PEDIATRIC SURGERY

Volume 2 Second Edition

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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, Bennett[1] 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 Sussman[14] 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 Coury[2] 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 Guthorn[6] in 1957 reported the first case, in a 21-day-old infant, and in 1967, Cremin[8] 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 Aberdeen[7] 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

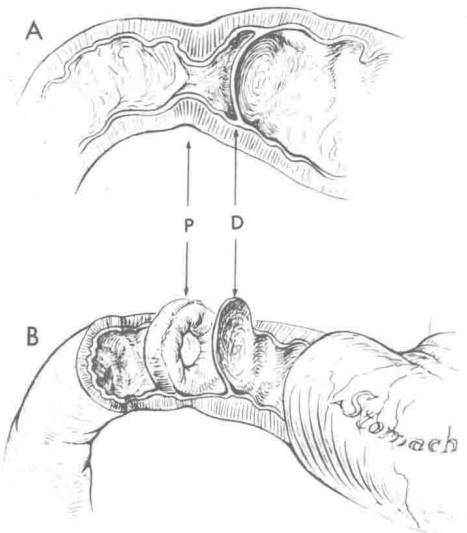

Fig. 52-1.—Drawings showing, **A,** prepyloric membrane **B,** antral diaphragm. **P**=pylorus; **D**=diaphragm.

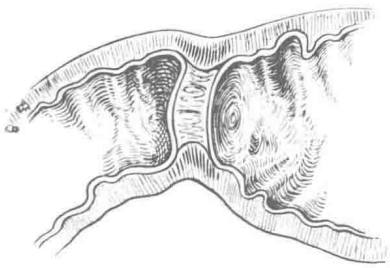

Fig. 52-2. – Pyloric atresia: drawing of a case. Note that the seromuscular layers are intact. The tissue between the gastric and duodenal mucosa was of two types, fibrous and areolar. Gastroduodenostomy in end-to-end fashion was successful.

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).

Talwalker[11] 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 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.

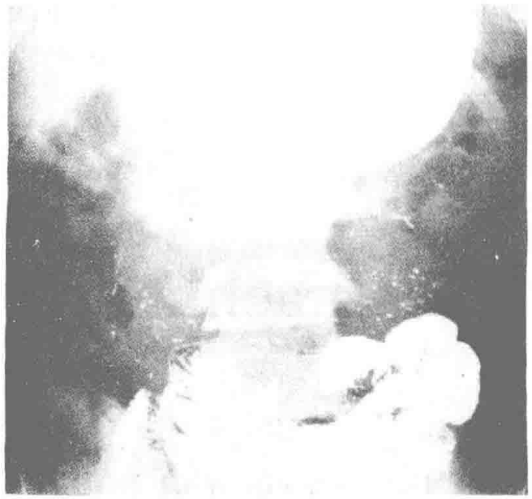

Fig. 52-4. – Partial prepyloric diaphragm. Upper gastrointestinal roentgenogram shows some narrowing at the prepyloric antrum with gastric retention at 4 hours. The patient, a boy of 11, had had intermittent vomiting since age 18 months. Excision of the incomplete mucosal diaphragm completely relieved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

