Article

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Operative Management of Digestive Duplications in Children

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Abstract: The study aimed to review the management of pediatric digestive duplications (DD) with regards to clinical presentation, operative procedures and complications in a single institution in Bucharest, Romania. A retrospective analysis was performed using patient records from 'Grigore Alexandrescu' Emergency Children's Hospital from 2013 to 2023. All patients with a postoperative diagnosis of DD were selected. A total of 39 children (19 boys, 20 girls) with DD have been treated in our hospital in the past 10 years. Only N=5 of them have been diagnosed antenatally. In N=22 cases patients were referred to our hospital for tumors, while N=17 cases involved an acute setting. Comorbidities were encountered in N=16 patients. Duplications involved several parts of the digestive tract: ileal (N=19), colonic (N=10), esophageal (N=2), jejunal (N=2), gastric (N=2), duodenal (N=1), ileocecal (N=1), esogastric (N=1) and rectal (N=1). Two of the duplications were intraluminal, while the rest were cystic. There were N=28 open interventions and N=11 laparoscopies. Conversion was needed in N=2 cases due to (a) overdistended bowel and (b) dense adhesions between the ileum and the tumor. The tumor was resected in N=17 cases, while intestinal resection was required in N=22 cases. A drain was used in N=21 patients. The average hospital stay for open procedures was 10 days and for laparoscopic procedures 8 days. Complications arose in N=7 patients who had an open procedure and N=1 patient who had a laparoscopic procedure. There were no mortalities. With a low rate of antenatal diagnosis (12.82%), 43.58% of DD appeared in an acute setting. Open procedures were favored in acute cases and patients with comorbidities, but required a longer hospital stay. Laparoscopic management had a 18.18% conversion rate and a low rate of complications (9.09%).

Keywords: digestive duplications; open; laparoscopy; children; gastrointestinal; alimentary tract duplications

1. Introduction

Digestive duplications (DD), also referred to as alimentary tract duplications or gastrointestinal duplications are rare congenital anomalies that occur in approximately 1 in 4500 live births [1]. These anomalies can manifest in any segment of the digestive tract, extending from the oral cavity to the anus [2]. Typically, they present as single, cystic lesions that are lined with alimentary tract mucosa. While DD can potentially emerge at any age, a significant majority of cases are detected before the age of two [1]. Advances in prenatal ultrasonography have enabled more in utero diagnosis of these anomalies [1].

Several theories were proposed as pathogenesis. The split notochord theory suggests there is an abnormal adherence of the endoderm of the roof of the primitive gut to the notochord which could explain DD associated with vertebral anomalies [2]. Other etiologic events can be an intrauterine mesenteric vascular accident [2] or aberrant recanalization of the alimentary tract lumen [1]. This variety of processes can explain the high rate of comorbidities (30-50%) in patients with DD [1].

Histopathologically, the lesion is defined by three features: a well-defined smooth muscle coat, the presence of intestinal epithelium and attachment to the alimentary tract [3]. A DD is lined with heterotopic tissue in most cases and ectopic tissue in 25-30% cases [1]. Ectopic tissues include gastric and pancreatic tissue which can lead to perforation or hemorrhage [1].

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DD can communicate with the lumen of the adjacent bowel or may be found as a single, separate lesion [2].

Due to their variety of locations and forms, they can either generate acute or chronic symptomatology, with a large palette of clinical pictures. Symptoms can include: abdominal pain, vomiting, bleeding, chronic respiratory complaints and abdominal distension [1] [4].

Imaging is a very important preoperative tool [2]. Esophageal and thoracoabdominal masses need extensive work-ul involving radiographies, CT scan or MRI and sometimes upper gastrointestinal endoscopy. These can reveal the extent of the duplication cranially and caudally, as well as communications with the lumen and vertebra and the effect on neighboring organs. Ultrasound is the main tool for diagnosing an abdominal mall [4]. The typical sonographic appearance of duplications is an inner hyperechoic rim of mucosal-submucosal tissue and an outer hypoechoic muscular layer [1]. Plain radiography and barium studies can demonstrate a mass effect [1], while computer tomography (CT) can show pelvic lesions and associated defects. Moreover, magnetic resonance angiography, endoscopic retrograde cholangiopancreatography or magnetic resonance cholangiography may aid in planning surgery for large retroperitoneal lesions [2]. In the case of pelvic duplications, contrast enemas, fistulograms, endoscopies and urinary tract sonographies are used preoperatively [2].

