GIANT DOUBLE ILEAL DUPLICATION CYST - A CASE REPORT -

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Abstract

Alimentary tract duplication cysts are malformations that occur during the embryologic development of the organs that form the digestive system. These malformations can appear at any level of the alimentary tract. The incidence at which these malformations occur is estimated at 1 every 4500 newborns. We present a case of a new-born suffering from intestinal duplication, with two ileal duplication cysts, a giant tubular shaped one and a spherical one. Although the vast majority of intestinal duplications develop in the small intestine, they are solitary and small. Multiple intestinal duplications in the same patient are a rarity, as well as the giant ones.

Keywords: giant intestinal duplication, echography, alimentary tract malformation

Introduction

Alimentary tract duplication cysts are malformations that occur during the embryologic development of the organs that form the digestive system. These malformations can appear at any level of the tract, beginning from the oropharynx and leading towards the anus (1, 2).

Calder in 1733 first mention this condition, but the modern term of alimentary tract duplication cysts has been introduced by Dr. William E. Ladd in 1937, in the attempt to better organize the individual pathologies that affect the alimentary tract system (3).

The incidence at which these malformations occur is estimated at 1 every 4500 newborns[2]. It has been observed that 80% of the tumors can be found in the abdominal cavity, 20% can be found in the thorax, neck and head regions (Fig.1). Within the group found in the abdomen, 75-80% are formed along the length of the jejunum and ileum, always on the mesenteric side (4).

From a morphological point of view, we cam distinguish two types of tumors, cystic and tubular. The cystic ones are rather round shaped and have no lumenal connections with the intestines, whereas the tubular one do communicate with their respective segment. Histologically the insides of these tumors can present heterotopic gastric mucosa that has acidic secretion capacity, which can lead to complications.

Clinical manifestations depend on the size, location, intimacy with surrounding structures and whether or not the tumor presents gastric mucosa. Frequently the symptoms can falsely lead the examiner to think of an acute abdominal pain syndrome, but most of these embryological defects are detected incidentally either ante-partum or after birth. The presence of other associated malformations is very common, most of them regarding the pancreas or the vertebral column.

Pain, regurgitation and a palpable abdominal tumor, are signs and symptoms associated with the presence of these malformations. Nevertheless, this condition is an exclusion diagnosis in any physical exam, given the rarity of its occurrence. Pain is felt when the affected intestinal segment gets infected and expands. In the case that gastric mucosa is present, signs of superior digestive hemorrhage can appear, like hematemesis or melena, and haematochezia if the tumor appears in the distal part of the digestive system (5).

The treatment is exclusively surgical, and the way it is performed depends on the site duplication, blood supply, and the relationship with surrounding structures. It is considered that small cysts can be enucleated without the need to perform intestinal resection, if the tumor and the healthy intestine segment do not share a common blood supply. In most cases, enteric resection is performed with end-to-end or end-to-lateral anastomosis of the remaining segments, and the tumor is removed alongside the healthy intestinal segment which is stripped of blood supply after the operation (6).

Case presentation

We present a case of a new-born suffering from intestinal duplication, with two ileal duplication cysts, a giant tubular shaped one and a spherical one.

The patient, coming form unmonitored pregnancy, born at 38 weeks of gestation with a weight of 2200 grams, 47 centimetres in size, 32,5 centimetres of head circumference, was admitted in Neonatology Department, ICU Unit of our Hospital at the age of 9 days presenting vomiting, abdominal distension and a palpable abdominal mass.

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No significant family history was found.

Surgical consultation at admission detect a large abdominal mass from the epigastric region to the right iliac fossa. Plain abdominal radiography discloses hypo-aeration in the lower abdominal quadrants, with no airfluid levels or pneumoperitoneum, also a T6-T8 vertebral block is observed incidentally. Abdominal echography shows a tubular mass with thin but well delimited walls, with no peristalsis, low echogenicity content and a calibre of 4 centimetres. The mass expands from the liver to vicinity of urinary bladder (Fig. 2).

Medical treatment and preoperative preparation with double antibiotherapy with Meropenem and Amikacin, Etamsylate, Calcium gluconate, Pantoprazole, Metoclopramide, Dopamine, as well as parenteral nutritional support and electrolytes were established. Blood tests shows hyperbilirubinemia with mild hepatocytolysis, slightly rise of creatinine levels, hypoglycemia, low total serum protein level and monocytosis.