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-

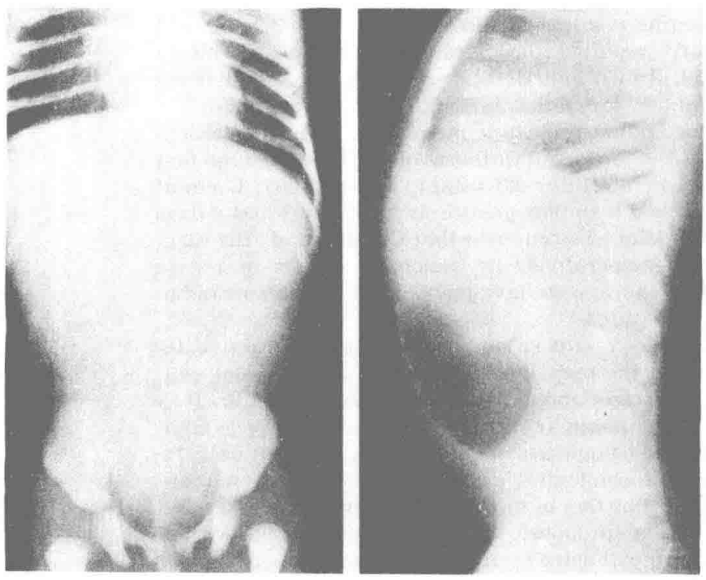

Fig. 52-3. – Prepyloric atresia in a newborn. Plain and lateral views of the abdomen show distention of the stomach, no gas beyond the pylorus and absence of gas in any portion of the small or large bowel. (From Benson and Coury.[2])

pyloric diaphragm operated on by Cremin,[5] and 4 with pyloric atresia – 2 newborns operated on by Thompson,[13] 1 by Talwalker[12] 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.

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C. D. Benson

Drawings by

Robert Mohr

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 phenomenon[5] 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

be a significant factor: 33 of the infants in this series were born prematurely, and 20 patients weighed less than 4 lb. Of the 61 patients, 37 (60.6%) experienced definite asphyxia, and the records of 19 babies strongly suggested an episode of shock stress or hypoxia. In only 5 infants was there insufficient evidence in the clinical record to indicate asphyxia.

These findings, correlated with the work of Scholander[9] and Elsner[2] and Corday[1] and their associates unequivocally implicate ischemia as the primary etiologic factor not only of neonatal gastrointestinal perforations but of all so-called spontaneous perforations regardless of the patients' age. Scholander and Elsner have studied diving mammals and birds and demonstrated an asphyxial defense mechanism that shunts blood away from areas of the body which tolerate prolonged ischemia (mesenteric, renal and peripheral circulations) to those areas which suffer irreversible damage when allowed to become ischemic for comparatively short periods (heart, brain and lungs). Corday *et al.*[1] demonstrated that intestinal ischemia can be induced by remote circulatory disturbances and called attention to "a renewed awareness of a syndrome characterized by hemorrhagic and/or necrotic lesions of the gastrointestinal tract for which there is no apparent anatomical explanation." "These changes . . . may follow such remote systemic disturbances as myocardial infarction, shock states, arrhythemias, heart failure, extensive surgery and severe burns." The "asphyxial defense mechanism" of Scholander and the "circulatory disturbance syndrome" of Corday are very closely related if not identical physiologic mechanisms of selective circulatory ischemia. Although James[4] demonstrated clinical evidence to support the contention that this mechanism functions in the perinatal period, there is indirect evidence as well. There is a period of time during birth before spontaneous respirations are established during which the infant is deprived, if not entirely, almost entirely of the umbilical circulation for varying periods, in many instances sufficiently long to produce irreversible damage to the heart and brain were it not for this asphyxial defense mechanism. In the past, the apparent lack of anoxic injury to the infant has been loosely ascribed to the traditional belief that the brain of the newborn can tolerate long periods of hypoxia. At times, the accumulated minutes of anoxia are far in excess of the known limits of physiologic tolerance.

Whether the ischemic lesions of the gastrointestinal tract that develop are due to such anatomic abnormalities of the mesenteric blood supply, prolonged local arteriolar spasm in the bowel wall and platelet thrombi is not clear, but areas of ischemia related to some circulatory disturbance occur and, if sufficiently severe, may involve the entire thickness of the gastric or intestinal wall and go on to perforation.

SYMPTOMS AND SIGNS.—The average age for onset of symptoms is the fourth day of life. In our series, symptoms occurred on an average of 2.6 days (range, 12 hours to 5 days) after an episode of asphyxia. Perforations in some instances were apparently present at birth or discovered soon afterward, ranging from birth to 18 days, or an average of 3.8 days after birth.