In the case of digestive bleeding, a ^{99m}Tc-pertechnetate scan can detect the presence of gastric mucosa in an intestinal duplication, but it cannot distinguish between a digestive duplication or a Meckel's diverticulum [5].

To avoid future complications, DD are best treated by early complete excision [2]. If there is an antenatal diagnosis, the surgery is usually performed after 6 months of age [6].

The study aimed to review the management of pediatric DD with regards to clinical presentation, operative procedures and complications in a single institution in Bucharest, Romania.

2. Materials and Methods

A retrospective analysis was performed using records from 'Grigore Alexandrescu' Emergency Children's Hospital in Bucharest, Romania. Data was collected using patient files and electronic records from the last 10 years. The inclusion criteria were as follows: children (age 0 to 18 years) and a postoperative diagnosis of DD.

The primary endpoints of the analysis were the type of operative procedures and outcomes (complications and morbidity). In order to do this, several parameters were followed, where available:

- age at surgery
- initial diagnosis
- DD location
- type of procedure (open/ laparoscopic)
- technique (tumor resection/intestinal resection)
- drainage use
- associated conditions
- postoperative morbidity and complications
- follow-up interval

All data was extracted to an Excel spreadsheet and analyzed with Excel functions. The patients were then split into two groups: an open group and a laparoscopy group and statistical analysis was made using the parameters detailed above.

3. Results

In the last 10 years, a total of 39 children were treated in our hospital for DD. There were 19 boys and 20 girls aged 1 day - 15 years. The demographics of the patients are detailed in Table 1.

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Table 1. Demographics of patients with DD.

Patient number	Age at surgery	Duplication	Procedure	Complications
1	9 months	transverse colon	laparotomy	-
	3 days	terminal ileum	laparoscop	
2			y	-
3	2 months	terminal ileum	laparotomy	-
	3 years	terminal ileum	laparoscop	
4			y	-
	2 days	jejunum	laparoscop	
5			y	-
	1 months	duodenum	laparoscop	
6			y	-
7	10 months	ileocecal	laparotomy	-
8	9 months	cecum	laparotomy	-
9	14 days	terminal ileum	laparotomy	-
10	2 days	terminal ileum	laparotomy	-
11	5 years	terminal ileum	laparotomy	-
12	2 years	terminal ileum	laparotomy	-
13	5 months	ileum	laparotomy	-
14	1 day	ileum	laparotomy	vomiting
	9 day	cecum	laparotomy	sepsis, pneumonia,
15				malabsorption
16	9 years	ileum	laparotomy	intestinal obstruction
	17 years	gastric	laparoscop	
17			y	-
18	6 years	rectum	laparotomy	abscess
19	13 years	esophagus	laparotomy	pachypleuritis, thoracic pain
20	15 years	esophagus	laparotomy	pneumonia
21	6 years	ileum	laparotomy	-
			laparoscop	
22	3 weeks	jejunum	y	-
			laparoscop	
23	13 years	cecum	y	-
24	6 years	ileum	laparotomy	-
			laparoscop	
25	14 years	ascendent colon	y	-
26	10 years	terminal ileum	laparotomy	-
27	6 years	terminal ileum	laparotomy	-
28	4 years	terminal ileum	laparotomy	-
29	7 months	colon	laparotomy	-
30	13 days	ileum	laparotomy	-

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31	6 months	ascendent colon	laparotomy	-
32	8 months	ileum	laparotomy	-
33	23 days	terminal ileum	laparotomy	-
			laparoscop	
34	5 months	cecum	y	-
35	16 years	ileum	laparotomy	volvulus
36	4 months	cecum	laparotomy	-
		cecum	laparotomy	
37	9 months	intraluminal		-
			laparoscop	
38	12 months	gastric	y	vomiting
			laparoscop	
39	15 years	esophagus+gastric	y	-

Five patients have been diagnosed antenatally with tumors. In N=22 cases patients were referred to our hospital for tumors. As for the location of tumors there were: N=15 abdominal, N=2 pelvic, N=2 retroperitoneal, N=1 mediastinal, N=1 thoraco-abdominal.

Other N=17 cases involved an acute setting. Four patients were admitted to the hospital for abdominal pain and vomiting, another N=4 patients had intestinal obstruction and N=3 were found to have intussusception. Rectorrhagia was present in N=3 cases, fever in N=2 cases and N=1 patient had pneumoperitoneum. There were N=1 case of peritonitis, N=1 case of volvulus and N=1 case of perianal abscess. In one case the patient was diagnosed initially with an ovarian cyst. Another patient was operated for ovarian torsion and a DD was found incidentally.

