

Alimentary Tract Duplications in Children

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This report reviews the experience with 78 alimentary tract duplications found in 64 patients over a 40 year period at the Children's Hospital of Pittsburgh. Ten patients had the duplication discovered at autopsy. Multiple duplications were found in 15% of patients. The symptoms and physical findings of a duplication varied with location, size and mucosal pattern. About one-fifth of the duplications contained ectopic mucosa, usually gastric. Two-thirds of the patients were diagnosed prior to one year of age. Vertebral anomalies, as a clue to the presence of the lesion, were present in 15% of the patients. The most common indications for surgery included a mediastinal or abdominal mass, intestinal obstruction, and gastrointestinal bleeding. The results of surgery were favorable, with a mortality of 20%. Surgical complications accounted for six deaths, while four children died of severe associated anomalies. Three others died without surgical treatment, but with symptoms from the duplication.

DUPLICATIONS OF THE ALIMENTARY TRACT are congenital lesions, cystic or tubular in shape, and composed of muscular walls with gastrointestinal mucosal lining. They are almost invariably adjacent to the alimentary tract, may communicate with the lumen, and may be multiple. These lesions can occur throughout the alimentary tract, but lesions of the tongue¹⁰ and pharynx are much less common than those originating in the esophagus, stomach, small bowel and colon.

In 1884, two years prior to his classical description of appendicitis, Reginald Fitz used the word "duplication" to describe what he thought were remnants of the omphalomesenteric duct.⁷ The terms "enterogenous cyst," "ileum duplex," "giant diverticula" and "unusual Meckel's diverticula" have been used to describe congenital cystic or tubular abnormalities of the gastrointestinal tract. In 1937, W. E. Ladd, reporting ten

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cases, used the phrase "duplications of the alimentary tract . . . in the hopes of simplifying the nomenclature."¹² Gross, et al. supported the use of the term "duplication" in their classic work in 1952 in which they explained the inaccuracy of terms.¹⁰

Material

We have reviewed 78 duplications in 64 patients seen at Children's Hospital of Pittsburgh from 1935 to 1975. Ten patients had multiple duplications. Five patients were black and 59 were Caucasian. Included in this report are 27 previously reported patients.^{13,18}

Fifty-two of the 64 patients had either symptoms or physical findings leading to the surgical discovery of the duplication. Six patients had the diagnosis made at autopsy, and four of the thoracic duplications were unsuspected until discovered by chest x-ray. The duplications were diagnosed in 54 patients (85%) prior to age two years, and in 39 patients (60%) prior to age six months. However, a significant number of patients may not become symptomatic until adult life.¹

This series includes 16 thoracic, 12 gastric or duodenal, 34 jejunal or ileal, 14 colonic or appendiceal, and two retroperitoneal duplications (Fig. 1). The embryology, symptomatology, diagnosis, therapy and results vary with the anatomical location of the lesion.

Clinical Pictures

Thorax

Thoracic duplications are best explained by the endodermal-ectodermal adhesion,¹⁶ or the split notochord theory. An embryological error allowing endoderm to herniate through the notochord at three to six weeks of development is currently the best explanation for posterior mediastinal duplications. Vertebral defects are often associated with these duplications as part of the embryological error (Fig. 2).⁴

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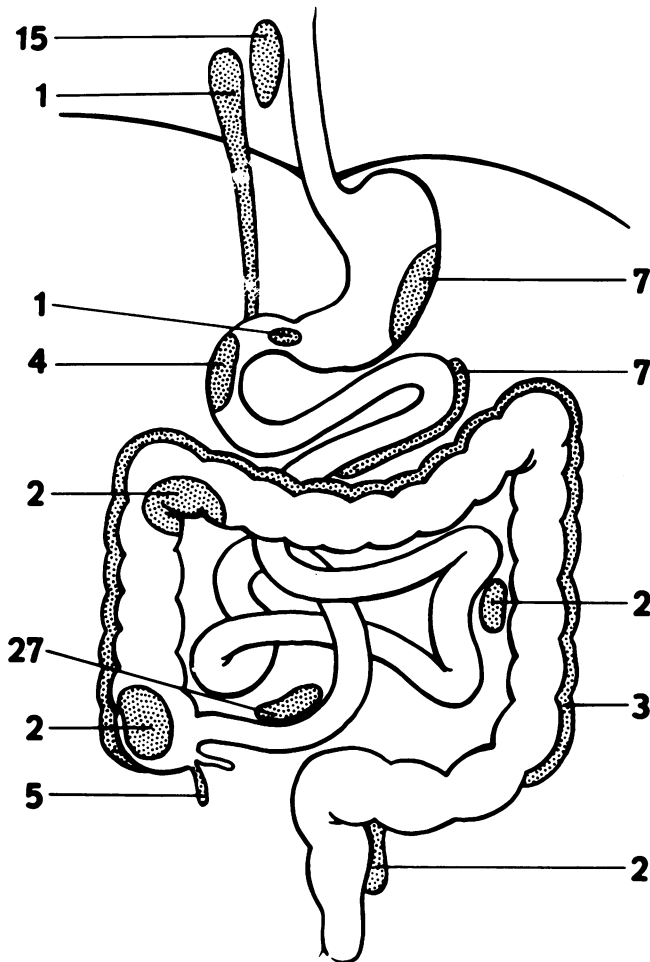


