Duodenal Duplication: *

Case Report and Literature Review

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THIS PAPER will review 54 cases of duodenal duplication from the literature and report a case with a 4-year follow up. To our knowledge, a preoperative diagnosis of duodenal duplication has not been made by any previous authors (Table 1).

Duplication of the alimentary tract is a term suggested by Gross [21] to encompass enterocystomas, enterogenous cysts, enteric cysts, giant diverticula, ileum duplex and thoracic cysts of enteric origin. The first case of duodenal duplication was reported by Sanger [44] in 1880: a stillborn female found to have a duodenal duplication, a choledochol and three ileal duplications. Waugh [51] in 1923 performed the first operation on a 19-day-old girl presenting with vomiting and a palpable mass; a duplication of the second portion of the duodenum was aspirated and packed; a gastroenterostomy was made at a subsequent operation but the patient died. In 1927 Sohn [49] was successful in resecting a duodenal duplication in a 8-year-old child.

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Case Report

The patient (HUP #11-59-13) was born on July 27, 1957 in the Hospital of the University of Pennsylvania and was examined regularly in the Pediatric Clinic. On April 27, 1958 she was admitted to the hospital with a history of vomiting and constipation of 1 day's duration. A diagnosis of appendicitis was made; mesenteric adenitis was found at operation and the patient was discharged without sequelae. On March 21, 1961 she was readmitted with a palpable right upper quadrant mass. Intermittent peri-umbilical pain had been present for 1 week. There was no history of vomiting, fever or jaundice. An 8×12 cm. firm, cystic, movable mass was palpated and peristalsis in the mass was noted by the examiner. Roentgen examination of the chest was normal. In the upper gastrointestinal study a large mass was seen to displace the duodenum superiorly, posteriorly and laterally to the right (Fig. 1). In the cholecystogram the gallbladder was displaced superiorly by an extrinsic mass. This lesion was noted in an excretory urogram, but the kidneys and ureters were not displaced. A preoperative diagnosis of duplication of the duodenum was made.

At operation a cystic, spherical mass, 10 cm. in diameter, was found anterior to the first and second portions of the duodenum (Fig. 2). It had a thick muscular wall, had peristaltic activity, and contained clear, slightly viscid fluid with a pH of 8.0. No direct communication could be established between the cyst and the true duodenum or the stomach. A cystojejunostomy and a distal jejunojejunostomy were performed (Fig. 2). Biopsy of the wall of the mass was reported as duplication of the duodenum with duodenal mucosa.

The child was discharged and has been examined repeatedly in the Pediatric Clinic. A gastrointestinal study was performed in June, 1964,

TABLE 1. *Preoperative Diagnoses Reported by Previous Authors for Duodenal Duplication*

	Diagnosis	Author	Year
Group I, pyloric obstruction	Pylorospasm, pyloric obstruction, etiology?? Pyloric stenosis	Meyer[36]	1919
		Maddox[32]	1927
		Smith[48]	1930
Group II, tumors	Abdominal tumor, pyloric tumor, duodenal tumor	Waugh[51]	1923
		Bassman[5]	1938
		Booher & Pack[9]	1946
		Peple[40]	1948
		Lorber & Machella[31]	1948
		Mendl *et al.*[35]	1954
		DuBerger[16]	1957
	Leiomyoma of duodenum	Shallow *et al.*[47]	1947
		Kirtley & Matuska[27]	1957
	Polyp of dyodenum, duodenal filling defect, or deformity	Polson & Isaac[42]	1953
		Brooks & Weinstein[12]	1943
		Booher & Pack[9]	1946
	Retroperitoneal lymphoblastoma, renal tumor	Hicken *et al.*[22]	1948
		Jelenko[25]	1962
Group III, cysts	Abdominal cyst, mesenteric cyst	Pachman[39]	1939
		Ladd[28]	1940
		Orgias[38]	1943
		Annamunthodo[3]	1959
	Choledochol cyst	Gardner & Hart[19]	1935
Group IV, miscellaneous	Intussusception, appendicitis	Sohn[49]	1927
		Gordimer & Bluestone[20]	1950
		Anderson *et al.*[2]	1962
	Duodenal perforation into lesser sac, intestinal obstruction	McLetchie[34]	1954
		Pinkerton & Annamunthodo[41]	1955
	Shock with peritonitis, obstructive jaundice	Basu *et al.*[6]	1960
		Basu *et al.*[6]	1960

4 years following operation; the duplication (and its anastomosis with the jejunum) was still present but was much smaller and not palpable (Fig. 3). The patient is asymptomatic.

