Malrotation of the Intestine in Children

EDWARD G. FORD, M.D., MELVIN O. SENAC, JR., M.D., M. S. SRIKANTH, M.B.B.S., and JORDAN J. WEITZMAN, M.D., F.A.C.S.

Intestinal malrotation may be complicated by volvulus and intestinal necrosis. One hundred two children (64 male, 38 female) undergoing surgical abdominal exploration from 1977 to 1987 had malrotation. Fifty-two patients were less than 7 days of age, 13 from 8 to 30 days, 26 from 31 to 365 days, and 11 were older than 1 year of age. Of infants, 39 of 65 had 40-week gestations, 18 of 65 had 36- to 39-week gestations, and 8 of 65 had less than 36-week gestations. Chief symptomatology included: bilious emesis (47), intestinal obstruction (19), abdominal pain (11), and bloody stools (7). Seventy patients had congenital anomalies (50 single, 20 multiple). Diagnostic evaluations included 56 upper gastrointestinal series and 27 barium enemas. Each patient underwent correction of malrotation and appendectomy, and correction of congenital anomalies (omphalocoele—9, gastroschisis—6, diaphragmatic hernia—7). Complications included short gut (2), sepsis (5), feeding difficulties (2), pneumonia (3), small bowel obstruction (2), and other (15). Nine patients (8.8%) died (trisomy 18—1, trisomy 13—1, intestinal necrosis—3, hepatic failure—1, prematurity—1, other sepsis—2). Two hundred sixteen children with intestinal malrotation have been treated from 1937 to 1987. Mortality rate has improved from 23% to 2.9%.

From The Division of Pediatric Surgery and the Department of Radiology, Children Hospital of Los Angeles, The University of Southern California Medical School, Los Angeles, California, and The Department of Surgery, The Keesler Technical Training Center Medical Center (ATC), Keesler Air Force Base, Mississippi

SYMPTOMS OF MALROTATION of the intestinal tract usually present in infancy and childhood. Malrotation may present acutely as a bowel obstruction and intestinal ischemia associated with midgut volvulus in newborns, or chronically as vague abdominal discomfort and malnutrition in children and adolescents.[1] Seventy per cent of children affected with malrotation may have associated congenital anomalies, including intestinal atresia, omphalocele or gastroschisis, congenital diaphragmatic hernia, or Hirschsprung's disease. Sequelae of symptomatic malrotation range from infants who are parenteral nutrition (TPN)-dependent because of short-bowel syndrome, to chronic learning and psychomotor disabilities in adolescents.

We review our recent 10-year experience of children operated on for abnormalities of intestinal rotation, detailing their clinical presentation, associated congenital anomalies, operative interventions, and follow-up. We compare and contrast our recent experience with two other reports from this institution, culminating a 50-year experience in surgical treatment of rotational disorders of the intestinal tract in children.[2,3]

Materials and Methods

Hospital charts of children found to have intestinal malrotation during surgical abdominal exploration at Childrens Hospital of Los Angeles from 1977 to 1987 were reviewed. We tabulated presenting signs and symptoms, radiographic evaluations, timing of operative intervention, and procedures performed, follow-up, and standard epidemiologic data. Results of the current 10-year experience are compared with two previous reports from this institution to demonstrate trends in presentation, management, and survival. The three combined reports constitute an ongoing 50-year institutional experience of 216 children with malrotation.

Results

Sixty-four male and 38 female children underwent surgical correction of intestinal malrotation as an isolated procedure or in combination with repair of other con-

The opinions expressed herein are those of the authors and not necessarily those of the Air Force or the Department of Defense.

Address reprint requests to Edward G. Ford, Maj, USAF, MC, KTTCMC/SGHS, KAFB, MS 39534.

Accepted for publication September 20, 1991.

genital anomalies. Most patients (50%) presented within the first 7 days of life, 64% presented at less than 1 month of age, and 89% presented under 1 year of age. Term infants were more likely to have malrotation as compared with premature infants (40 weeks' gestation—60%, 36 to 39 weeks—28%, <36 weeks—12%). Bilious emesis (48%) and abdominal distention (21%) were the predominant signs in patients younger than 1 year of age. Abdominal pain followed by bilious emesis was evident in 72% of children older than 1 year of age. Diagnostic radiographic evaluation included 56 upper gastrointestinal series (UGI) and 27 barium enemas (BE).

One hundred twenty-four congenital anomalies were identified in 70 patients; 50 patients had single and 20 had multiple anomalies. Gastrointestinal anomalies were most common (54/102) (Table 1). Central nervous system and cardiac anomalies were next most common (12 each), followed by respiratory (11), genitourinary (6), musculoskeletal (5), splenic (5), oropharyngeal (2), and hematologic (1).

Operative procedures included correction of malrotation and appendectomy in each case, with correction of congenital anomalies as indicated (omphalocele—9, gastroschisis—6, diaphragmatic hernia—7). There were no duodenocolopexies or patients with recurrent volvulus. Complications included: short gut syndrome (2), sepsis (5), feeding difficulties (2), pneumonia (3), small bowel obstruction (2), and other (15). Nine patients (8.8%) died (trisomy 18—1, trisomy 13—1, intestinal necrosis 3, hepatic failure—1, prematurity—1, other sepsis—2).

The Fifty-year Experience

We have treated 216 children for malrotation over the past 50 years (141 males, 75 females). Most patients presented as infants younger than 30 days of age (65%). Neonates uniformly presented with bilious emesis or other clinical and radiographic evidence of high small-bowel obstruction. Eighteen patients (8%) had bloody stools.

