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Enteric Duplications in Children

– An Analysis of 6 Cases –

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This is an analysis of 6 patients with enteric duplications seen over an 8 year period at the Department of Pediatric Surgery, Dongsan Medical Center. They were all males but one. All duplications were cystic, and single except one. Locations of duplications were in the duodenum in one patient, in the jejunum in one, and in the terminal ileum in four. Five of the 6 patients were seen within 1 year of life. Three were newborn infants who had symptoms of intestinal obstruction with palpable mass since birth. Duplication cyst acted as a leading point of intussusception in 4 month and 8 month old infants respectively. One jejunal duplication was found in an 11-year-old boy who had malrotation of the midgut with Ladd's bands. Clinical presentation, embryogenesis of duplication, and management are discussed.

Key Words: *Duplication, Enteric duplication, Enterogenic cyst*

INTRODUCTION

Duplications of the gastrointestinal tract are rare congenital lesions of uncertain etiology. Clinical manifestations of enteric duplications are variable, and are determined by the type, site, and size of the duplication. Most commonly, they present in infancy or early childhood (Grosfeld et al.,1970; Gross and Holcomb, 1952), but late presentations have been reported (Gordimer and Bluestone, 1950; Polson and Issac, 1953; Thompson and Labow, 1967). Duplications of the small intestine are of two main types, cystic and tubular. They present as a simple abdominal mass with or without intestinal obstruction, and may act as a leading point for intussusception. Sometimes large cysts lead to intestinal obstruction simply by pressure on the adjacent bowel. Gastrointestinal bleeding may be a presenting feature, and is due to ectopic gastric tissue in the lining mucosa . This report reviews an 8 year experience with enteric duplications in 6 patients and discusses etiology and operative therapy.

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MATERIALS AND METHODS

A retrospective study of 6 patients with surgically and pathologically proven enteric duplications was undertaken by reviewing the hospital records at the Department of Pediatric Surgery, Dongsan Medical Center during the period between 1985 and 1993. The record analysis comprised of patient age, clinical manifestations, location, size, presence of ectopic tissue, presence of additional anomalies, and operation.

RESULTS

There were six patients during the past 8 years. All the patients were boys except one. The age at operation ranged from 2 days to 11 years. Five were seen within 1 year of life; three were admitted within the first 9 days of life and two were 4 month and 8 month old infants. Two of the three newborns were strongly suspected of having enteric cyst preoperatively by symptoms of intestinal obstruction with palpable abdominal mass but in a

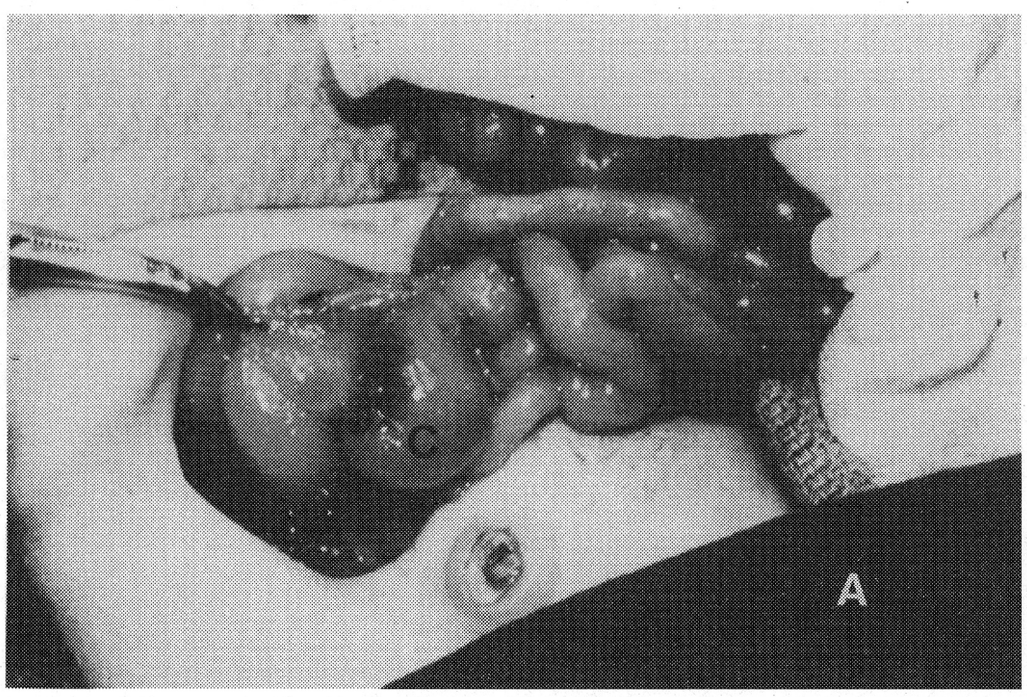
premature 2-day-old very low birth weight infant, the preoperative diagnosis was duodenal atresia with characteristic "double-bubble" appearance on abdominal x-rays. At laparotomy, an isolated duodenal duplication cyst situated between the dilated proximal and collapsed distal segments of duodenal atresia was found (Fig. 1). The two patients, 4 months and 8 months of age, were diagnosed to have intussusception. Intussusceptions were reduced successfully by barium enema but barium was retained in the small bowel with a filling defect in the terminal ileum near the ileocecal valve. The latter case had a history of barium reduction of intussusception 2 months previously.