Initially, there is abrupt abdominal distention which may become so severe as to produce marked respiratory distress. The infant appears listless, dusky and frequently vomits bile-stained material. X-ray examinations are prompted by either the respiratory distress or the obstructive symptoms. In a few of our patients, a clinical diagnosis of perforation was entertained and confirmed by aspiration, but most commonly the diagnosis is established by the radiologic demonstration of free intraperitoneal air on either the upright abdominal films (Fig. 52-5) or the chest films.

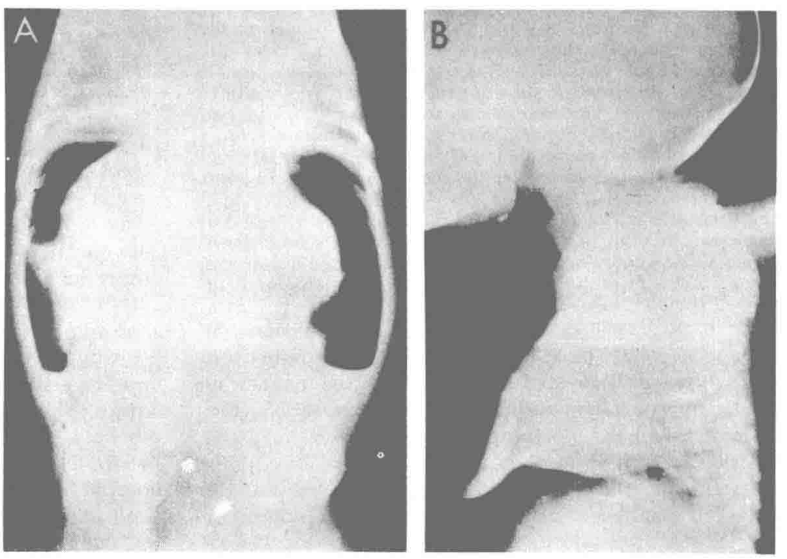

Fig. 52-5.—Spontaneous perforation of the stomach. Plain upright posteroanterior **(A)** and lateral **(B)** films of the abdomen, showing massive pneumoperitoneum.

Several criteria have been reported for recognizing free intraperitoneal air on the flat film, but this is risky at best and upright films should always be obtained. These infants may also show evidence of hypovolemic shock and/or sepsis. Large quantities of air may be released from a gastrointestinal perforation at any level, although massive pneumoperitoneum is usually associated with gastroduodenal perforations. On the other hand, we were unable to demonstrate a significant correlation between the site of perforation and the amount of free air in our patients. The lesion may occur anywhere along the gastrointestinal tract, and the surgeon should be prepared to cope with the lesion regardless of its location.

TREATMENT.—*Preoperative preparation.*—Early recognition and prompt treatment are essential to the survival of these infants. If the abdomen is so tightly distended that there is severe respiratory distress or if inferior vena caval return is compromised, aspiration of the peritoneal cavity may be life-saving. Nasogastric intubation should be instituted and a venous cutdown established. Administration of electrolyte solutions should be started to correct dehydration, and, when there is significant shock, 10 ml/lb. of Ringer's lactate should be given. Blood is made available, antibiotics and vitamin K are given intravenously, and the infant is carefully protected against hypothermia.

Operation.—The operating table should be equipped with a warm-water mattress and rectal temperature continuously monitored. General anesthesia is preferred, but if the situation demands its use, local anesthesia is satisfactory. The abdomen is opened through a right upper quadrant transverse rectus transecting incision placed about 2 cm above the umbilicus. When necessary for greater exposure, the incision can be carried across the left rectus. The entire abdominal portion of the gastrointestinal tract of the infant is readily accessible through this incision. The nature of the peritoneal contamination encountered sometimes suggests the location of the lesion; for instance, milk curds are indicative of a gastric lesion, while pure meconium favors a colonic lesion. Less commonly, there may be a large walled-off abscess in neglected cases with no free air or fluid. Material for cultures is obtained and the abdominal cavity is irrigated with warmed normal saline to wash out all contaminants. The site of perforation may be readily apparent, but when not, a routine search is conducted, starting with the exposed surfaces of the stomach. The entire small and large bowel are inspected, and if the lesion is not located, the lesser omental sac is entered and the posterior wall of the stomach carefully explored. In 6 of our infants, the site was not identified. Two of them had small amounts of air which gradually disappeared on repeated x-ray examination. They were treated conservatively and survived. The site of perforation was not found in the other 4 on surgical exploration. Two of these infants survived. Discrete perforations are closed with two layers of fine silk. Any devitalized tissue around the perforation is excised and a two-layer closure accomplished. Extensive lesions of the intestine are best treated by resection and end-to-end anastomosis. Exteriorization procedures are usually not indicated unless there is a suspected obstruction distal to the lesion, such as a possible aganglionic segment of distal colon. The abdomen is closed without drainage and the infant transferred to a prewarmed incubator.