FIG. 1. Diagrammatic location of the 78 duplications.

There are 16 thoracic duplications in this report. Most often, respiratory symptoms led to the diagnosis (Table 1). Three infants presented with severe respiratory obstruction on the first day of life. Haller¹¹ has stressed that duplications should be considered in the differential diagnosis of severe respiratory obstruction in the newborn. Duplications are second only to neural tumors as a cause of posterior mediastinal masses in children. Ectopic gastric mucosa was present in seven of these cases. Peptic ulceration can cause pain, bleeding, or rarely perforation of the duplication, and one of our patients had anemia and one had hematemesis. Spinal cord compression is rare in spite of severe vertebral anomalies.

Chest roentgenograms usually show the duplication as a posterior mediastinal mass. Occasionally, a barium swallow shows esophageal compression from the cyst. Because of the incidence of ectopic gastric mucosa, it is sometimes possible to diagnose duplications preoperatively by the use of a technetium scan.⁶

Three of the 16 cases had hemivertebrae (Fig. 2).

Only one of the 16 cases had an abdominal extension of the duplication, and two other patients with thoracic duplications had separate jejunal duplication.

Of the 15 patients undergoing surgical resection, 12 had a right thoracotomy and three a left; others have also noted the preponderance of right-sided lesions.¹⁰ Operation is undertaken because of the possible complications of bleeding, perforation, respiratory obstruction, and difficulty in differentiating the mass from a posterior mediastinal malignancy. If there is a common cyst-esophagus wall, only the cyst mucosa of the shared wall need be resected while removing the remainder of the mass *in toto*. Care should be taken to be sure that there is no neuroenteric communication.

One patient died of pneumonia at age two weeks without operation and a duplication between the esophagus and trachea was found at autopsy. Another patient died postoperatively of aspiration while a third has severe brain damage secondary to aspiration. The other 13 patients underwent resection of the duplication without problems (Table 1).

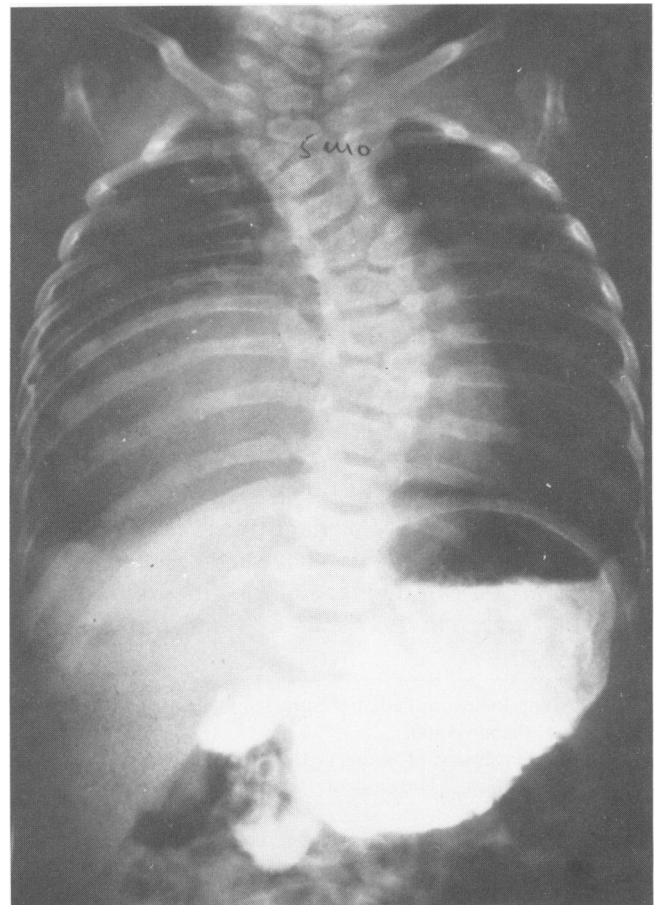


FIG. 2. Hemivertebrae in five-month-old with right-sided thoracic duplication.