As summarized in Table 1, these were all cystic duplications and all cysts were noncommunicating type except one (Fig. 2). They were located in the duodenum in one patient, in the proximal jejunum in another one, and in the terminal ileum near the ileocecal valve in the remaining four patients. The size of the duplications was variable from 1.3cm to 7cm. The lining epithelium was intestinal mucosa in four patients and gastric mucosa in the remaining two. One of two patients with gastric mucosa had the histologic appearance of peptic ulcer. In the other patient, who had recurrent intussusception, two separate duplication cysts (Fig. 3) were noted in the terminal ileum near the ileocecal valve and acted as leading points and each cyst was lined with gastric mucosa (Fig. 4). One jejunal du-

Table 1. Summary of 6 cases with enteric duplications

Case	1	2	3	4	5	6
Sex	F	M	M	M	M	M
Age	8 days	4 months	11 years	9 days	8 months	2 days
Clinical Manifestations	vomiting diarrhea abd. mass	vomiting diarrhea abd. distention	abd. pain	vomiting abd. distention abd. mass	vomiting abd. mass	vomiting
Symptom duration	2 days	3 days	5 days	9 days	1 day	1 day
Special study	US	BE	US	BE Tc^{99m}-scan	BE	abd. X-ray
Preoperative diagnosis	duplication	intussusception	mesenteric cyst	duplication	intussusception	duodenal atresia
Location	terminal ileum	terminal ileum	proximal jejunum	terminal ileum	terminal ileum	duodenum
Size (cm)	1.3×1.2	4.2×3.5	7×6	4×3	4×5 Ł 2×2	1.8×1.2
Lining mucosa	gastric mucosa	columnar epithelium	columnar epithelium	cuboidal epithelium	gastric mucosa	intestinal mucosa
Communication to bowel lumen	(+)	(−)	(−)	(−)	(−)	(−)
Surgery	ileoascending colectomy	segmental resection	segmental resection Ladd procedure	partial excision	ileoascending colectomy	segmental resection DJ stomy
Associated disease			malrotation			DA

* Abbreviations
US : Ultrasound BE : Barium enema
DJ stomy : Doudenojejunostomy
DA : Duodenal atresia



