

PEDIATRIC SURGERY

Volume 2 Second Edition

PEDIATRIC SURGERY

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1. The abdominal cavity is divided into four quadrants by a vertical line passing through the midline and a horizontal line passing through the umbilicus. The quadrants are labeled as follows: RU (Right Upper), RL (Right Lower), LU (Left Upper), and LL (Left Lower).

2. The abdominal cavity is also divided into nine regions by two vertical lines and two horizontal lines. The regions are labeled as follows: RU (Right Upper), RL (Right Lower), LU (Left Upper), LL (Left Lower), and the central regions are labeled as RU, RL, LU, and LL.

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PART IV

Abdomen

SECTION FOUR

PLATE V

A. Chylous Cyst of the Mesentery. A 4-year-old boy had abdominal colic with vomiting, as well as a lifelong history of intermittent attacks of a similar nature. There was dullness in the upper abdomen, but no palpable mass. The involved segment had twisted, producing a volvulus, and this twisting had probably occurred intermittently. The chylous cyst, filled with a creamy material, bulges its thin lobulations through the mesentery of the small bowel (*center*), stretched out over the tumor. As in most mesenteric cysts, it was necessary to resect the bowel, together with the cyst, and perform an end-to-end anastomosis. Other cysts occasionally become manifest by the development of acute inflammations, suggesting appendicitis in their symptomatology.

C. Hirschsprung's Disease. Typical narrowing of the aganglionic rectosigmoid area with dilatation of the sigmoid were found at operation in a 3-week-old infant. A colostomy, performed in the sigmoid after frozen section, determined the presence of normal ganglion cells. Resection of the aganglionic segment and Swenson's procedure were successfully accomplished when the child weighed 30 pounds.

E. Intestinal Duplication. A large enteric cyst near the ileocecal junction in a 12-day-old child who had great abdominal distention. Serosa-covered muscle-walled and mucosa-lined cysts of this kind, when localized, as in this instance, usually do not communicate with the bowel. A common muscular coat between the bowel and the cyst usually requires resection of the attached bowel, as was performed in this instance, with an end-to-end anastomosis. Long tubular duplications frequently communicate with the bowel; and they are lined in whole, or in part, by gastric mucosa, so that junctional ulcers occur, presenting with massive hemorrhage. If resection of these duplications is precluded by the length of bowel involved, it is possible to strip the mucosal-submucosal lining from the duplication, leaving the innocuous muscle behind. Enteric cysts in the duodenum and other special locations require individual operative procedures.

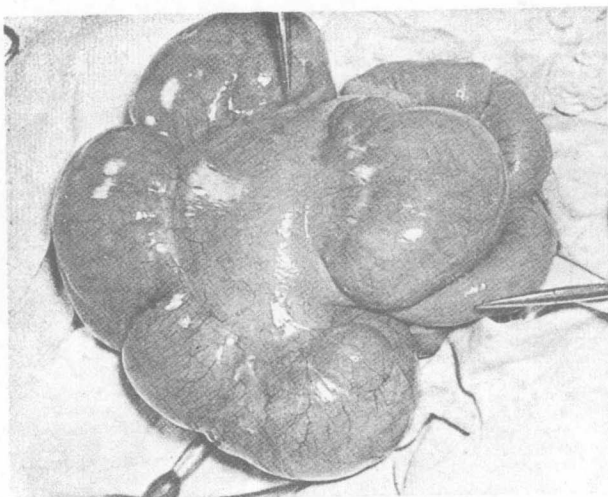
B. Jejunal Atresia. A 21-hour-old infant vomited bile, and there was an absence of meconium. A complicating volvulus of the small bowel had already been reduced. Death, without apparent relief of obstruction, may occur in the presence of a patent anastomosis, because of failure of the dilated proximal loop to empty. The enormous distention of the blind proximal loop requires resection of a good portion of this section and postoperative decompression by a fine catheter sewn into the loop and brought out through the abdominal wall.

