

Gastrointestinal tract duplications in children

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Abstract. – **AIM:** Gastrointestinal tract duplications (GTD) are rare congenital abnormalities that can occur anywhere along the gastrointestinal tract. These anomalies may present as a single, multiple, or a vague pathologies. Diagnosing and treating these diseases may be difficult in some patients. We aimed to present 32 patients who were followed and treated in our clinic.

PATIENTS AND METHODS: This study included the patients between 2000 and 2013. Evaluations included clinical presentations, diagnostic strategies and algorithms, surgical procedures and associated anomalies, and presence of ectopic tissue, complications, and prognosis.

RESULTS: Common clinical presentations included vomiting (n=8; 25%), palpable abdominal mass (n=4; 13%). Twenty-eight patients (2 of them antenatally) were diagnosed preoperatively while four of them were diagnosed at surgery. Ileal duplications constituted the most common type (34%) while the least common ones were located in appendix, thoracoabdomen and rectum. One of our patients was present with a gastric duplication which was closely interconnected to a tubular duplication of esophagus, which had never been encountered in the literature before.

CONCLUSIONS: It is crucial to note that duplications are likely to occur in various types and numbers and also may accompany other anomalies. Computed Tomography (CT) remains the method of choice since Magnetic Resonance (MR) is likely to cause the use of sedation and analgesia at very young ages and it may also be relatively costly despite being more sensitive in soft tissues. Mucosal stripping is an ideal method for the patients requiring restricted surgery. The antenatal asymptomatic cases can be operated after their 6th months of age.

Key Words:

Child, Gastrointestinal duplication, Surgery.

Introduction

Gastrointestinal tract duplications (GTD) are uncommon congenital abnormalities which can occur anywhere throughout the gastrointestinal

tract (GI). They mostly occur in distal ileum and have an incidence rate of 4,500¹-12,500². GTD may be single, multiple, or complex and may present challenges for diagnosis and treatment³. The pathogenesis still remains vague though errors in embryological canalization or a part of the notochord syndrome have been previously suggested¹. GTD symptoms depend on the location of the lesion. Common diagnostic methods include ultrasonography (USG), computed tomography (CT), magnetic resonance imaging (MRI), scintigraphy, and upper GI series. Several antenatal cases have been reported to be diagnosed by USG and MRI⁴.

Total excision is the method of choice in the treatment of GTD. Nevertheless, other methods of therapy may be needed depending on the localization and type of the GTD along with the length of the segment involved. We aimed to present the procedures employed in the diagnosis and treatment of 32 GTD patients who had different locations of duplications along GI tract.

Patients and Methods

This retrospective study included 32 patients who were diagnosed with GTD and received treatment at Dicle University Medical School Pediatric Surgery Clinic between 2000 and 2013. Age, gender, clinical presentations, diagnostic strategies and algorithms, surgical procedures and the patients' age at surgery, location of GTD, associated anomalies, presence of ectopic tissue, complications, and prognosis were evaluated. As per our clinical algorithm, the patients presenting with abdominal complaints were primarily diagnosed with abdominal graphy + USG. Upper GI series was performed in the patients suspected with passage problems such as vomiting and digestion. Depending on these examinations, CT was used in most of the patients and MRI was

used only for a limited number of patients. Scintigraphy was rarely used, only for studying the ectopic mucosa.

Results

The 32 patients consisted of 9 (28%) male and 23 (71%) female patients with a mean age of 2.7 years (5 days-10 years). Eight of them were infant and 5 of them were neonate at admission. Common clinical presentations included vomiting (n=8; 25%), palpable abdominal mass (n=4; 13%), and abdominal distension (n=5; 16%). USG was performed in 28, CT in 21, MRI in 3, and scintigraphy in 5 patients. Twenty-eight patients (2 of them being antenatal) were diagnosed preoperatively, whereas 4 patients were diagnosed incidentally during the exploration of invagination, acute abdomen, cloacal malformation and anal atresia + RV fistula. Ileum (n=11; 34%) constituted the most common site of GTD, followed by duodenum (n=5; 16%), jejunum (n=3), colon (n=3), stomach (n=2), ileocecum (n=2), appendix (n=2), thoracoabdomen (n=2), esophagus (n=1), and rectum (n=1) (Table I and Figures 1, 3).