TABLE 1. Thoracic Duplications

Pts.	Age	Symptoms	Size (cm)	Ectopic Mucosa	Result
1.	1 day†	Respiratory Obstruction	5 × 3	Gastric	Postop aspiration; recovery
2.	1 day*	Respiratory Obstruction	6 × 3	Adrenal Cortex	Resection; recovery
3.	1 day	Respiratory Obstruction	3	—	Died without operation
4.	2 day	Aspiration	3	—	Died 2 mos. postop aspiration
5.	6 wks	Pneumonia	4.5	—	Resection; recovery
6.	4 mos	Pneumonia	4	Gastric	Resection; recovery
7.	4 mos	Pneumonia	4	Gastric	Resection; recovery
8.	6 mos	No symptoms	3.5	Gastric	Resection; recovery
9.	10 mos†	Anemia	2.5	Gastric	Resection; recovery
10.	11 mos*†	No symptoms	6 × 5	—	Resection; recovery
11.	19 mos	Wheezing	7	Gastric	Resection; recovery
12.	8 yrs	Cough	4	—	Resection; recovery
13.	8 yrs	Pain	11 × 6	Gastric	Resection; recovery
14.	10 yrs	Hematemesis	2.5	—	Resection; recovery
15.	11 yrs	No symptoms	16 × 3	—	Resection; recovery
16.	16 yrs	No symptoms	7	—	Resection; recovery

* Also had separate jejunal duplication

† Hemivertebrae

Stomach and Duodenum

Gastric and duodenal duplications were once considered quite rare, but recent reviews report over 50 examples in each location.¹³ Pyloric duplications are extremely rare with only three reported up to 1970.⁹

The etiology of duplications of this area is unknown, but the diverticulum theory of Lewis and Thyng¹⁴ and the Bremer³ theory of failure to recanalize are often mentioned; neither completely explains the origin. It is of interest that gastric duplications are twice as common in girls;² three of our four were found in girls.

Vomiting, secondary to partial or complete obstruction and an upper abdominal mass are almost always present in the clinical course (Table 2). The diagnosis can sometimes be made by an upper G.I. series which may show an extrinsic gastric mass (Fig. 3) and depression of the transverse colon. Peptic complications of pain, bleeding and perforation, as well as increased gastrin secretion, have been reported. The one pyloric duplication was found after a preoperative diagnosis of hypertrophic pyloric stenosis.

Gastric duplications are most often found on the greater curvature and usually do not communicate with the stomach.² A carcinoma has been found in an adult gastric duplication.¹⁵

Because of their location, gastric duplications can usually be excised by a limited partial gastrectomy. Unusual locations may necessitate more complete gastric resection. Duodenal duplications often lie behind the duodenum and usually do not communicate with the bowel lumen.¹³ The first reported surgical

TABLE 2. Gastroduodenal Duplications

Patients	Age	Symptoms	Size (cm)	Location	Result
1.	7 mos	None (autopsy)	1	Stomach	Autopsy finding
2.	2 mos	Obstruction	5	Stomach	Died 2° to perforation of gastric duplication
		Masses	3 & 3	Duodenum	
		Melena			
3.	1 mo	Obstruction	4 × 3	Stomach	Resection; recovery
		Masses	2 × 1	Stomach	
4.	2 days	Vomiting	2.5	Duodenum	Died after resection
		Mass			
		Jaundice			
5.	3 mos	Mass	4	Stomach	Resection; recovery
6.	2 wks	Vomiting	4 × 2	Stomach	Resection; recovery
		Mass			
7.	3 wks	Vomiting	1.5	Pylorus	Excision; recovery
		"olive"			
8.	1 mo	Mass	3	Duodenum	Roux Y; recovery

procedure for duodenal duplication was that of excising the common wall so that the duplication opened into the duodenum.⁸ Cystoenterostomy by the Roux-Y technique has been reported with satisfactory results in some cases.¹³ Occasionally, extraluminal excision of duodenal duplications is possible.