Morbidities were encountered in N=16 patients. There was a large spectrum of pathologies associated with DD. There were N=8 digestive pathologies. In N=2 cases, patients with DD also had a Meckel's diverticulum. Enterocolitis with *Adenovirus* was encountered in a patient. A child born prematurely had necrotising enterocolitis as well as a duplicated cecum. Two patients had volvulus and an ileal duplication. A neonate had duodenal stenosis with annular pancreas as well as three ileal duplications. A five-year-old patient had gastroesophageal reflux, spina bifida and a horse-shoe kidney. There was one patient, aged five days, who presented with ovarian torsion. DD was also found to be associated with neurologic pathologies in N=2 cases: one patient with spina bifida mentioned above and one patient diagnosed with epilepsy. In N=2 cases, patients under 6 months old had a persistent foramen ovale. In one case, an accessory spleen was found intraoperatively. One patient had a maternal-fetal infection. Two patients were found to have anemia during preoperative investigations.

Duplications involved several parts of the digestive tract. The majority of DD were found in the small bowel. There were N=19 ileal duplications and N=2 jejeunum duplications. In another N=10 cases, patients were found to have colonic duplications. These were distributed as follows: (a) N=6 duplications in the cecum, (b) N=2 duplications in the ascending colon, (c) N=1 duplication in the transverse colon and (d) in one case the exact location was not noted. There were N=2 esophageal duplications. Another N=2 tumors were found in the stomach, while N=1 started in the esophagus and continued intra-abdominally to the stomach. There was one ileocecal duplication and one rectal duplication. In one case the patient had a duodenal duplication. Two of the duplications were intraluminal, one in the terminal ileum, one in the cecum, while the rest were cystic.

Regarding surgical management, there were N=28 open interventions and N=11 laparoscopies. Open interventions included several procedures. In N=7 cases, the tumor was completely resected. In N=11 cases, the tumor was resected along with the adjacent ileal segment. For N=10 colonic duplication, colectomy was required. Furthermore, N=2 appendectomies were performed, as well as N=1 desinvagination. When the tumor was approached minimally invasively, a tumor resection was

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possible in N=9 cases. Laparoscopic jejunum resections were performed în N=2 cases. In total, the tumor was resected in N=17 cases, while intestinal resection was required in N=22 cases.

Considering the location of the duplication, in the case of esophageal duplications, both tumors were resected leaving the native esophagus intact. In the case of the esogastric tumor, a partial resection of the duplication was required and the common wall mucosa was then stripped. Both gastric duplications were managed by tumor resection, without intrusion into the stomach. One duodenal duplication was completely resected without involving the native duodenum. In the case of jejunal duplication, an intestinal resection was required and an end-to-end anastomosis was performed. For ileal duplications, there were N=5 tumor resections and N=14 intestinal resections. An ileocecal duplication required removal along with the native bowel. Tumors located in the colon were either resected totally in N=3 cases, or with colectomy in N=6 cases. In one case, a cecal duplication required partial resection and mucosal stripping. The rectal duplication was removed by complete excision.

Conversion was needed in N=2 cases. One patient with transverse colon duplication had an overdistended bowel, leaving little room for laparoscopic maneuvers and therefore was converted to open surgery. Another patient with an ileal duplication had dense adhesions between the ileum and the tumor, causing the surgeon to convert from laparoscopy to laparotomy.

A drain was used in N=21 patients, after N=13 open procedures and N=8 laparoscopic procedures. Hospital stay ranged from 4 to 30 days. In the open procedure group the hospital stay ranged from 6 to 30 days, with a median of 10 days. In the laparoscopic group, the hospital stay ranged from 3 to 15 days, with an average of 8 days. Both patients with a hospital stay longer than 20 days were premature newborns under 1 month old when they were admitted and needed special care in the neonatal intensive care unit.

Complications arose in N=7 patients who had open procedures and N=1 patient who had a laparoscopic procedure. All complications are detailed in Table 1. Immediate complications consisted of either gastric (vomiting) or respiratory problems (pneumonia). One patient developed sepsis with *Klebsiella.* Late complications arose 2 months – 2 years after surgery and consisted of intestinal occlusion and infection. In the two cases of esophageal duplication, both patients had an open procedure and then developed pneumonia, thoracic pain and pachypleuritis. They were then medically treated. Two patients with an ileal duplication had an open procedure and then developed intestinal occlusion requiring further surgery. A patient with rectal duplication developed an abscess 2 months after surgery.