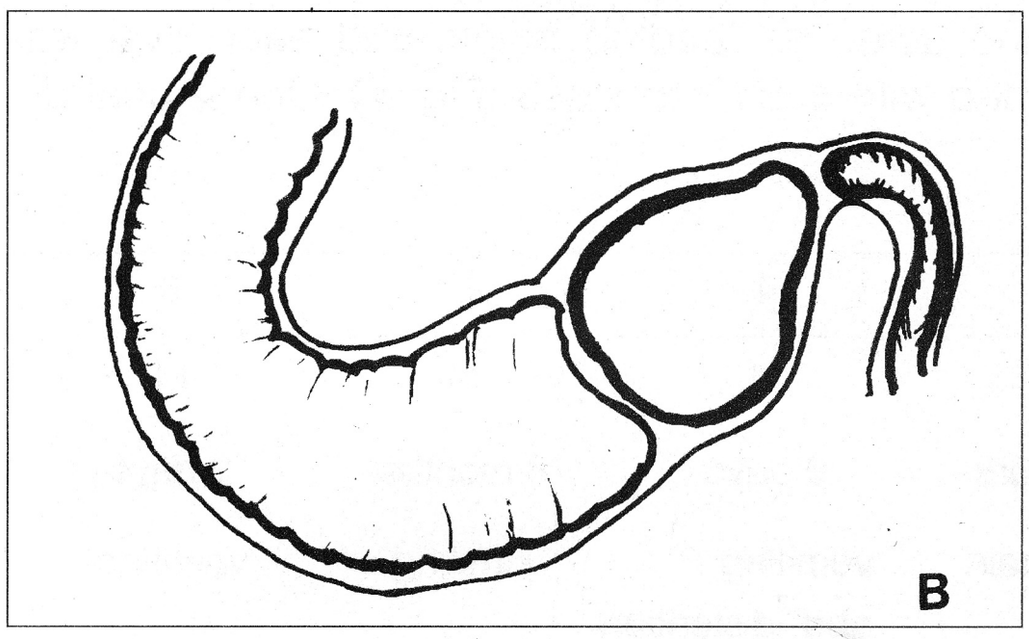


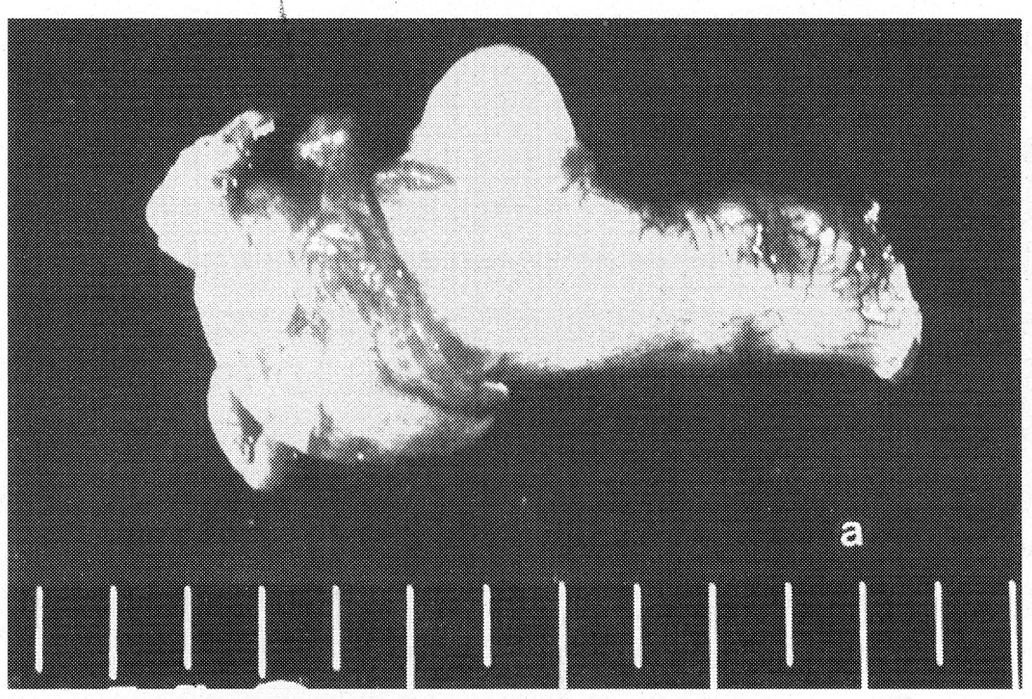
Fig. 1. Operative finding (A) of an isolated duodenal duplication cyst (C) partially dissected from the dilated blind end (indicated by clamp) and diagrammatic drawing (B). A cyst is situated between the dilated proximal and collapsed distal segments of the duodenal atresia without luminal communication in a premature 2-day-old very low birth weight infant.

plication, which was located in the proximal jejunum 15cm beyond the Ladd's bands crossing the duodenal second part to the cecum in an 11-year-old boy, was associated with malrotation.

The results of surgical treatment were good except for the premature infant (case 6) weighing 1350gm, who died shortly after who died shortly after postoperative recovery.