D. Hirschsprung's Disease. Photomicrographs from a resected specimen in a child with a classic history of constipation and distention from birth and with typical operative findings. The left view, from the dilated hypertrophied proximal bowel, shows clearly the numerous large ganglion cells of the myenteric plexus of Auerbach and the inconspicuous nerve fibers. The right view, a section through the narrowed distal segment, shows a complete absence of ganglion cells and a great prominence of whorls of neurofibrillae. These are regular findings in the narrowed distal segment in Hirschsprung's disease. Distally, the aganglionic segment begins with the internal sphincter; and proximally it extends uninterruptedly until normal bowel is reached, usually in the sigmoid but occasionally in the more proximal areas of the colon, and, at times, well up into the small bowel. Occasionally the entire small intestine is involved, along with the colon. "Skip" areas are not reliably known to occur.

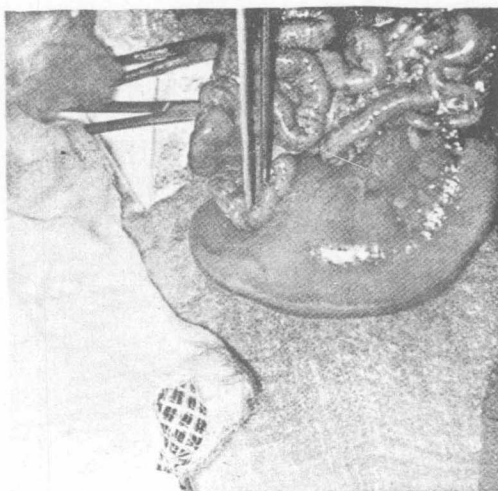
F. Chronic Ulcerative Colitis. The left view shows ulcerations and pseudopolypoid mucosal thickening. The right view demonstrates shortening and narrowing of the colon.

PLATE V

A



B



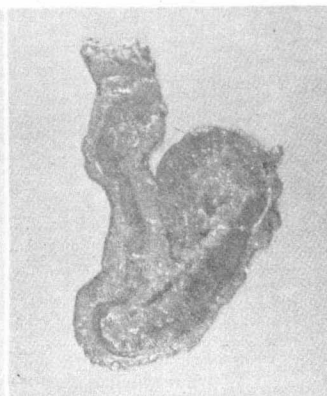
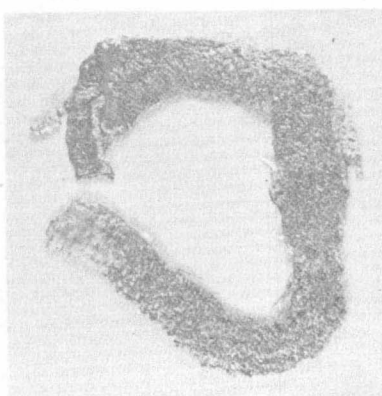
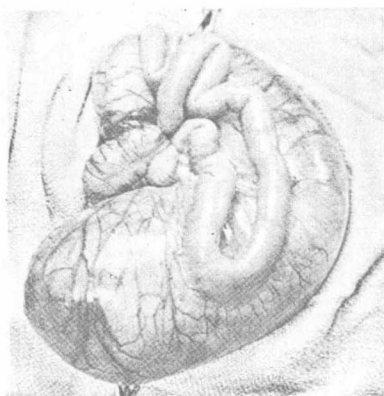
C



D



E



The Stomach and Duodenum

Prepyloric and Pyloric Obstruction

NEONATAL GASTRIC OUTLET OBSTRUCTION

Congenital gastric outlet obstruction is not common in the newborn period and has been recognized in only comparatively recent years. In 1937, Bennett¹ reported on an infant 4 days old who had been operated on for pyloric stenosis and died 36 hours later. Autopsy revealed the cause of obstruction to be a complete prepyloric diaphragm. In 1940, Touroff and Sussman¹⁴ successfully removed a complete prepyloric diaphragm in a 1-day-old infant. Metz *et al.*¹⁰ in 1951 reported on an infant 3 days old who had a double diaphragm which caused a cystlike structure between the two membranes. Incision of the diaphragms resulted in recovery. In 1951, Benson and Coury² reported the third successful case, and in 1959, Brown and Hertzler reported successful treatment of 2 premature infants 10 and 7 days old.