Although a few patients have survived gastrointestinal perforations with conservative management, this course is generally not advised. When employed, the infant must be watched almost constantly. Should there be a progression of the pneumoperitoneum or evidence of deterioration, surgical intervention is mandatory. It should also be emphasized that neither the degree of peritoneal contamination nor the size of the perforation is necessarily related to the amount of free air present in the peritoneal cavity.

POSTOPERATIVE MANAGEMENT.—Most of these infants will have severe peritonitis and present problems of hypovolemic or septic shock in the postoperative period. Antibiotics are given intravenously, with type and dosage dictated by the results of sensitivity studies based on cultures of material obtained at the time of surgery. Nasogastric suction is continued until adequate gastrointestinal function is restored, after which oral feedings are cautiously resumed.

PROGNOSIS.—The survival figures for infants with gastrointestinal perforations continue to be discouragingly poor. Of 301 patients with gastrointestinal perforations (literature plus Children's Hospital of Michigan series), only 75 survived. As with the majority of lesions that result in contamination of the peritoneal cavity, the interval between the onset of symptoms and definitive treatment as well as the degree of soilage significantly influence the outcome of management. The degree of prematurity and the prolonged hypoxia that some of these infants endure are additional factors that may explain the unexpected death of those patients who are expertly and expeditiously managed.

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Duodenal Obstruction: Atresia, Stenosis and Annular Pancreas

HISTORY.—Congenital obstruction of the duodenum was first reported by Calder[5] in 1733, and the first successful operation for its relief by Ernst[7] in 1916. Benson and Coury[4] could find reports of only 57 survivors of this procedure during the next 36 years.[6] Between 1952 and 1960, the number of reported survivors more than doubled, and undoubtedly a great number of successfully treated cases have not been reported, including our own.

TERMINOLOGY.—The term atresia is used here to denote a complete obliteration of the intestinal lumen. Stenosis denotes a partial or incomplete obstruction of the lumen.

It has been customary to classify these obstructions as intrinsic, including any internal diaphragm, anatomic variant or embryologic arrest of maturation, and extrinsic, due to outside forces compressing the bowel wall.

PATHOLOGIC ANATOMY

Intrinsic anomalies (Fig. 52-6) occur in several forms: (1) The duodenum may end in a blind dis-

