Jr., Benton C. Tubular colonic duplication—review of 1876-1981 literature. *Pediatr Radiol* 1983;13:65-71.
8. Soper RT. Tubular duplication of the colon and distal ileum: case report and discussion. *Surgery* 1968;63: 998-1004.
9. Blickman JG, Rieu PH, Buonomo C, Hoogeveen YL, Boetes C. Colonic duplications: clinical presentation and radiologic features of five cases. *Eur J Radiol* 2006;

- 59:14-9.
10. Puligandla PS, Nguyen LT, St-Vil D, Flageole H, Bensoussan AL, Nguyen VH, et al. Gastrointestinal duplications. *J Pediatr Surg* 2003;38:740-4.
 11. Mourra N, Chafai N, Bessoud B, Reveri V, Werbrouck A, Tiret E. Colorectal duplication in adults: report of seven cases and review of the literature. *J Clin Pathol* 2010;63:1080-3.
 12. Wu X, Xu X, Zheng C, Li B. Tubular colonic duplication in an adult: case report and brief literature review. *J Int Med Res* 2018; 46: 2970-5.
 13. Al-Jarroof AH, Al-Zayer F, Meshikhes AW. A case of sigmoid colon duplication in an adult woman. *BMJ Case Rep* 2014;2014.
 14. Heiberg ML, Marshall KG, Himal HS. Carcinoma arising in a duplicated colon. Case report and review of literature. *Br J Surg* 1973;60:981-2.
 15. Kusunoki N, Shimada Y, Fukumoto S, Iwatani Y, Ohshima T, Arahi E, et al. Adenocarcinoma arising in a tubular duplication of the jejunum. *J Gastroenterol* 2003;38:781-5.
 16. Di Serafino M, Mercogliano C, Vallone G. Ultrasound evaluation of the enteric duplication cyst: the gut signature. *J Ultrasound* 2016;19:131-3.
 17. Bhattacharya A, Samujh R, Rao KL, Mittal BR. Long segment jejuno-ileal duplication cyst with ectopic gastric mucosa detected on 99mTc-pertechnetate scintigraphy. *Indian J Nucl Med* 2013;28:96-8.
 18. Jia HM, Zhang KR, Qu RB. Ileal duplication with extensive gastric heterotopia in a girl. *World J Pediatr* 2009;5:322-4.
 19. Stern LE, Warner BW. Gastrointestinal duplications. *Semin Pediatr Surg* 2000;9:135-40.