Incomplete prepyloric membrane in the newborn is rare. DeSpirito and Guthorn⁶ in 1957 reported the first case, in a 21-day-old infant, and in 1967, Cremin⁸ described a similar picture in 2 infants 3 and 4 days old. It is of great interest that Cremin made the diagnosis preoperatively by radiologic studies in 1 case and is the first to have pointed out the salient radiologic features.

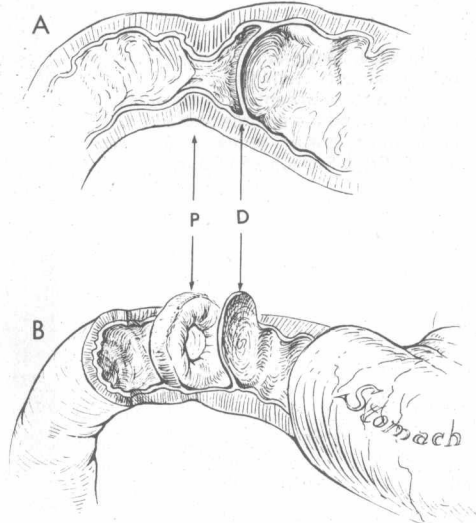
ETIOLOGY AND PATHOLOGY.—These obstructive lesions in the prepyloric and pyloric areas are of congenital origin and may have a vascular basis, such as infarction, similar in nature to the origin of jejunoileal and colonic atresia or stenosis. In about half the cases of congenital gastric outlet obstruction there will be a history of maternal hydramnios.⁹ Gerber and Aberdeen⁷ proposed a very plausible classification after an extensive review of the literature:

- I. Pyloric
 - A. Membrane
 - B. Atresia
- II. Antral (1 cm or more proximal to pylorus)
 - A. Membrane
 - B. Atresia

At operation, there may be no external evidence of abnormality at the gastroduodenal junction, and the obstruction can only be detected on gastrotomy (Fig. 52-1). This is especially true of the diaphragmatic obstructions. The complete atresia usually can be recognized from a fibrous cord which connects the proximal and distal portions of the stomach across the atresia, although not in all cases (Fig. 52-2).

The newborn with complete gastric outlet obstruction due either to a complete diaphragm or to a segmented atresia will vomit only gastric contents. In

Fig. 52-1.—Drawings showing, A, prepyloric membrane B, antral diaphragm. P=pylorus; D=diaphragm.



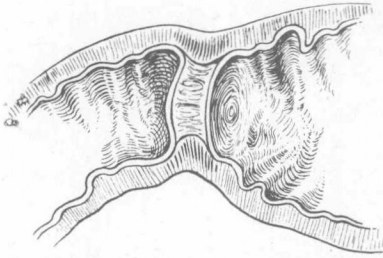


Fig. 52-2.—Pyloric atresia: drawing of a case. Note that the seromuscular layers are intact. The tissue between the gastric and duodenal mucosa was of two types, fibrous and areolar. Gastroduodenostomy in end-to-end fashion was successful.

addition, he may be dyspneic, cyanotic and have excessive salivation. The last is a definite hazard to these infants. Distention, when present, is confined to the upper abdomen. A normal meconium stool is passed soon after birth. The symptoms occur when an incomplete diaphragm is present because the aperture in the diaphragm is so small.

DIAGNOSIS.—The diagnosis can be made preoperatively if the possibility of such a lesion is entertained by the examiner. Roentgen study is of great aid because the gas pattern is limited to the stomach in the case of complete obstruction (Fig. 52-3).

Talwalker¹¹ has emphasized the difficulty of diagnosis of true pyloric atresia at laparotomy and makes the significant point that it is always worth while to make an opening in the distended proximal bowel in cases of neonatal obstruction.

TREATMENT.—In the newborn with a complete diaphragm or a diaphragm with an aperture, local excision or incision of the diaphragm with a Heineke-Mikulicz pyloroplasty is effective. In infants who