Patients were followed up for up to two years. There were no mortalities

4. Discussion

In the last ten years, a comprehensive analysis reveals that there were 39 DD with an equal distribution between sexes, with a 1:1 incidence ratio and a median age of 9 months among the affected individuals. Only 12.82% patients were diagnosed antenatally, emphasizing the challenge in early detection.

Symptomatology arises from obstruction, hemorrhage, infection, inflammation and intussusception [2]. This is connected to their location, presence of ectopic tissue and communication with the digestive tube.

In the case of esophageal duplications, patients may present with respiratory distress or pneumonia due to the extrinsic compression of the trachea. Narrowing of the esophageal lumen can lead to dysphagia. If gastric mucosa is present in the duplication, patients may develop hematemesis and subsequent anemia. If the duplication extends downwards to the stomach, the patient may have hemorrhage and necrosis from ectopic gastric mucosa [1].

Gastric duplications can present as abdominal pain. If the duplication communicates with the lumen of the stomach, a peptic ulceration can occur. This can lead to upper gastrointestinal hemorrhage manifested as hematemesis and melena or gastric perforation. They can also associate anemia [1,2].

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In the case of duodenal duplication, symptoms arise according to the location of the tumor. If the tumor obstructs the duodenal lumen, this can lead to occlusion with emesis, abdominal distension and gastrointestinal bleeding. If the tumor obstructs the biliopancreatic ducts, this can cause jaundice, abdominal pain, pancreatitis and vomiting.

Patients with small bowel duplications might present with gastrointestinal bleeding, anemia, vomiting. The duplication might act as a lead point for intussusception or volvulus [1].

If the duplication appears in the colon, there is a low rate of it containing gastric mucosa [1], therefore symptoms usually arise from obstruction, intussusception and volvulus. Patients are vomiting, have abdominal distension and no bowel movement.

Associated pathologies are usually found in 30-50% of cases with DD [1]. This series found a 41.02% rate of comorbidities which is in accordance with the literature. There is a large spectrum of morbidities. Digestive pathologies are found in 47.36% of patients, uro-genital, neurologic and cardiac morbidities in 10.52% cases each.

As described in the literature, esophageal duplications appear in 17% of patients, gastric duplications in 8% cases, ileum and jejunum in 45% cases and colon duplications in 15% cases [1]. In our hospital, there was a high rate of ileum duplications (48.71%), followed by cecum and colon duplications (25.64%). Esophageal, gastric and jejunal duplications accounted for 5.12% cases each, while esogastric, duodenum, ileocecal and rectum duplications were present in 2.56% cases each. Only 2 DD in this study were intraluminal, one in the ileum and one in the cecum, both leading to ileocolic resection.

DD are usually solitary tumors [1]. In this study, there was a 1-day-old boy who underwent surgery for occlusion. During laparotomy, three duplication tumors were found in the ileum, as well as duodenal stenosis. All three tumors were resected and a duodenojejunostomy was performed.

Before the widespread use of the abdominal ultrasound, DD typically resulted în intestinal complications [3]. Most commonly, DD caused bowel obstruction, intussusception and volvulus. Almost half of the cases in this series (43.58%) presented as a medical emergency. Gastrointestinal hemorrhage and long term malignant potential are also of concern; therefore pediatric surgeons do not recommend observation of DD [3]. Due to their wide variety of presentations and locations, the pediatric surgeon must be familiar with many techniques in managing DD.

In the management of DD, several factors are to be considered. First of all, the location can influence both the clinical picture and interventions. As DD can arise from any digestive structure, it is important to determine the exact origin.

Esophageal duplications might extend into the spinal column or downward through the diaphragm [1], thus requiring a multidisciplinary and staged approach. A minimally invasive approach is feasible, without significant complications [1,2]. Esophageal duplications usually develop in the right side of the thoracic esophagus and do not share a common wall [1]. In the case of esogastric duplications, they usually present as tubular masses with ectopic mucosa [1].

Gastric duplications usually occur along the greater curvature and rarely communicate with the stomach or pancreas [2]. A complete excision is required due to the risk of hemorrhage, perforation and malignancy [1]. For small greater curvature cysts, a complete resection is possible, while larger lesions could be managed by partial resection and stripping the mucosa of the common wall.

Duodenal cysts are problematic due to their relationship with the biliopancreatic ducts. Intraoperative cholangiopancreatography could show the anatomy and dictate a surgical procedure: either a partial resection and mucosectomy or a complete excision. Some even describe fenestrating an ampullary cyst into the duodenal lumen [2].