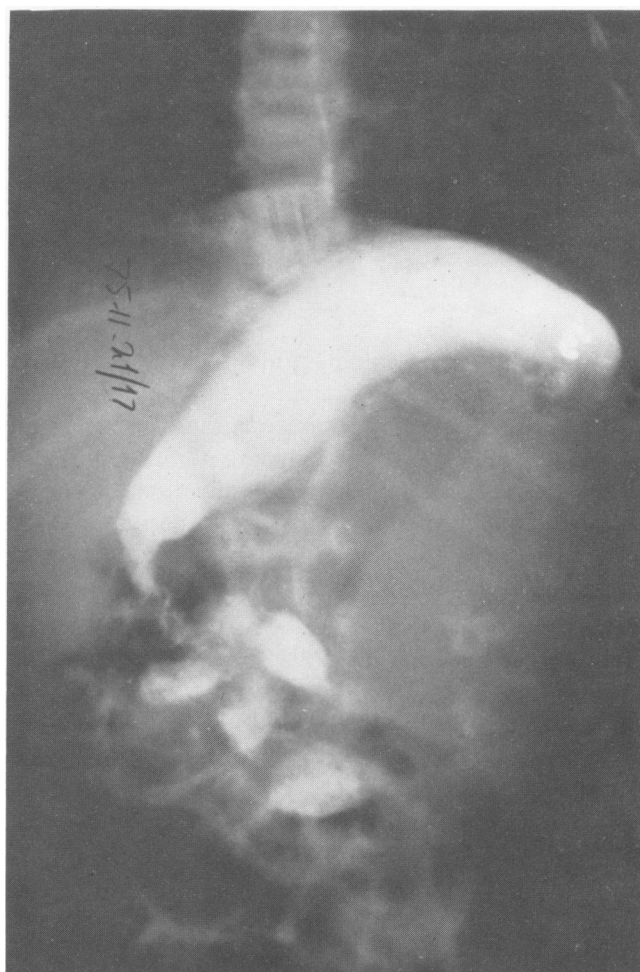


FIG. 3. Large gastric duplication on greater curvature.

TABLE 3. Small Bowel Duplications

Patients	Age	Size (cm)	Clinical Presentation	Location	Ectopic Mucosa	Result
1.	3 mos	1	none (autopsy)	Ileum	—	Died; congenital heart disease
2.	2 mos	1	none (autopsy)	Ileum	—	Autopsy finding
3.	5 mos	20*	B	Jejunum	Gastric	Resection; recovery
4.	4 mos	3	M, I	Ileum	—	Died postop
5.	13 yrs	12, 9, 4	O, B, V	Ileum × 3	—	Resection; recovery
6.	2 mos	8 × 7	O, M	Ileum	—	Resection; recovery
7.	4 mos	1.2	I	Ileum	—	Resection; recovery
8.	2 yrs	2	O	Ileum	—	Resection; recovery
9.	1 day	5 × 4	O	Ileum ^a	—	Resection; recovery
10.	7 days	4	O, M, B	Ileum	—	Resection; recovery
11.	4 days	2	O	Ileocecal	Colonic	Resection; recovery
12.	21 mos	5	I, M	Ileocecal	Colonic	Resection; recovery
13.	2 wks	2.5	B	Ileocecal	Colonic	Resection; recovery
14.	11 mos	2	O	Ileocecal	Colonic	Resection; recovery
15.	1 day	3.5	O, M	Ileum	—	Died; sepsis, volvulus
16.	2 mos	2.5	O, I	Ileum	—	Resection; recovery
17.	1 day	15*	O	Ileum	Gastric	Died; anastomotic leak
18.	1 day	1	O, M	Jejunum ^a	—	Died; respiratory failure
19.	5 wks	4	O	Ileum	—	Resection; recovery
20.	4 days	3	none (autopsy)	Ileum	—	Died after TEF repair
21.	2 days	10	none (autopsy)	Jejunal ^v	—	Died after omphalocele
22.	10 mos	16*	B	Jejunal ^{t,v}	Gastric	Resection; recovery
23.	11 mos	3	O	Ileal ^a	—	Died; sepsis
24.	6 wks	7 × 5	M	Ileal	—	Resection; recovery
25.	8 mos	10 × 7	M	Jejunum	—	Resection; recovery
26.	5 mos	5	M	Ileum	—	Resection; recovery
27.	3 wks	15*	M	Ileum	—	Resection; recovery
28.	8 mos	2.5	M	Jejunum ^t	Gastric	Resection; recovery
29.	18 mos	7*	B	Ileum	Gastric	Resection; recovery
30.	2 days	4	O	Ileum ^a	—	Died postop
31.	5 days	3, 2	O, M	Ileum × 2	—	Resection; recovery