Transfonatanellar ultrasound detect grade II intraventricular haemorrhage.

Cardiac evaluation shows a Patent Foramen Ovale (PFO), which is normal at this age.

Thoracic, abdominal and pelvic computed tomography (CT) reveal a cystic giant formation that occupies the entire anterior abdominal cavity as follows: it has its own wall and liquid content, with debris deposits. It has a sinuous tract, it is predominantly developed in the anterior abdominopelvic compartment, exerting a displacing compressive effect on intestine and parenchymal organs. Behind the transverse colon is a similar but smaller image (3.6 cm long). T6-T8 spinal vertebral anomalies and T5 hemi-vertebra are revealed (Fig. 3).

The patient is transfered to Surgery Department for surgical treatment and after proper preparation radical treatment is applied. Surgical treatment consist of medial laparotomy that reaveal a tubular ileal duplication cyst that starts upward from about 5 cm from the ileocecal valve, in intimate contact with the mesenteric face of about 8-10 cm of the ileum to subsequently lose intimate contact with the ileum. The described formation continues through an atretic portion with another spherical duplication cyst that is not intimate with the normal intestinal segment. The block resection of the two duplication cysts is performed with the loss of approximately 10 cm of the terminal ileum that shares common blood supply with the cyst segment being in intimate contact with it. The excised piece is taken for histopathological examination. Reconstruction of the digestive tract is accomplished by end-to-end anastomosis in double layer and "cut-back" on the distal portion due to the large incongruence between the two ends of the intestinal anastomosis segments (Fig. 4). Appendectomy, peritoneal drainage and incision closure is performed, as well as CVC into the right subclavian vein.

First Author	Institution	No. D (No. Pts)	Oral	Esophagus	Thoracoabdominal	Stomach	Duodenum	Jejunum/ileum	Colon	Rectum	Other
Gross, 1952[2]	Children's, Boston	68 (67)	1	13	3	2	4	32	10	3	0
Basu, 1960[21]	A. H. Children's, Liverpool	33 (28)	0	7	0	1	3	16	4	2	0
Grosfeld, 1970[22]	Children's, Columbus	23 (23)	0	4	2	1	0	9	7	0	0
Favara, 1971[12]	Children's, Denver	39 (37)	1	6	0	3	4	20	4	0	1
Bower, 1978[23]	Children's, Pittsburgh	78 (64)	0	15	1	6	6	34	12	2	2
Hocking, 1981[24]	RHSC, Glasgow	60 (53)		8	2	8	1	32	4	5	0
Ildstad, 1988[25]	Children's, Cincinnati	20 (17)	0	6	0	1	0	5	8	0	0
Bissler, 1988[26]	Children's, Akron	11 (11)	0	1	0	1	2	4	2	1	0
Holcomb, 1989[14]	Children's, Philadelphia	101 (96)	0	21	3	8	2	47	15	5	0
Pinter, 1992[27]	Hungary	30 (28)		6	2	4	3	9	3	3	0
Bajpai, 1994[28]	IIMS, New Delhi, India	15 (14)	0	8	1	0	1	1	3	1	0
Stringer, 1995[13]	Hospital for Sick Children, London	77 (72)	2	15	6	10	3	21	10	6	4
Iyer, 1995[29]	Children's, Los Angeles	29 (27)	2	0	0	3	1	9	8	6	0
Yang, 1996[30]	NTUH, Taipei, China	20 (17)	0	2	0	1	0	14	3	0	0
Karnak, 2000[31]	Ankara, Turkey	42 (38)	1	7	2	1	3	17	9	2	0
Puligandla, 2003[32]	Montreal Children's	73 (73)			0	6	7	51	5	4	0
TOTALS		719 (665)	7 (1%)	119 (17%)	22 (3%)	56 (8%)	40 (6%)	321 (45%)	107 (15%)	34 (6%)	7 (1%)

Fig. 1. Alimentary Tract Duplications by Location as Described in Literature Reports (George W. Holcomb III et al. Ashcraft's Pediatric Surgery 6th Edition, Ed. Saunders Elsevier, 2014, eBook ISBN: 978-0-323-18736-7, p. 539-547)