DISCUSSION

Duplications may be located anywhere along the gastrointestinal tract from the mouth to the anus. Those of the small intestine are the most common and the ileum is by far the most common location, with the jejunum the second and the duodenum the third in frequency (Bower, 1978; Houston and Lynn, 1966). Duplications are seen primarily in patients in their first year of life. In Gross and Holcomb's series (1952), 65% of patients were seen during this period. The age at time of



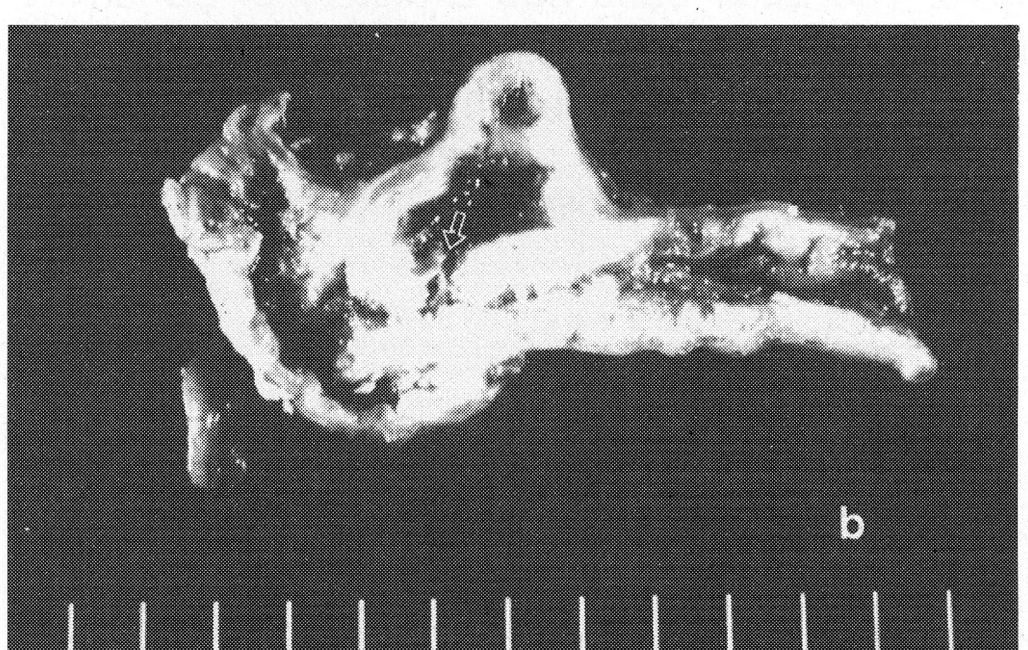


Fig. 2. Resected specimen of the terminal ileum and cecum of case 1(a) and cut section showing the communication indicated by white arrow(b).

detection and operation in our series ranged from 2 days to 11 years. Five of our 6 patients presented within the first year of life including the 3 newborn infants. However, there are occasional reports of a duplication found in older patients (Gordimer and Bluestone, 1950; Polson and Issac, 1953). Thompson and Labow (1967) reported a case of duodenal duplication in a 51-year-old man.

The clinical manifestations of enteric duplications are variable, and are determined by the type, site, and size of the duplication. They may present as an abdominal mass. They may lead to intestinal obstruction simply by pressure on the adjacent bowel or act as a leading point for intussusception, as in two of our 6 patients. Gastrointestinal bleeding may be a presenting feature in case of duplications lined by gastric mucosa (Jewett, 1958; Newmark et al., 1981; Waterston et al., 1980; Wilson et al., 1977). The ulcer is usually located in the duplication or in the adjacent small bowel at the site of their communication. In these cases a technetium scan is useful in localizing the site of bleeding. All of our patients presented with