One of our patients was present with gastric duplication which was closely interconnected to tubular duplication of esophagus (Figure 2). In

this patient, a cystic formation in the greater curvature of the stomach with the size of 10 × 6 cm was detected to be thoroughly linked to a tubular duplication of 4-5 cm stretching along the esophagus gut. The gastric cyst was managed with mucosal stripping from the common wall and the tubular esophagus segment was excised through total excision. Histopathological examination on the samples obtained from the gastroesophageal junction revealed two distinct patterns of mucosal tissue (gastric and oesophageal) each of which complying with their source.

Seven patients were present with associated anomalies. Common anomalies included diastematomyelia, BlockVertebra, malrotation, congenital dislocation of the hip, hydronephrosis, Cloaca, and anal atresia with rectovestibular fistula. Cystic GTD were present in 25 (78%) and tubular in 7 (22%) patients. Surgery was performed in 30 patients following the diagnosis, whereas 2 asymptomatic patients were diagnosed antenatally and received surgery after the 6th month of age. Twenty-seven patients received laparotomy, 2 laparoscopy, 2 laparotomy + thoracotomy, and 1 thoracotomy. Nineteen patients underwent resection anastomosis, 12 mucosal stripping, and 1 patient received both. Ectopic gastric mucosa was observed in 6 (19%) patients (5 in the stomach and 1 in the pancreas). One patient developed wound area infection and another

Table I. Clinical characteristics of duplications.

	Patient n (%)	Type (n)	Complaint (n)	Ectopic tissue (n)	Additional anomalies (n)
Esophagus	1 (3)	Cystic (1)	Respiratory distress (1)	Gastric (1)	
Gastric	2 (6)	Cystic (2)	Vomiting (2)		
Duodenum	5 (16)	Cystic (5)	Vomiting (5)		
Jejunal	3 (9)	Cystic (3)	Vomiting (1) Mass (1)		
Ileal	11 (34)	Tubular (1) Cystic (10)	Abdominal distension (4) Mass (3) Abdominal pain (2) Bleeding (1)	Pancreas (1) Gastric (1)	Malrotation (1) Congenital hip dislocation (1)
Thoracoabdominal	2 (6)	Tubular (2)	Respiratory distress (2)	Gastric (1)	Diastematomyelia (1) BlockVertebra (1)
Ileocecal	2 (6)	Cystic (2)	Abdominal distension (1) Abdominal pain (1)		Hydronephrosis (1)
Appendix	2 (6)	Tubular (2)	–		Cloaca (1) AA + RV fistula (1)
Colonic	3 (9)	Tubular (2) Cystic (1)	Constipation (1) Bleeding (1) Abdominal pain (1)	Gastric (1)	
Rectum	1 (3)	Cystic (1)	Constipation (1)		
Total	32			5	7