Theories on Development of Duodenal Duplication

Meckel's diverticulum has been considered a possible precursor of intestinal duplication. This has not been widely accepted since Meckel's diverticula are usually found in the antimesenteric border, in contrast to the usual mesenteric location of duplication.

Lewis and Thyng [30] described in 20 to 30 mm. embryos of the pig, rabbit and human, minute knob-like groups of epithelial cells derived from the alimentary tract. These occur along the course of the esophagus, stomach and small intestine, chiefly along the antimesenteric surface of the intestinal tract. These diverticula were more numerous in the older embryos and in the distal part of the small intestine and disappeared with growth. If persistent, the lesions either remained connected with the lumen by a narrow pedicle or survived as separate closed cysts. In some instances the cysts remained in the submucosa and by expanding, form an intraintestinal cyst. This theory suffers the same inconsistency as that in Meckel's diverticulum.

Bremer [10] proposed a theory which appears to explain best intra-abdominal duplications. During the sixth week of development, the intestinal tract undergoes a sudden rapid increase in length which is preceded by an increase in the number of

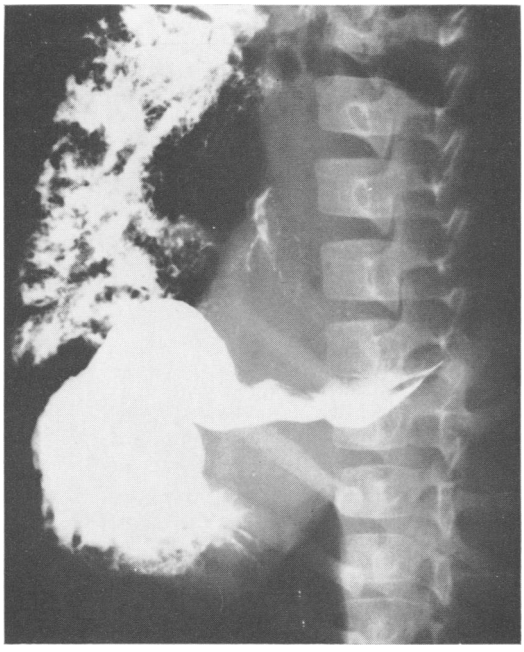

FIG. 1. Preoperative x-ray showing duplication of duodenum. Note mass displacing the duodenum superiorly, posteriorly, and laterally.

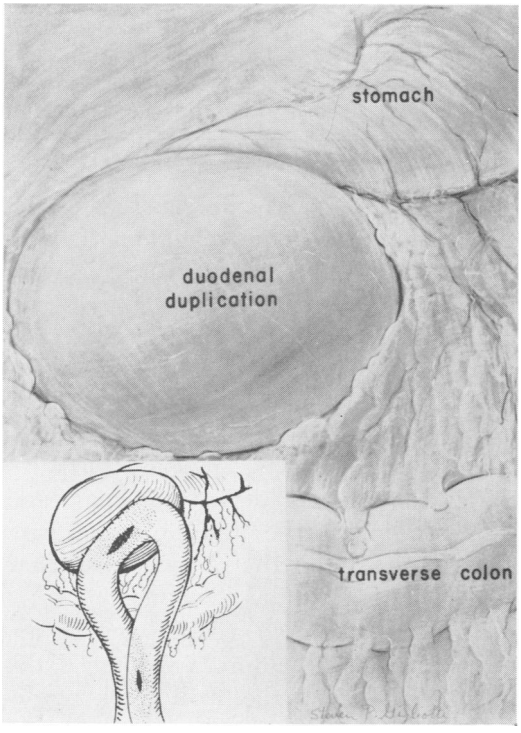

FIG. 2. Operative finding in duodenal duplication. Insert shows cystojejunostomy and enteroenterostomy.