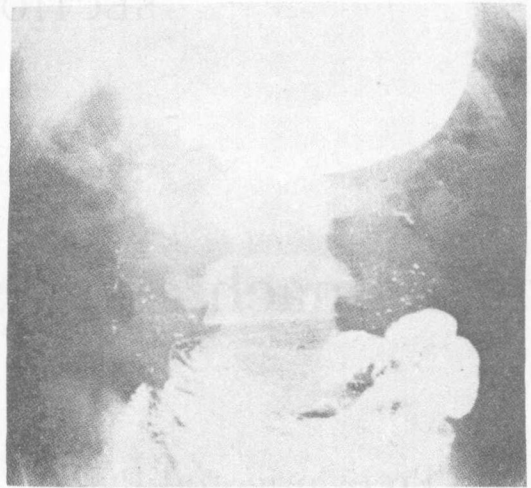
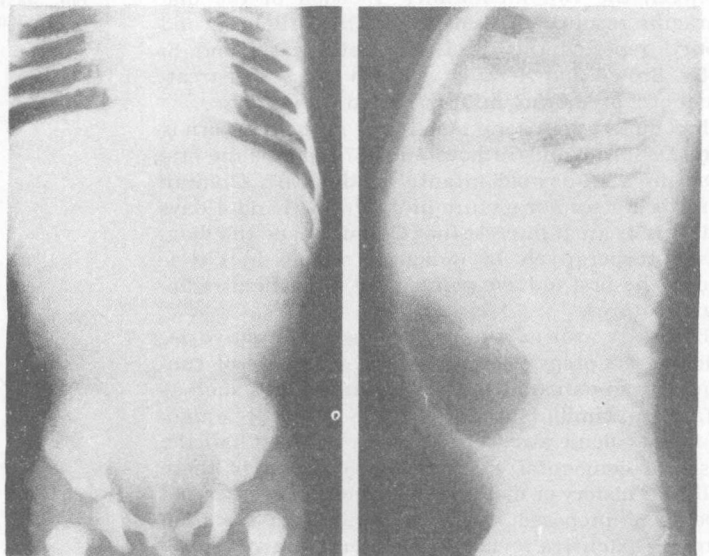


Fig. 52-4.—Partial-prepyloric diaphragm. Upper gastrointestinal roentgenogram shows some narrowing at the prepyloric antrum with gastric retention at 4 hours. The patient, a boy of 11, had had intermittent vomiting since age 18 months. Excision of the incomplete mucosal diaphragm completely relieved symptoms.

have a segmental atresia, the atretic segment is resected along with the diaphragmatic mucous membrane pouch and an end-to-end gastroduodenostomy performed, closing the anterior wall as in a Heineke-Mikulicz pyloroplasty.

PROGNOSIS.—With early diagnosis and surgical treatment, the prognosis is excellent. Of 19 infants reported on in the earlier literature, 14 survived. To these can be added the 1967 experience, each case with successful outcome: the 2 with incomplete pre-

Fig. 52-3.—Prepyloric atresia in a newborn. Plain and lateral views of the abdomen show distention of the stomach, no gas beyond the pylorus and absence of gas in any portion of the small or large bowel. (From Benson and Coury.²)



pyloric diaphragm operated on by Cremin,⁵ and 4 with pyloric atresia—2 newborns operated on by Thompson,¹³ 1 by Talwalker¹² and a 2-day-old infant by Benson. This makes a total of 25 patients treated surgically in the newborn period, with 20 recoveries.

INCOMPLETE PYLORIC-PREPYLORIC DIAPHRAGM IN INFANTS AND CHILDREN

This is a quite rare lesion, especially in the infant and child. The degree of obstruction is related to the size of the aperture in the diaphragm. Such lesions have been reported in a 5-week-old infant,¹⁵ in a 2-year-old,³ in another 2-year-old,⁷ in a 4-year-old,⁸ and I have operated on 1 patient of 11 years (Fig. 52-4). All recovered. They were managed by excision of the diaphragm or excision of the diaphragm and a pyloroplasty of the Heineke-Mikulicz type.