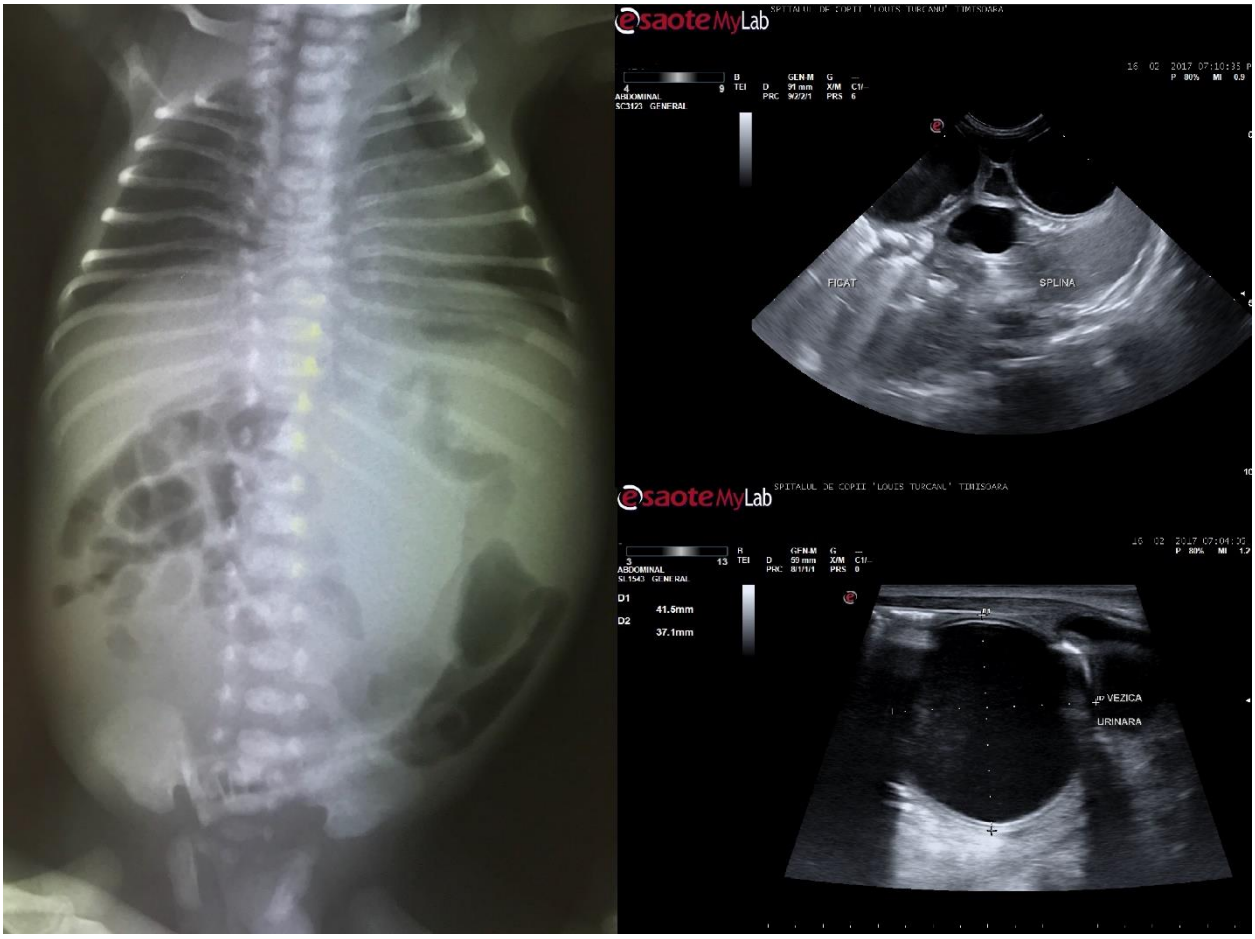

Fig. 2. Plain Thoracic and Abdominal Radiography and Ultrasound images showing the characteristics of abdominal duplication cyst.