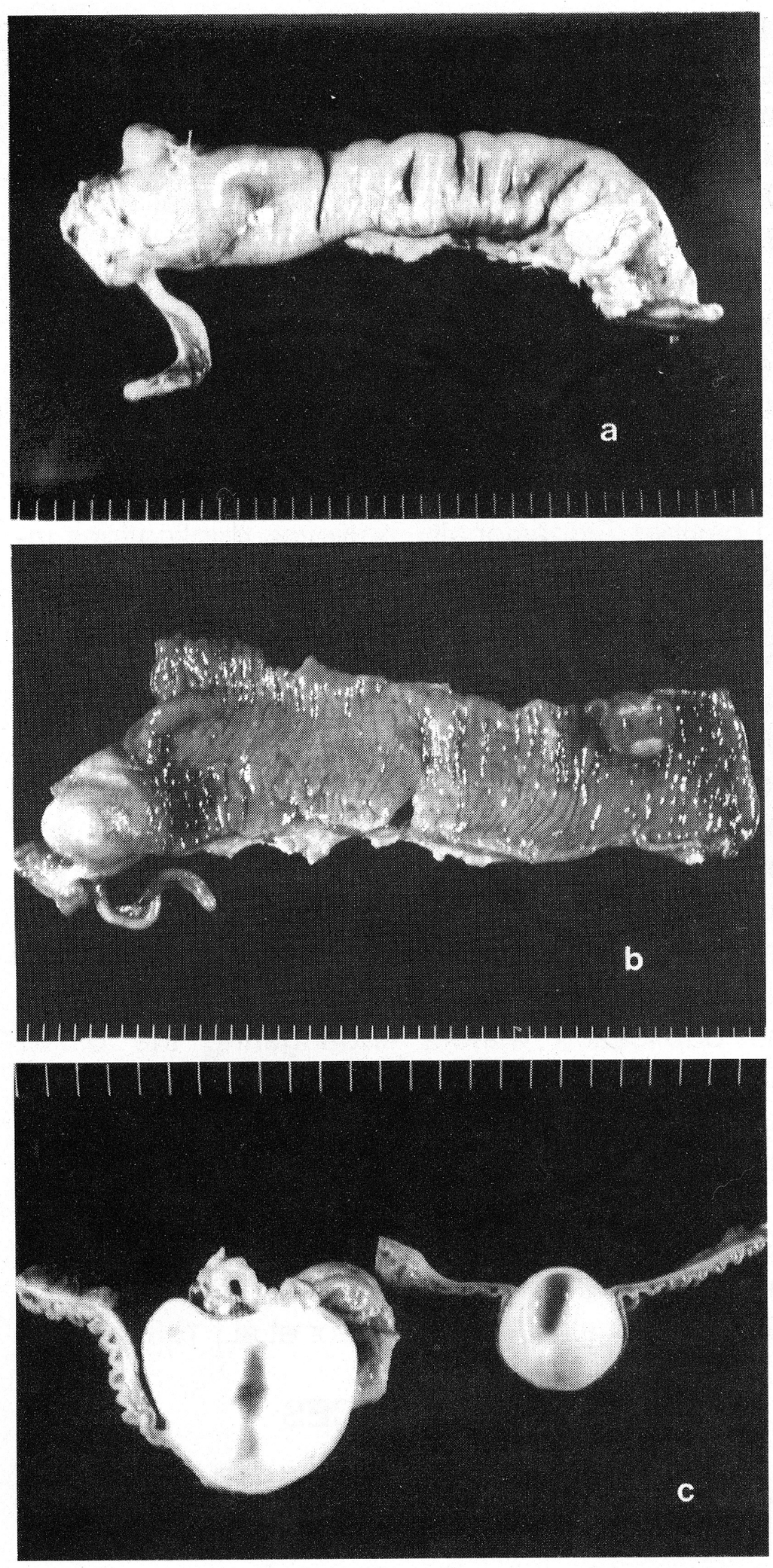


Fig. 3. Gross appearance of resected specimen(a and b) and cut section(c) showing two cysts. A distal duplication near the ileocecal valve acts as a leading point of recurrent intussusception in case 5.