* tubular O — obstruction a — atresias
M — mass t — thoracic duplication also
B — bleeding v — vertebral defects
I — intussusception
V — volvulus

Of the six gastric duplications (in five patients) in this series, one was an incidental finding at autopsy, one was found at autopsy and was the cause of death (perforation), and three patients are doing well following resection, one having two duplications. The one pyloric duplication had a satisfactory result from the type of excision described by Grosfeld.⁹ Two duodenal duplications were found incidentally in the child who died from a perforated gastric duplication. One duodenal duplication patient died shortly after attempted resection, and the other patient had a cysto-Roux-Y enterostomy and is doing well nine years later.

Small Bowel

The ileum is the most common location for duplications of the alimentary tract. These are located on the mesenteric border and have a common blood supply with the adjacent small bowel. The duplications may be spherical or tubular. Approximately 50% of patients in this series had small bowel duplications. Two cases had multiple lesions (Table 3).

Six duplications occurred in the jejunum and 28 were found in the ileum or ileo-cecal area (Fig. 4). Five lesions were tubular (6.8–20 cm in length); the remainder were spherical (1–12 cm). Five of the duplications contained ectopic gastric mucosa and three of these presented with bleeding. A preoperative diagnosis of small bowel duplication has been made in some cases by use of the technetium scan.²⁰ In addition to blood loss, the clinical findings in this group included obstruction in 15 cases and a palpable abdominal mass in 12 cases. Coexistent intestinal atresias were present in four cases and a vascular etiology for such duplications has been proposed by Favara.⁵

Spherical duplications usually require a segmental resection of the duplication and the adjacent small bowel. Occasionally, long tubular duplications would require resection of too much normal small bowel; in this situation, the distal portion of the tubular duplication can be opened into the normal lumen with the duplication being left in place.

Twenty-one of the 31 patients had resection of their duplication without problems. Four patients had an asymptomatic duplication discovered at autopsy.

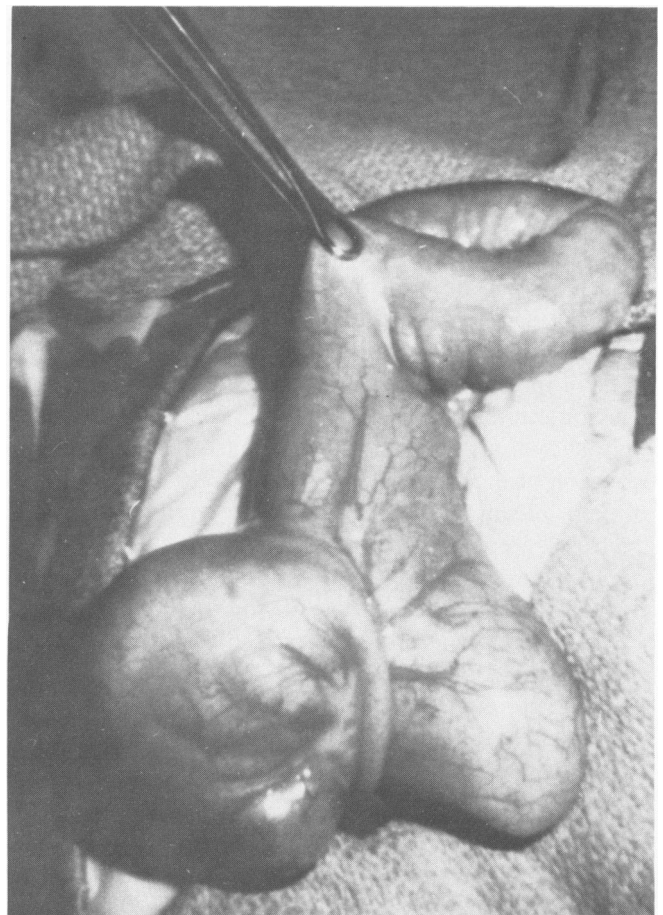


FIG. 4. Cystic duplication in mesentery near ileo-cecal valve.