Small bowel duplications are the most common DD and complete excision is usually possible [2]. The duplication is usually encountered on the mesenteric side of the bowel and shares a common blood supply with the adjacent segment [1]. Depending on vascularization, enucleation or intestinal resection is warranted. In the case of long tubular duplications where too much intestinal resection could lead to short bowel syndrome, partial resection and mucosal stripping is a choice.

Colonic duplications might associate genital or urinary duplications or fistulas [1]. Small colonic cysts can be resected, while larger lesions require removal of the colonic segment. In the case of

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tubular colonic lesions whose resection is not possible, a distal communication can be created between the colon and the duplication [1].

Finally, rectal duplications pose a high risk of malignization [2] and can be managed by excision through a perineal approach or a posterosagittal approach.

Secondly, the type of procedure is tailored to every case considering symptoms, location and surgical expertise. With the advent of minimally invasive surgery, laparoscopy became a popular option for both diagnostic and treatment. The first literature report of laparoscopic resection of a DD came from Lee et al [5] in 2000.

Laparoscopy for DD started back in 2015 in our hospital and was performed on patients as old as 3 days. Even though this approach was favored in slightly older children, this is not statistically significant (Table 3). Open procedures were evenly distributed each year, while laparoscopy gained pace after 2015 and reached a peak in 2023 when all cases were managed minimally invasively.

Table 3. Statistical analysis between open procedures and laparoscopy

	Open	**Laparoscopy**	***p-value***
Age (months) (median)	9	12	0.3
Acute presentation (nr of patients)	16	0	<0.0001
Morbidities (nr of patients)	13	3	0.16
Tumor resection (nr of patients)	8	9	0.003
Hospital stay (days)	10	8.5	0.04
Drain (nr of patients)	13	8	0.09
Complications (nr of patients)	7	1	0.211

All of the patients who had laparoscopy for DD were operated in a non-acute setting. All patients with a diagnosis of emergency had open procedures. The association between an acute diagnosis and an open procedure is statistically significant (Table 3). Open procedures were also favored in children with comorbidities, although there is no statistical difference (Table 3).

It is considered that "laparoscopy gives a much better view of the vascularization of the lesion and adjacent organs allowing for resection of the lesion only without jeopardizing the blood supply of the remaining organ" [5]. Therefore laparoscopic enucleation is almost always possible. In this series, during laparoscopy, most surgeons performed tumor resection, while, during an open procedure, intestinal resection was most commonly performed. In one patient with cecal duplication, the cyst walls were excised and the common wall mucosa was cauterized during laparoscopy. Another patient with esogastric duplication was operated laparoscopically and needed cyst resection and cauterisation of the common wall mucosa.

A drain was used in 72.72% of laparoscopic cases and 46.42% of open cases. Having a drain was not statistically associated with the type of procedure.

Hospital stay was lower in the laparoscopy group, but not statistically significant (Table 3).

Complications arose in 25% of open cases and 9.09% of laparoscopic cases. Reintervention was required in N=3 cases. A 9-year-old patient first presented with occlusion and had an open procedure where a duplication of ileum was found. An ileal resection was performed. Two months after laparotomy, the patient developed occlusion and needed subsequent surgery. Another laparotomy was performed for adhesiolysis.

In the case of a 6-years-old child with rectal duplication, an abscess appeared 2 months after initial surgery and required reintervention for drainage.

One patient was referred to our clinic for an abdominal tumor. She was operated on at the age of 14. An open ileal duplication resection and appendectomy was performed. Two years later, she presented with occlusion. She had a redo laparotomy where intestinal volvulus was found, requiring ileal resection.

The median follow-up time was 3 months. There was no mortality.

In conclusion, patients with DD presented with acute symptomatology 43.58% of cases. There is a low rate of antenatal diagnosis accounting for 12.82% of patients. Surgeons preferred to operate on acute cases and patients with comorbidities using an open approach. The open procedure, however, required a longer hospital stay than a minimally invasive one. During laparoscopic procedures surgeons were more likely to resect the tumor, while during an open procedure, they usually performed intestinal resection. Laparoscopic management had a 18.18% conversion rate and a low rate of complications (9.09%). There was no mortality în the DD cases in our institution in the last 10 years.

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Informed Consent Statement: Informed consent was obtained from all subjects involved in the study.

Data Availability Statement: The data presented in this study are available on request from the corresponding author. The data are not publicly available due to privacy reasons - hospital records.

Conflicts of Interest: The authors declare no conflict of interest.

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