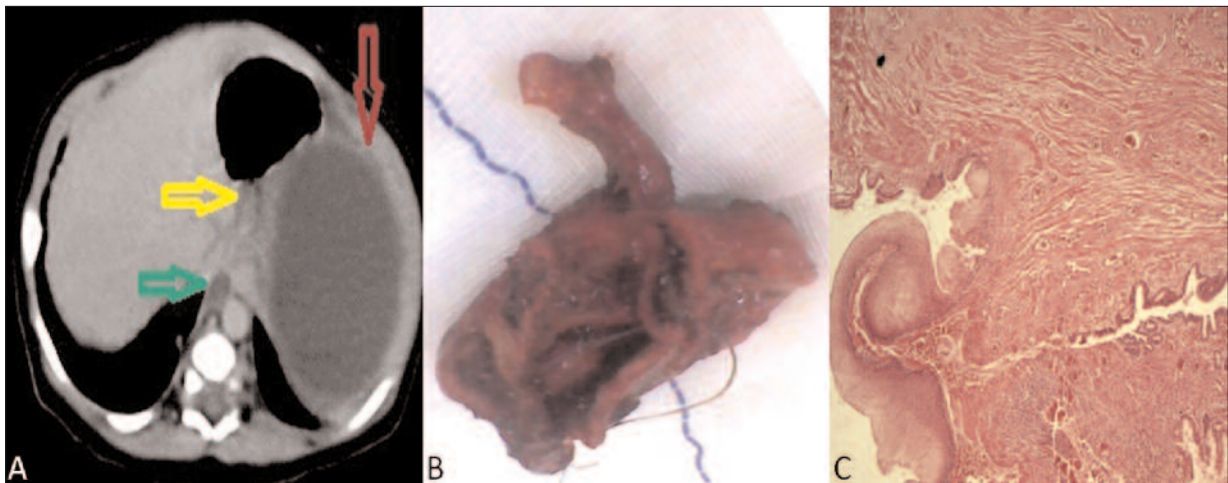


Figure 1. *A*, Axial CT section; stomach (yellow arrow), gastric duplication cyst (red arrow), esophageal duplication cyst (green arrow). *B*, Macroscopic appearance. *C*, Microscopic appearance and transition zone

patient developed treatable ileus. One patient died of sepsis. Mean hospital stay in our series was 6 days.

Discussion

GTD are rare abnormalities which are generally diagnosed at early ages. These anomalies may occur anywhere along the gastrointestinal tract from the mouth down to the anus. GTD may be asymptomatic or present with vague symptoms mimicking some other common pathologies (e.g.

acute abdomen invagination, Hirschsprung's disease, etc.) or may accompany these diseases as well^{5,6}. These duplications are best explained by the development of fetal intestinal diverticulitis, vacuolization, caudal duplication and split notochord theory^{7,8}. GTD cysts may be single or multiple and they are twice more common in females than males^{9,10}. Similarly, the females in our study had an outstanding preponderance (71%) over males. Jejunal and/or ileal duplications (44-53%) comprise the most common type, followed by colonic (13-15%), gastric (7-9%), duodenal (4-6%), and rectal (3-4%) duplications^{4,10,11}. In a

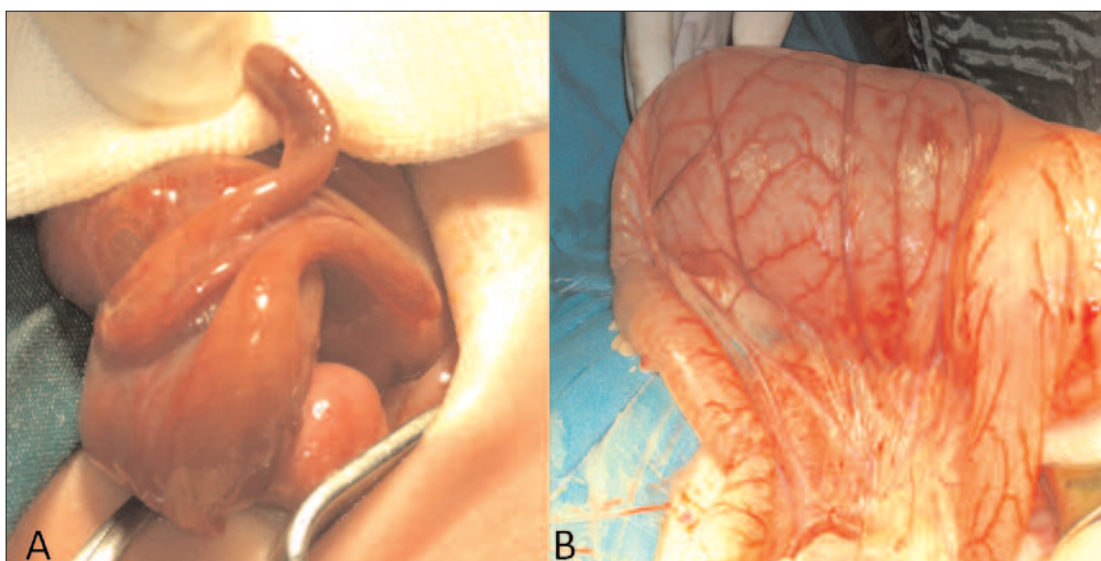


Figure 2. *A*, Appendiceal. *B*, Colonic duplication.