epithelial cells, so that the lumen of the tube is markedly reduced or occluded. A transverse cleft forming across the mass of epithelial cells allows ingrowth of submucosa with or without muscle layers. Normally, epithelial cells secrete a fluid which results in formation of vacuoles which coalesce to form a lumen. Formation of a duplication results from failure of these vacuoles to coalesce into a single channel. The duplication may share a single mesentery and a single basement membrane or form a discrete tubular structure with its own walls and mesentery.

Veeneklaas,[50] in 1952, advanced the theory that best explains duplications found within the thoracic cavity. During the third week of embryonic life the notochord, which has its origin in cells from Hensen's node of the ectoderm, begins to form within the mesodermal mass. The notochordal process fuses with and becomes temporarily

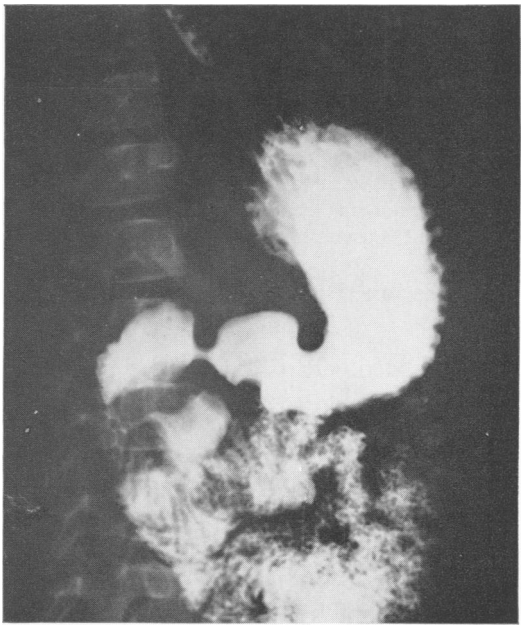

FIG. 3. Postoperative roentgenogram showing duplication of duodenum. Note that following enteroenterostomy, the duodenal duplication fills with barium from the jejunum. There is no connection with duodenal bulb or biliary system.