There were six deaths in this group, two of the deaths resulting from complications of the multiple atresias that were associated with the duplication. One death was caused by agenesis of the lung and respiratory failure, and one death was the result of volvulus with extensive gangrene of the small bowel. Two final patients died directly as a result of complications related to resection of the duplication.

Appendix, Colon and Rectum

Fourteen duplications of the appendix, colon or rectum were found in this review (Table 4). Three (#9, #10, #12) were discovered incidentally at autopsy, three presented because of obstruction (#1, #2, #3) and the remainder were discovered by the presence of a rectal or abdominal mass.

Of the 14 patients with hindgut duplications, bladder anomalies were present in the two patients (#4, #5) who had duplicated appendices, and three had total colonic duplication with genitourinary anomalies (Fig. 5). One of the appendiceal patients is alive and well after duplication resection numerous procedures to reconstruct the urinary tract, while the other one died postoperatively from aspiration. One of the colonic patients (#3) did well after the two lumens were joined distally as suggested by Soper.¹⁹ Another (#1) was cured by partial colon resection only to die of

congenital heart disease six months later; the third (#2) died in 1944 of "megacolon" without operation.

Many comments have been made concerning the relationship between duplications of the hindgut and genitourinary anomalies. Although any hindgut duplication may have related genitourinary anomalies, the tubular duplications of the entire colon are the type most commonly found to have double bladders, double urethras or double external genitalia.¹⁹ Ravitch concluded that these associations occur as a result of partial twinning.¹⁷

Total colonic duplications, which communicate proximally but not distally, are best treated by distal fenestration. Because of the frequency of associated urinary anomalies in this type, appropriate evaluation of the genitourinary tract is indicated preoperatively. Cystic duplications of the colon are handled by partial colectomy; this was done in our three symptomatic cases with one death from sepsis, the two additional cases being incidental autopsy findings. Rectal duplications may be totally resected or the common rectal-duplication wall removed, one of our patients was so treated successfully and the other had his duplication as an unexpected autopsy finding. The two retroperitoneal duplications were treated by excision with uneventful recoveries in both instances.

Colonic duplications occasionally have ectopic gastric mucosa; peptic ulceration therefore is a potential complication. Another theoretical risk of any

TABLE 4. Appendix, Colon, Rectum

Patients	Age	Size (cm)	Clinical Feature	Location	GU or Vertebral Anomaly	Result
1.	18 mos	Total colon + appendix + ileum 6	Obstruction Rectal bleeding	Anti-mesenteric	Extrophy of bladder Meningomyelocele	Died 6 mos. postop; truncus arteriosus
2.	8 mos	Total colon + appendix	Obstruction	Anti-mesenteric	Double bladder	Died without surgery (1944)
3.	14 yrs	Total colon + appendix	Constipation	Anti-mesenteric	Double bladder Hemivertebra	Distal fenestration; recovered
4.	1 mo	Double appendix	—	—	Double bladder Urethra & penis Hemivertebra	Doing well after multiple procedures for other anomalies
5.	1 mo	Double appendix	—	—	Cloacal extrophy Meningomyelocele	Died of aspiration
6.	2 wks	17 × 4	Obstruction Mass	Transverse colon	—	Resection; recovered
7.	3 mos	4 × 3	Mass	Cecum	—	Died of sepsis
8.	7 yrs	4	Mass	Cecum	—	Right colectomy; recovered
9.	3 days	3	None (autopsy)	Transverse colon	—	Incidental at autopsy
10.	7 mos	1	None (autopsy)	Hepatic flexure	—	Incidental at autopsy
11.	6 yrs	5	Obstipation Mass	Rectum	Hemivertebra	Excision; recovered
12.	1 day	5 × 3	None (autopsy)	Rectum	—	Died of congenital heart disease
13.	3 mos	8	Mass	Left flank Retroperitoneal	—	Excision; recovered
14.	4 mos	10	Mass	Left flank Retroperitoneal	—	Excision; recovered



FIG. 5. Cystogram in 15-year-old with total colonic duplication and septate bladder.

procedure which leaves a portion of a duplication in place is the malignant potential as reported by Orr and Edwards.¹⁵

Summary

Duplications of the alimentary tract are relatively rare. They usually become symptomatic early in life. The symptoms depend on the location of the lesion and the type of mucosal lining. Peptic complications may occur when ectopic gastric mucosa is present. The cause of duplication formation is unknown, but the results of surgical therapy are good except for the patients who have other anomalies which in themselves carry a high morbidity and mortality.

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