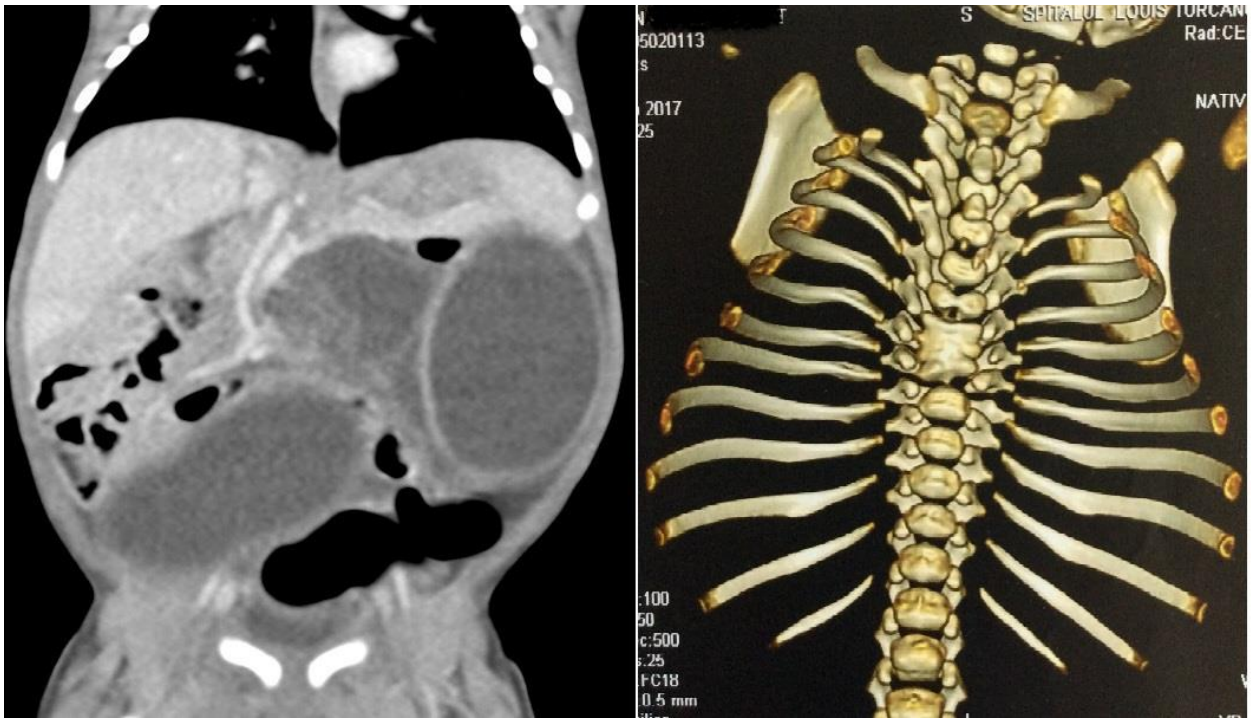

Fig. 3. Computed Tomography Image of the Duplication and 3D Reconstruction to better highlight the vertebral malformation.