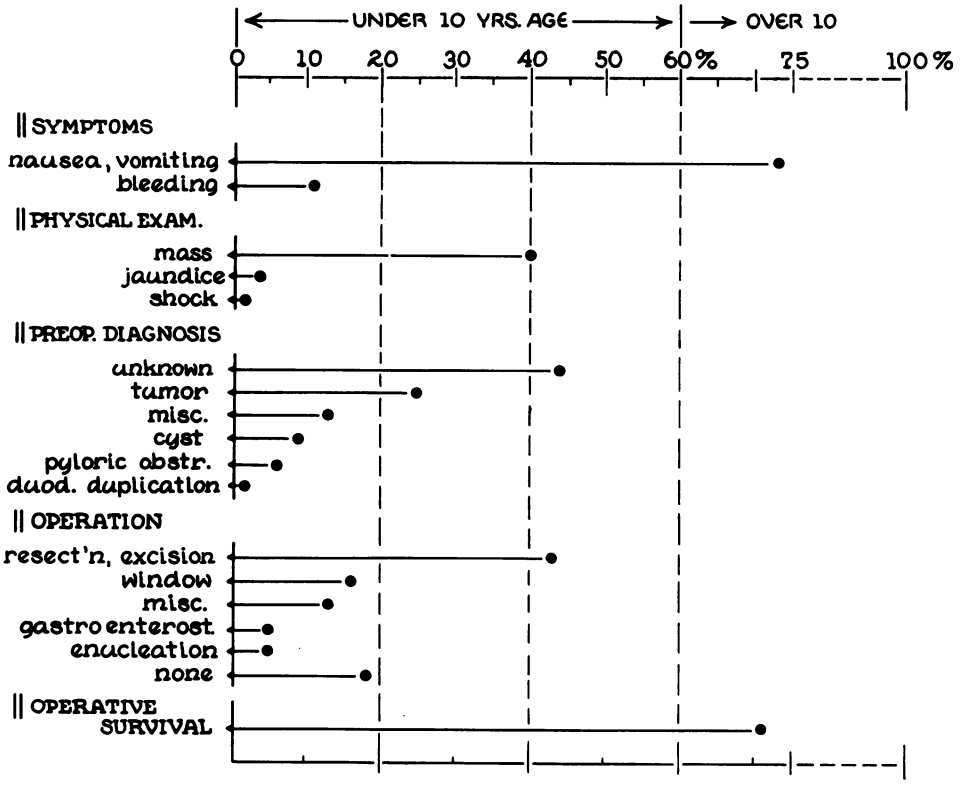

FIG. 4. Duodenal Duplication: a review of 55 case reports.

intercalated in the embryonic endoderm, and the notochordal plate is formed. By the fourth week the notochordal plate has separated from the endoderm and formed a vertebral mass around the notochordal remnants, producing the nucleus pulposus of the intervertebral discs. Adhesions between the ectoderm and entoderm may result in a cyst lined with epithelium of the intestinal tract or nervous system.[8]

As the embryo grows, the intestinal tract elongates and carries the cyst or tubular structure caudad; the cyst itself often remains in the posterior mediastinum, most often on the right side. A fibrous tract may or may not be traced down through the diaphragm into the peritoneal cavity. In a few instances, both thoracic and abdominal duplications have been reported in the same patient.[8]

The presence of a thoracic duplication is probably responsible for failure of fusion of the vertebral body. Tomograms may be necessary to determine the presence of these vertebral abnormalities.[50] In support of this theory, Saunders found that patients with anterior or posterior, or combined spina bifida, often demonstrated a diverticulum or fibrous strand projecting from the alimentary tract toward the vertebral cleft and spinal cord.[45]

Clinical Findings Associated with Duodenal Duplication (Fig. 4)

Age. Duplications are usually discovered during infancy and childhood. Eighteen patients were under the age of 4 months and 15 were less than 10 years of age.

Sex. There were an equal number of males and females.

Location and Operative Findings. Duodenal duplications, single or multiple, are

TABLE 2. *Surgical Treatment for Duodenal Duplication*

Treatment	Author
Aspiration	Waugh[51], Bassman.[5]
External drainage	Smith.[48]
Resection	Ladd,[28] Sohn,[49] Orgias,[38] Pinkerton,[41] Annamunthodo,[3] Basu *et al.*,[6] Hicken,[22] Chamberland *et al.*,[15] Leonard[28]
Window operation	Gardner & Hart[19], Lorber & Machella,[31] Gross *et al.*[21] Ploson & Isaac,[42] Broker & Hay,[11] Faegenburg & Bosniak,[18] Browning.[13]
Marsupialization	Pachman.[39]
Enucleation	Broker & Hay,[11] Peple,[40] Hicken.[22]
Stripping of mucosa	Hicken.[22]
Cystojejunostomy	Adams,[1] Inouye.
Gastrojejunostomy	Bassman,[5] Maddox.[32]
Biopsy of wall only	Browning.[13]

usually found along the first and second portions of the duodenum on its mesenteric side on the anterior wall; both submucosal and intermuscular sites have been described. The blood supply is intimately associated with that of the duodenum so that surgical dissection of the duplication from the duodenum is often impossible. Only the rare tubular duplications have been found to communicate with the lumen of the gastrointestinal tract. Communications with the biliary tract have been found, and calculi have been present.[6, 16, 17, 20, 42, 43]

Structure and Contents. Duodenal duplications are usually cystic structures, 2 to 12 cm. in diameter, lined by one or more layers of smooth muscle. They may have peristaltic activity. The mucosal lining, when not destroyed by pressure necrosis, may be identical to that found in the duodenum or resemble another portion of the gastrointestinal tract. In the cases reviewed, duodenal mucosa predominated; gastric, small intestinal, pancreatic, "columnar" and "flattened cells" were also present.[3, 18, 29] The cyst content has been described as mucilaginous, mucoid or watery and the color has been described as clear to reddish brown.