REFERENCES

1. Bennett, R. J., Jr.: Atresia of pylorus, *Am. J. Digest. Dis.* 4:44, 1937.
2. Benson, C. D., and Coury, J. J.: Congenital intrinsic obstruction of the stomach and duodenum in the newborn, *Arch. Surg.* 62:856, 1951.
3. Berman, J. K., and Ballenger, F.: Prepyloric membranous obstruction, *Quart. Bull. Indiana Univ. M. Center* 4:14, 1948.
4. Brown, R. P., and Hertzler, J. H.: Congenital prepyloric gastric atresia, *Am. J. Dis. Child.* 97:857, 1959.
5. Cremin, B. J.: Neonatal pre-pyloric membrane, *South African M. J.* 41:1076, 1967.
6. DeSpirito, A. J., and Guthorn, P. J.: Recovery from meconium peritonitis associated with diaphragm-like obstruction of the prepyloric mucosa, *J. Pediat.* 50:599, 1957.
7. Gerber, B. C., and Aberdeen, S. D.: Prepyloric diaphragm: An unusual abnormality, *Arch. Surg.* 90:472, 1965.
8. Liechti, R. E.; Mikkelsen, W. P., and Snyder, W. H., Jr.: Prepyloric stenosis caused by congenital squamous epithelial diaphragm—Resultant infantilism, *Surgery* 53:670, 1963.
9. Lloyd, J. R., and Clatworthy, H. W., Jr.: Hydramnios as an aid to the early diagnosis of congenital obstruction of the alimentary tract: A study of the maternal and fetal factors, *Pediatrics* 21:903, 1958.
10. Metz, A. R.; Householder, R., and DePree, J. F.: Obstruction of the stomach due to congenital double septum with cyst formation, *Tr. West. S. A.* 50:242, 1951.
11. Salzberg, A. M., and Collins, R. E.: Congenital pyloric atresia, *Arch. Surg.* 80:501, 1960.
12. Talwalker, V. C.: Pyloric atresia: A case report, *J. Pediat. Surg.* 2:458, 1967.
13. Thompson, N. W.: Personal communication.
14. Touroff, A. S. W., and Sussman, R. M.: Congenital prepyloric membranous obstruction in a premature infant, *Surgery* 8:739, 1940.
15. Wurtenberger, H.: Gastric atresia, *Arch. Dis. Childhood* 36:161, 1961.

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Drawings by

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Gastrointestinal Perforations in the Newborn

HISTORY.—Spontaneous perforation of the stomach in the newborn was first reported by Siebold in 1825.¹⁰ Over 100 years later, Stern *et al.*¹¹ reported one of the earliest attempts at operative intervention for this problem, but it was not until 1950 that closure of a neonatal gastric perforation was carried out successfully.⁶ Thereafter, reports of infants surviving surgical repair of perforations of the gastrointestinal tract appeared in the literature with increasing frequency. In 1964, Lloyd *et al.*⁷ found 132 cases of gastric perforation reported in the literature and added 31 from the Children's Hospital of Michigan, making a total of 163 cases, with 43 survivors. Since 1964, 4 more neonates have been operated on for gastric perforations at the Children's Hospital of Michigan. Fourteen of the 35 infants in our series have survived.

ETIOLOGY AND PATHOLOGY.—This discussion is concerned with perforations of the gastrointestinal tract unrelated to such well-documented causes as peptic ulcerations, trauma due to intubation, obstruction distal to the perforation and accidental gastric insufflation. In spite of numerous theories to explain these perforations, such as congenital muscular defects,³ hypophyseal-adrenal axis stress phenomenon⁵

and increased gastric acidity in the newborn,⁸ the etiology is still shrouded in mystery.

After a review of the clinical records of 61 infants with so-called spontaneous gastrointestinal perforations at the Children's Hospital of Michigan (stomach 31, duodenum 5, jejunum 1, ileum 7, colon 11, indeterminate 6), two important facts became apparent: (1) A large percentage of these patients had clinical evidence of shock, hypoxia or stress. (2) Postmortem studies of 39 of the 43 infants who died disclosed evidence of ischemic necrosis of the gastrointestinal tract not only at the sites of perforation but also in areas of the bowel remote from the perforation. There was a remarkable similarity in the histopathologic appearance of these ischemic lesions regardless of their location in the alimentary tract. In addition, other organs (brain, adrenals, liver, kidneys and skin) showed histologic evidence of asphyxia.

A detailed study disclosed maternal complications predisposing to compromise of 67.2% of the infants in utero. There were 4 infants with fetal distress, and these 4 plus 24 others had a poor Apgar rating at birth. Of 33 infants whose condition at birth was reported as fair or good, 15 had a cyanotic or asphyxial episode in the neonatal period. Prematurity may also