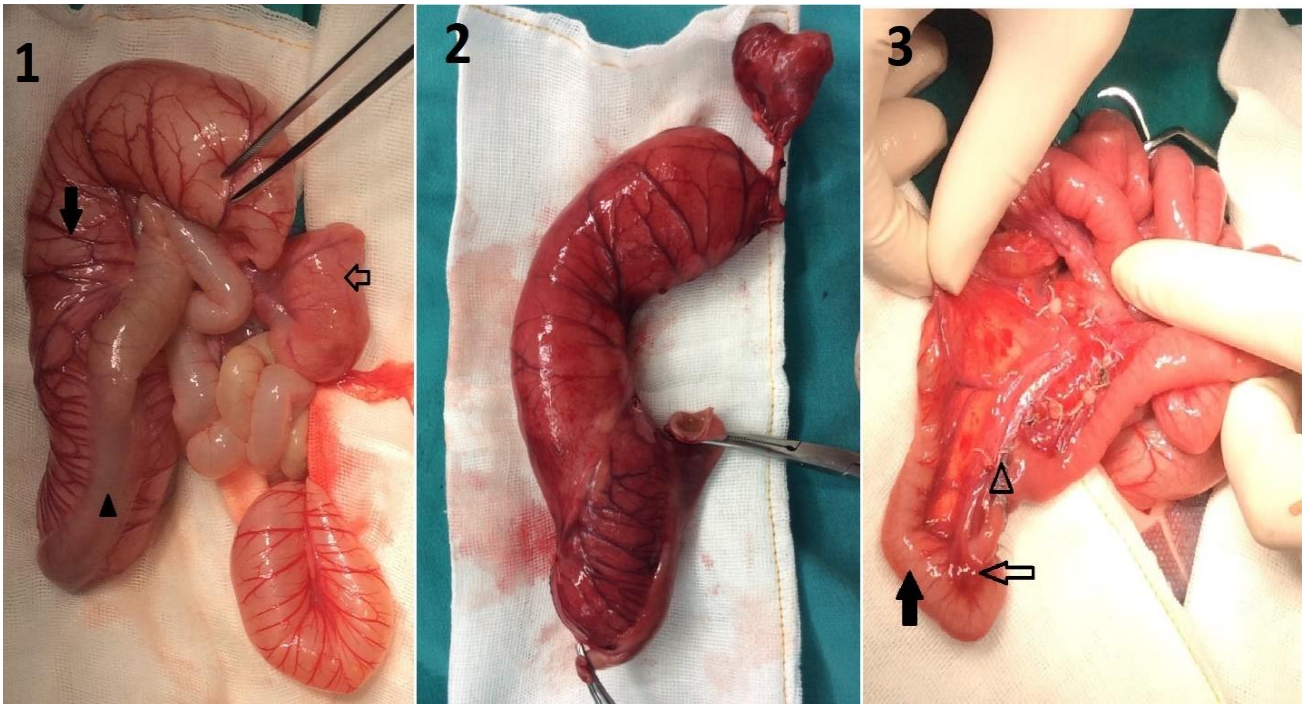

Fig. 4. First image show intraoperative aspect of the duplication. Full arrow marks the tubular duplication cyst; empty arrow marks the smaller duplication cyst; Triangle marks the terminal ileal segment of intestine where cyst is in intimate contact. In second image resection specimen includes both cysts and a segment of the small intestine which shared common blood supply. Third image showing the intestines after resection of the duplication and end-to-end anastomosis. Full arrow marks ascending colon which can be observed to be of abnormally small caliber; empty arrow marks ileocecal valve; triangle marks the end-to-end anastomosis.

The histopathological examination reveals the following aspects: the macroscopic examination - intestinal fragment of 8.5 centimeters length, presents a cystic formation of 21.5 / 3 / 3.5 centimeters attached to it. The cyst has liquid content, gray color, elastic consistency, wall thickness 0.1 - 0.2 centimeters. Microscopic examination - a small intestine duplication cyst with a wall consisting of muscle layer, submucosa and mucosa that is thinned in patches, sometimes denuded. Fibro-connective tissue that includes lymphoid structures with reactive modifications and lymphoid tissue with intestinal glandular structures, secreted by secretory cylindrical epithelium, including Paneth cells (ectopic gut) (Fig. 6).

Postoperatively major complications occurred.

Immediately postoperative evolution was good but the patient's condition progressively began to deteriorate despite proper medical management and two weeks postoperatively developed digestive intolerance with progressive increase in inflammatory markers and septic status (Fig. 5) and 18 days postoperatively, surgical reintervention is decided, intraoperatively, revealing complete disunion of anastomosis with generalized peritonitis and multiple peritoneal adhesions.

Resection of the ileocecal valve together with the cecum, closure of the ascending colon and cutaneous ileostomy is performed. The postoperative progression was favorable with slight weight gain, correction of electrolyte deficiencies. One month later, reconstruction of the digestive tract was performed by take down of ileostomy. Subsequently, the patient's evolution was to complete recovery and healing and was discharged from Surgery Department the 10th postoperative.

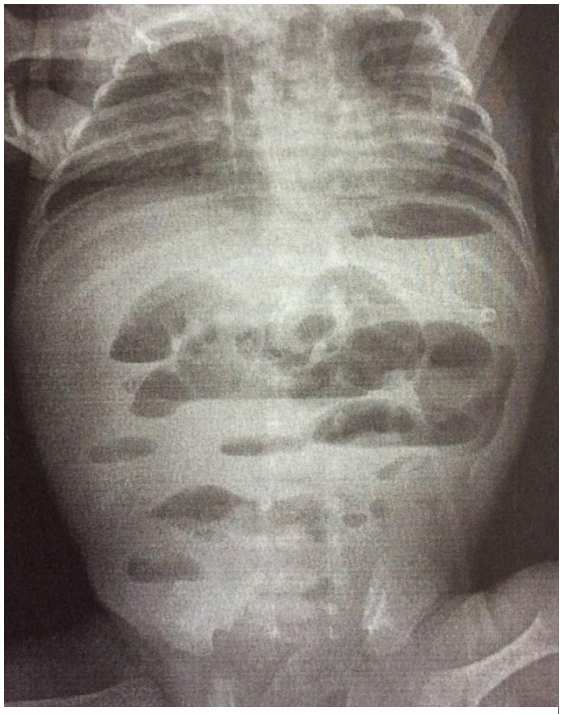

Fig. 5. Plain Abdominal Radiography showing multiple hydroaeric levels due to small-bowl occlusion.