Associated Abnormalities. Numerous congenital defects have been reported in patients with duodenal duplication and include: double gallbladder,[6] congenital dislocation of the hip,[1] ventricular septal defect,[17] patent ductus arteriosus,[3] ileal and gastric duplications,[2, 7, 17] and mediastinal and esophageal cysts.[7, 8] Although vertebral abnormalities are found with 22 to 30 per cent of mediastinal duplications, they are not found with duplications limited to the intra-abdominal intestine. Cancers have been reported in duplications found elsewhere in the gastrointestinal tract, but none have been reported in duodenal duplications.[33, 37]

Symptoms. The most common symptoms were vomiting and gastrointestinal bleeding. Massive melena reported with duplication of the entire small intestine was not observed in these patients.[26] A palpable mass was recorded in 22 patients. One patient, in shock, had a perforation of the duodenal duplication lined with gastric mucosa.[6] Obstructive jaundice was noted in a patient with a duodenal duplication (possessing gastric mucosa) that had ulcerated into the common bile duct.[6] Biliary colic was observed in another patient with partial common bile duct obstruction due to extrinsic pressure by the duodenal duplication.[52] Acute pancreatitis with partial obstruction of the distal duodenum by the duplication has been reported.[12, 23]

Treatment

Surgical therapy is directed toward relief of symptoms such as obstruction and bleeding. Of 55 patients, 45 had surgical treatment and 39 survived.

Aspiration, marsupialization and external drainage do not offer surgical cure (Table 2). Although resection of the duplication alone or with the duodenum is fraught with

danger due to the common blood supply and contiguous structures, this has been successfully performed. The "window operation" first described by Gardner and Hart [19] in 1935 has been utilized by others; however, in retrospect, Gardner and Hart would have preferred a Roux-en-Y anastomosis to the original procedure.[40] Enucleation has been performed successfully in three instances. The mucosa was stripped by Hicken [22] in an attempt to prevent reaccumulation of cyst fluid. Cystojejunostomy, first reported by Adams,[1] was elected in our patient because of the disparity in size between the large duplication and the true duodenum, making it difficult to ascertain the exact site of the common wall. It should be noted that the danger of future peptic ulceration exists if the duplication should contain gastric mucosa.

Summary

The embryology, symptomatology, radiology, and surgical treatment of duodenal duplication have been discussed. Fifty-four cases from the literature were reviewed, and a new case diagnosed preoperatively is presented.

Radiologic Findings. Complete obstruction with gastric distention and absence of gas in the gut distal to the lesion may be observed on abdominal roentgenograms. When the duodenal duplication does not communicate with the intestine, only an abnormal contour or the uniform density of a mass may be seen. Hope [24] believes that if a single supine radiograph of the abdomen fails to show obstruction, introduction of a catheter and aspiration of the upper gastrointestinal tract with air replacement may make the diagnosis of high obstruction. If intestinal obstruction is present but the site is not determined, barium enema x-ray should be done and if no obstruction is then demonstrated, a gastrointestinal x-ray study using barium should be performed. Obstruction caused by narrowing of the duodenal lumen, together with displacement superiorly and laterally, may be noted. The duodenal loop may be widened.[4, 14, 46] The duodenal bulb may be dilated or it may be crescent-shaped due to the mass indenting the inferior surface of this segment.[4, 18] Caffey reports that conclusive radiologic diagnosis can be made in rare instances when the roughened mucosal relief of ectopic rugae are seen in the duplication; roentgen findings may be normal if the duplication is large.[14]

The barium x-ray study in adults may show an oval filling defect or a well defined marginal defect in the concavity of the first and second portions of the duodenum.

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