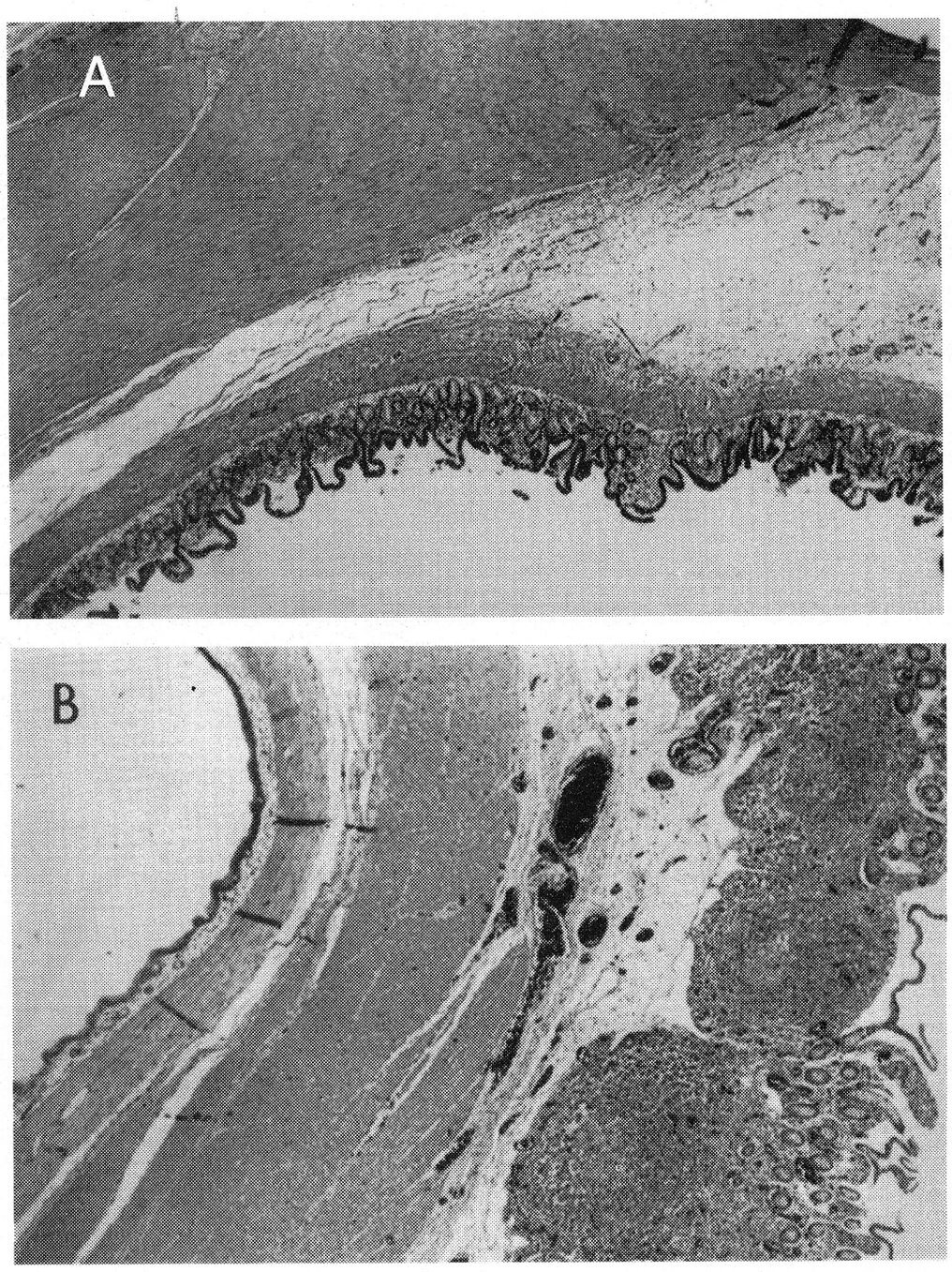


Fig. 4. Photomicrograph of the cystic duplication of case 5. The cyst is lined by gastric mucosal epithelium (A) and a partial common muscle layer between the terminal ileum and the cystic duplication is noted (B). (H & E, ×20).

intestinal obstruction and three of them had a palpable abdominal mass.

The diagnosis is difficult to make clinically due to variable symptoms and signs including occult bleeding, intestinal obstruction, abdominal mass, or abdominal pain. Conventional radiology is rarely helpful in aiding diagnosis. Sometimes a filling defect can be seen on barium meal and follow-through examinations, particulary in high jejunal lesions (Newmark et al., 1981; Royle and Doig, 1988). Ultrasonography may allow accurate diagnosis by identifying a double-layered wall, consisting of an inner echogenic layer and an outer sonolucent layer (Barr, 1990). Tc^{99m}-pertechnetate scan is also used for preoperative diagnosis of duplication especially in bleeding patients (Newmark et al.,1981; Royle and Doig, 1988; Waterston et al., 1980; Wilson et al., 1977). However, in more than half of the cases the duplication contains no gastric mucosa and the isotope scan will be negative. In general, investigations including barium enema and isotope scans are of limited value in the diagnosis of these lesions. The discovery may indeed be incidental at laparotomy for other conditions (Al-Salem and Khwaja, 1990).