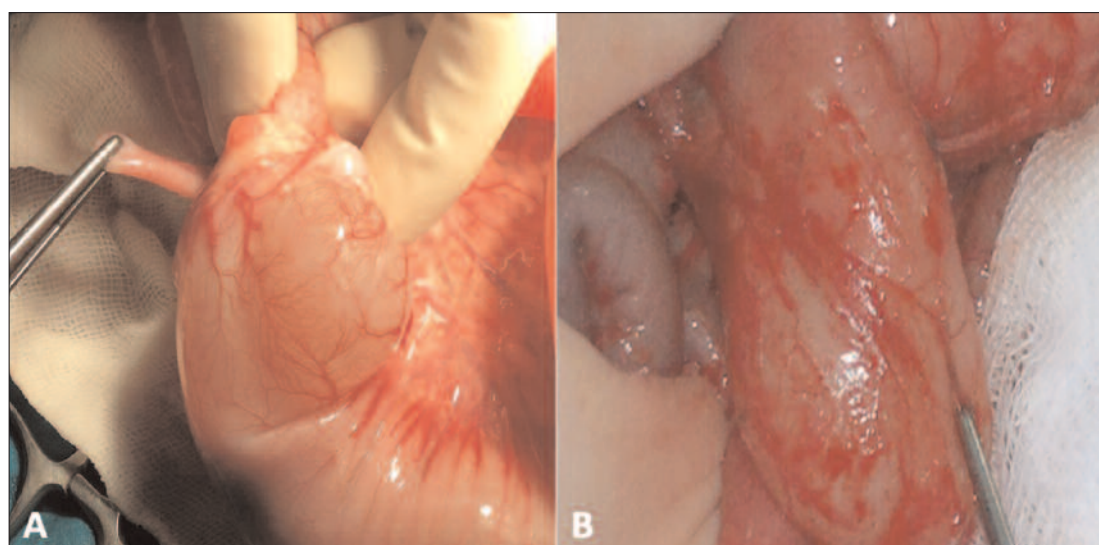


Figure 3. *A*, Ileocecal. *B*, Duodenal duplication.

similar vein, 47% of our patients had jejunal and/or ileal while 16% had colonic and 3% had rectal duplications.

Thoracoabdominal duplications are reported to constitute 2-4% of all GTD types¹¹⁻¹³. These duplications were only present in two of our patients (6%). Complex duplications are also reported in the literature; however, in one of our patients, who was present with gastric duplication, we observed something which had never been reported before: the patient presented with a cystic duplication in the stomach which was closely related to the tubular duplication in the esophagus. Keeping this notion in mind, the physician should conduct a detailed exploration in GTD patients since these anomalies are quite likely to occur in unexpected ways.

The symptoms of GTD are often related to the location, size and mucosal lining of the duplications. Oral and esophageal lesions may cause respiratory difficulties while gastrointestinal duplications may lead to nausea, vomiting, obstruction, hemorrhage, or perforation^{5,14}. In our series, the patients with esophageal duplications presented with respiratory difficulties and the ones with gastro duodenal lesions were mostly suffering from vomiting and nutritional problems. In ileal and jejunal duplications most common symptoms were palpable mass, abdominal distension, pain, and bleeding, respectively. One patient with ileal duplication was present with invagination. Another patient with ileocecal lesion was brought to our clinic with an abdominal

pain. Colonic and rectal duplications are mostly accompanied by constipation and bleeding. We consider that GTD must be suspected in the patients suffering from any of the symptoms aforementioned.