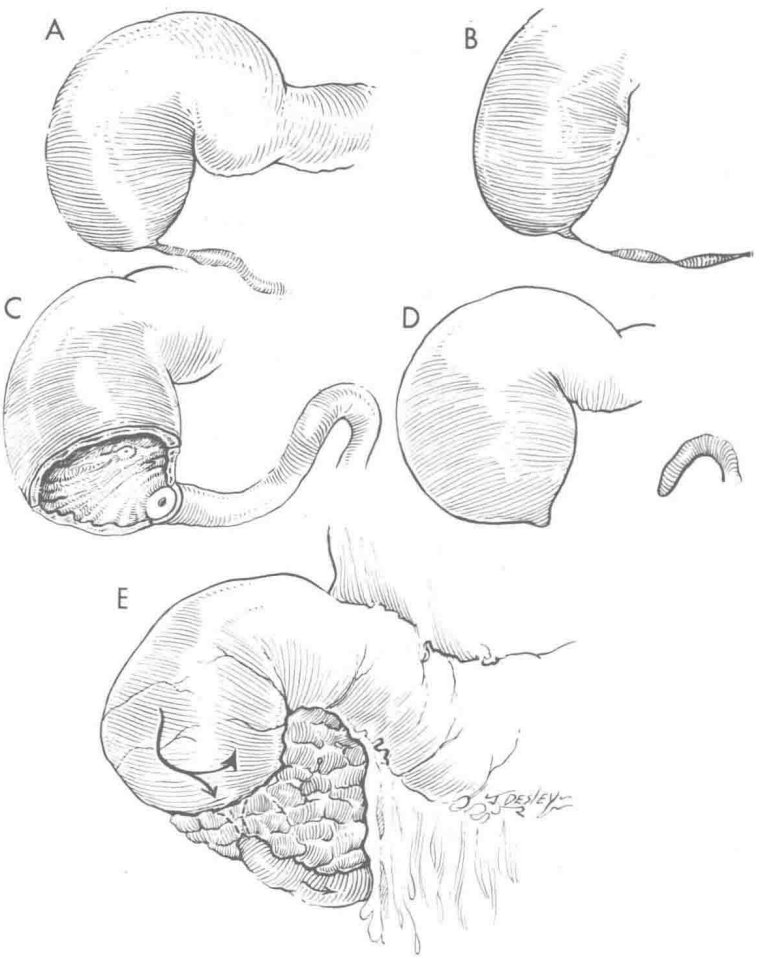

Fig. 52-6.—Intrinsic duodenal obstruction. **A,** atresia with continuity of bowel wall. **B,** atresia with fibrous cord joining segments. **C,** stenosis, internal diaphragm. **D,** atresia, loss of continuity of wall and blood supply. **E,** annular pancreas causing extrinsic pressure.

tended pouch with undilated bowel distal to the point of atresia. (2) A dilated blind end with a fibrous cord running to distal, undilated bowel may be present. (3) There may be a relatively complete diaphragm with only a small fenestration, or (4) a membranous ring within the duodenum with little effect on the lumen other than loss of distensibility. (5) There may be no evidence of any continuity, the proximal bowel lying completely free of any distal segment.

In extrinsic obstruction (Fig. 52-7), there may be continuity of bowel with only a modest compression of the duodenal wall by peritoneal bands, anomalous vessels or organs. Annular pancreas has become almost a household word in surgical circles and represents the best-known form of external pressure on the descending duodenum. Embryologically, the annular constriction is due to a persistence of a portion of the ventral anlage[2] which maintained its ventral location while being drawn around the right side of the duodenum to fuse with the main body of the pancreas. In this type of developmental arrest, it is possible to have either complete or incomplete encircling of the second portion of the duodenum. The degree of obstruction would depend on the firmness and completeness of the ring of tissue. Grossly, this pancreatic tissue is indistinguishable from the normally placed gland.

The other familiar form of extrinsic pressure obstruction is that due to the peritoneal folds or bands associated with incomplete rotation of the midgut. In this lesion, the weight of the distended ascending colon, which has never reached its proper position in the right lumbar gutter, tightens the peritoneal reflection across the lower portions of the duodenum.

Clinically, these will cause symptoms and signs in direct proportion to the completeness of the obstruction. The diagnosis may be delayed because of the minor degree of obstruction or, as frequently happens, may become apparent only when the anatomic stenosis becomes converted to a complete obstruction through factors such as edema and blockage by milk curds.

Reports vary as to the level of the atresia or stenosis. Actually, the exact level is of only academic importance. Most authors agree that atresia above the ampulla is very uncommon, the great majority of obstructions lying distal to the ampulla.

INCIDENCE

Congenital duodenal obstruction is relatively rare. It has been variously estimated as occurring once in 10,000 births to once in 40,000 births. Undoubtedly, the condition is being more frequently discovered, largely because of improved medical coverage of the newborn population. From 1954 to 1961, the surgical

Fig. 52-7. — **A,** extrinsic duodenal obstruction due to annular pancreas. **B,** usual course of duodenum down right side of the abdomen after division of bands.