Etiology is little known. Several mechanisms for their formation have been proposed: persistence of fetal enteric diverticula (Lewis and Thyng, 1908); failure of recanalization (Bremer, 1944); split notochord theory (Bently and Smith, 1960); and vascular insult theory (Favara et al., 1971) but none is supported by convincing evidence. The

most widely quoted theory is that of Bremer. Bremer(1944) thought that most of the spherical cysts were derived from the true diverticula, but a few of the spherical and most of the tubular duplications originated by an abnormal persistence of the vacuoles, which were normally present among the massed cells of the "solid stage of the intestine", in embryos of the sixth or seventh week. The same theory could not be applied to all enteric duplications. From the observations of enteric duplications in close proximity to the areas of intestinal atresia, ileal stenosis, and congenital short bowel, Favara et al (1971) proposed that intrauterine vascular occlusion may be the cause of certain enteric duplications in newborn infants. Mellish et al (1961) suggested that duplications are the result of some environmental stress in the early development of the fetus by observing the frequent association of the other anomalies in their series. In present study, the exact cause of enteric duplication was not demonstrated. We only strongly support Bremer's theory as a cause of duodenal duplication by experiencing an isolated duodenal duplication cyst, which was situated between the dilatated proximal and collapsed distal segments of the duodenal atresia without obvious change of serosal surface in a very low birth weight premature infant(case 6). Two of our series had associated gastrointestinal anomalies: duodenal duplication with duodenal atresia in a very low birth weight premature infant and malrotation with jejunal duplication in an 11-year-old-boy.

Duplications of the small intestine are of two main types, cystic and tubular. Occasionally multiple duplications (Gross et al., 1952; Bower et al., 1978; Buras et al., 1986) and coexistance of cystic and tubular duplications (Buras et al., 1986) occur in single individual. Cystic duplications seldom communicate with the bowel lumen and are lined by mucosa resembling that of the adjacent bowel. Ectopic gastric mucosa is more commonly associated with tubular duplications (Wrenn, 1962), although it may be found in cystic ones as well. 17-36% of small bowel duplications are lined with ectopic gastric mucosa (Gross et al., 1952; Bower et al., 1978). The Tc^{99m}-pertechnetate scan (Meckel's scan) is the most sensitive test for detection of ectopic gastric mucosa. The gastric mucosa lining coupled with the communication with the lumen of the small intestine frequently leads to peptic ulceration and bleeding. Bleeding, which may be either intermittent in small amounts or life threatening, is often the only clinical manifestation of tubular duplication (Wrenn, 1962). Perforation of the duplications have been reported in various sites of the small intestine by peptic ulceration (Dickinson et al., 1971; Rios-Dalenz et al., 1965; Royle and Doig; 1988). In present study, three ileal duplication cysts observed in 2 patients (one cyst in case 1 and 2 in case 5) were lined with gastric mucosa. One cyst lined by gastric mucosa (case 1) was communicated with the normal bowel and peptic ulcer was noted in ectopic gastric mucosa.

All duplications of the gastrointestinal tract should be excised, whether cystic or tubular type, if possible. Occasionally, however, duplications involving a long segment of the small intestine are difficult to manage. Jewett (1958) first suggested excision of the communication between the duplication and normal bowel with drainage of the duplication into the stomach. Wrenn(1962) first reported the use of a mucosal stripping procedure through multiple incision in the seromuscular layers of the duplication in an attempt to core out the entire mucosa of the duplication in a patient with a very long tubular duplication lined entirely by gastric mucosa. This technique effectively enucleates the duplication without endangering the blood supply or the function of the adjacent intestine. In cases of long duplication, the creation of an opening at both ends of the abnormal structure to form a double-barrel intestine has been used (Bower, 1978), or more rarely, the duplication can be separated from the normal bowel by careful dissection (Schwartz et al., 1980; Bar-Maor et al., 1985).

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