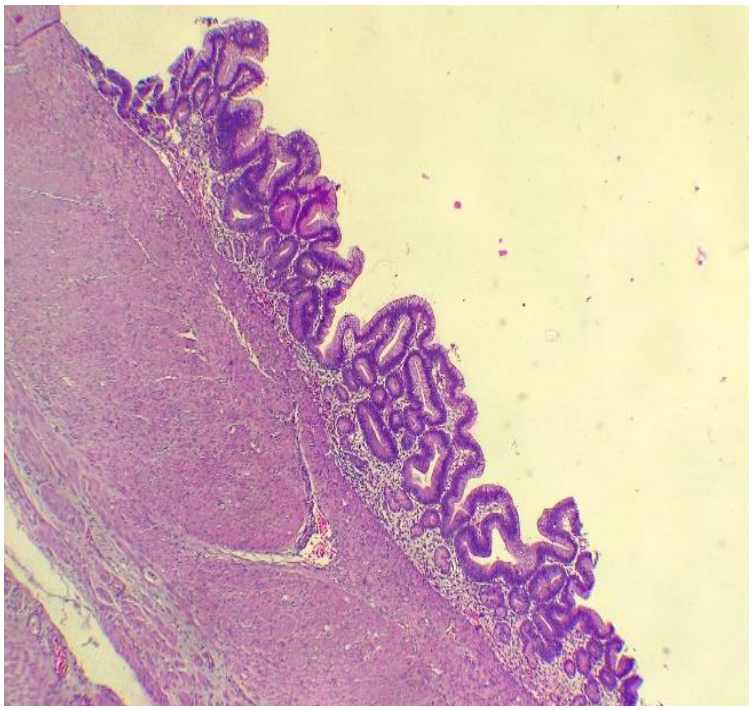

Fig. 6. Microscopic aspects of the duplication

Disscusions

Differential diagnosis was primarily made corroborating ultrasound aspects, CT and biological parameters and consisted mainly of the following pathologies without ultrasound flow:

- omental cyst;
- mesenteric cyst;
- meconium pseudocyst;
- urachal cyst;
- urinoma;
- renal congenital cyst;
- ureterohydronephrosis;
- hepatic cyst;
- choledochal cyst;

We consider echography has the highest diagnostic value in this pathology.

The cost of hospitalization for this patient was over 7,000 euros for a 64-days hospital stay. These calculations only refer to the period of hospitalization at the Surgery Department. When the patient was surgically healed, he was transferred back to the Neonatology Department and then to the Chronic Patients Care Department for nutritional and weight recovery, where he is at this time.

Although the vast majority of intestinal duplications develop in the small intestine, they are solitary and small. Multiple intestinal duplications in the same patient are a rarity, as well as the giant ones. Confrontation with this case has created the opportunity to treat a rare pathology, and its documentation represents a benefit for ourselves as well as for the medical community.

Conclusion

At the occurrence of complications (disunion of primary anastomosis), we consider contributed the poor quality of tissues due to pre-existing low serum proteins levels, but also the quality of the vascularization left after the resection of duplication cysts. Knowing that this type of cysts always develops on the mesenteric side of the healthy intestine can not be excised without resection in block with a healthy portion of the intestine, and in our case it has been preferred to excise the cysts with the resection of only a small segment of the adjacent intestine - the one with which the cyst was in intimate contact. The reasoning behind the decision was, on one hand, the fear of not cutting a large part of the gut (more than 20 centimeters) that could end up with Short Bowel Syndrome, and on the other hand that more than half of the cyst did not have intimate contact with the adiacetic intestine which allowed its dissection. But this seems to have resulted in a compromise of good vascularization necessary for proper healing of anastomosis and the occurrence of complications with the need for surgical reintervention and increasing the healing time of the patient as well as hospitalization.

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