USG, the graphy of upper GI, CT, MRI, and scintigraphy are commonly reported as useful visualization techniques for GTD¹⁵⁻¹⁷. Although usg is valuable for diagnosis, in some cases USG presents a restrictive use particularly in rectal duplications and also in detecting the relations of the lesions with their surroundings and associated anomalies. Instead, MRI or CT may be employed to provide a clear imaging of these situations. Moreover, MRI is reported to be quite sensitive in soft tissues¹⁸. On the other hand, MRI has some disadvantages such as causing sedation or anesthesia in the early years of age and being relatively costly. In our series, CT was the common technique used for visualization. We consider that CT must be the method of choice when MRI seems impractical. The series of upper gastrointestinal may be useful for the patients presenting with nutritional problems and vomiting.

Scintigraphy has been reported useful in detecting the ectopic gastric mucosa. Ectopic mucosa does not exist in 68-73% of the GTD patients. In the patients with ectopic mucosa, the mucosa is generally of gastric origin while pancreatic origin is also possible^{12,13}. Moreover, gastric mucosa can also originate from Meckel's diverticulum¹⁹. Conducting several experimentations using the gamma camera technique, Priebe

et al²⁰ reported that an ectopic tissue with a size of at least 1.8 cm² is needed to provide a clear imaging for the ectopic gastric mucosa. Other studies argue that the substance injected in the cases with peptic ulceration, inflammation, intussusception, hemangioma, arteriovenous malformations, bowel obstructions, and urethral anomalies may lead to nonspecific uptake²¹⁻²³. There is no current consensus as to whether the use of scintigraphy in the detection of ectopic tissue makes any effect on determining the surgical strategy.

In the cases where surgery is restricted due to the site of the lesion, the first thing to discover is to detect the presence of ectopic mucosa. In the partial resection of ectopic gastric mucosa, the cyst should be excised since acid secreting mucosa may lead to complications such as haemorrhage, erosion, or malignant degeneration^{6,24}. Scintigraphy was used in our two patients who presented with lower GI hemorrhage, and in another patient the colonic duplication was excised at surgery since it had been suggested by scintigraphy to be the origin of bleeding in the ectopic gastric mucosa. The patient who did not present with ectopic mucosa on scintigraphy was operated under elective conditions since the hemorrhage stopped spontaneously. Depending on scintigraphic images, 4 patients had negative and 1 patient had positive findings. These results were confirmed by the pathological analysis. We consider that scintigraphy could only be used in the patients undergoing restricted surgery, who present with unlocatable hemorrhage, to clarify whether the bleeding stems from the ectopic gastric mucosa.

Associated anomalies are reported with an incidence rate of 16-26%²⁵. The frequency of associated anomalies in our series (22%) was similar to the rate reported in the literature, and vertebral anomalies constituted the 6% of our patients. Surgical intervention is generally performed at the age of diagnosis; however, the surgery for antenatal patients may be performed after 6th month of age since they are generally asymptomatic^{4,5,14}. Most of our patients underwent surgery shortly after their diagnosis while only 2 antenatal patients were operated on in the 6th month of their ages. Commonly reported surgical procedures are thoracotomy, thoracotomy + laparotomy, laparotomy, or laparoscopy^{4,11,13}.

Mucosal stripping was used to excise the cysts in the cases with limited resection anastomosis. Mucosal stripping is an ideal method specifically

for ileocecal duplications and for the cases with multiple lesions who have the risk of short bowel. One of our patients received mucosal stripping in a combination with resection anastomosis. We believe that a detailed exploration is essential in GTD since these anomalies may occur as multiple duplications. Reported mortality rate is 4-20% while the rate in our series was only 3%. One patient died of sepsis following the operation for esophageal duplication. We attribute our low rate of mortality to the recent advancements in the diagnostic and treatment techniques.

Conclusions

It is commonly accepted that GTD cysts may occur in different shapes and numbers and also may accompany other anomalies. CT remains the method of choice since MRI is likely to cause the use of sedation and analgesia at very young ages and it may also be relatively costly despite being more sensitive in soft tissues. Scintigraphy is rarely needed in GTD cases. While resection is the method of treatment for duplications, mucosal stripping is quite useful in the cases with restricted surgery. Surgical intervention is generally performed at the age of diagnosis, whereas the surgery for the patients diagnosed at antenatal period may be performed following the 6th month of age since they are generally asymptomatic.

Conflict of Interest

The Authors declare that there are no conflicts of interest.

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