TABLE 1. *Congenital Gastrointestinal Anomalies Associated With Malrotation*

Congenital Anomaly	No. of Patients
Omphalocele	9
Gastroschisis	6
Duodenal atresia/stenosis/web	6
Meckel's diverticulum	7
Gastroesophageal reflux	6
Other intestinal atresia/web	6
Biliary atresia	4
Imperforate anus	3
Annular pancreas	2
Rectovaginal fistula	2
Colon duplication	1
Microcolon	1
Tracheoesophageal fistula	1

Melena was not associated with a significantly higher morbidity or mortality. Older children and adolescents presented with chronic abdominal pain, intermittent obstructive symptomatology, or chronic malnutrition. In the period 1937 to 1952, plain abdominal roentgenograms indicated duodenal obstruction in one half of patients, with the remaining one half requiring contrast evaluations to aid in diagnosis. Half of those requiring contrast studies underwent BE and half underwent UGI. The BE and UGI were equally diagnostic. In the period 1952 to 1977, plain films often suggested high intestinal obstruction, but most patients underwent confirmatory contrast studies, with the BE usually performed first. The BE suggested incomplete rotation in most patients, but was not uniformly diagnostic. The UGI uniformly demonstrated duodenal obstruction in patients with midgut volvulus, but was rendered technically difficult by barium remaining in the colon from the preceding BE. In the recent 10 years, a modified approach to contrast evaluations has been instituted to use advantages of both the UGI and BE. Each patient with a suspected malrotation first had a limited UGI (5 cc of barium) that demonstrated duodenal obstruction. If needed, the patient then underwent a contrast enema to show incomplete intestinal rotation as the cause of obstruction.

Associated congenital anomalies were common and most often gastrointestinal. One-hundred six gastrointestinal congenital anomalies were identified in 90 patients. Duodenal atresia/web were most common (11%), followed by Meckel's diverticulum (11%), omphalocele (9%), other stenosis/atresia (5%), and Hirschsprung's disease (2%). Many anomalies included only one or two patients per group and included gastroschisis, biliary atresia, pancreatic anomalies, microcolon, esophageal web, anal stenosis, intestinal duplication, and tracheoesophageal fistula. Eighty per cent of the patients in the 1937 to 1952 group had midgut volvulus associated with their malrotation, 40% of patients in the 1952 to 1977 group had volvulus, and 30% in the most recent group. This apparent reduction in the incidence of midgut volvulus is probably due to increased recognition of malrotation in association with other congenital anomalies. Midgut volvulus is more likely to occur when malrotation of the intestine is an isolated problem not associated with other anomalies such as gastroschisis, omphalocele, and duodenal atresia.

Fifteen patients died in the early postoperative period as a sequela of intestinal necrosis (6%). Mortality rate in the early group (1937 to 1952) was 23%. Intestinal necrosis and functional intestinal obstruction, presumably due to dysfunction after ischemia, and malnutrition with inadequate methods for nutritional support contributed significantly to the high mortality rate. In the subsequent group (1952 to 1977), earlier recognition of the disease process and improved methods of nutritional support led

to a decrease in the perioperative mortality rate to 4%. The trend has continued to the present report of a mortality rate of 2.9%. The difference between the interim group and the most recent report is not statistically significant.

Discussion

During the first 2 months of embryonic development, growth of the intestinal tract exceeds the capacity of the abdominal cavity, and the bowel develops outside the abdomen.[2] At 10 to 12 weeks' gestation, the intestinal tract returns to its usual intra-abdominal location. During its return to the abdominal cavity, the intestine undergoes a simultaneous two-part "rotation" to assume its normal anatomic relationships. In the first phase, the duodenojejunal junction passes behind the superior mesenteric artery and becomes fixed to the upper left retroperitoneum. In the second phase, the cecum passes from the left side of the abdomen, anterior to the superior mesenteric artery, and assumes its normal position right of midline. At completion of rotation, the mesentery becomes fixed to the retroperitoneum in a broad band from the upper left (duodenojejunal junction/ligament of Trietz) to the lower right abdomen (ileocecal junction). Malrotation of the intestine occurs when the two-part rotational process does not proceed normally; the duodenojejunal junction remains right of midline and the cecum remains in the upper left abdomen. The mesentery still attaches to the retroperitoneum; however, instead of a broad-based attachment, the attachment is very narrow. This "omega" configuration predisposes postnatal rotation about the narrow vascular mesenteric axis, leading to midgut volvulus with subsequent intestinal ischemia and necrosis.

Malrotation may occur as an isolated entity, but usually is found in combination with other congenital anomalies. The number of patients in our series presenting with isolated malrotation and volvulus (31%) is similar to percentages reported elsewhere.[1,3,4] When malrotation is associated with volvulus, the anomaly is usually the patient's only significant surgical problem. The most devastating consequence of midgut volvulus is death or loss of sufficient amounts of the intestinal tract to relegate the patient to a life dependent on TPN. Through our recent 10-year experience, we have accumulated six patients with short-bowel syndrome who are hoping that efforts in the field of small bowel transplantation will soon come to fruition.

Typically, the patient with malrotation and volvulus is a term neonate presenting with bilious emesis, abdominal distention, colicky abdominal pain, or overwhelming sepsis from an obvious intra-abdominal process.

Roentgenographic evaluation begins with plain abdominal films to indicate the level of intestinal obstruction. The major plain film findings of volvulus include: (1) a normal or nonspecific study, (2) duodenal obstruction, (3) gastric distention with paucity of intraluminal gas distally (Fig. 1A), and (4) generalized distention of small bowel loops. If the plain film shows complete duo-

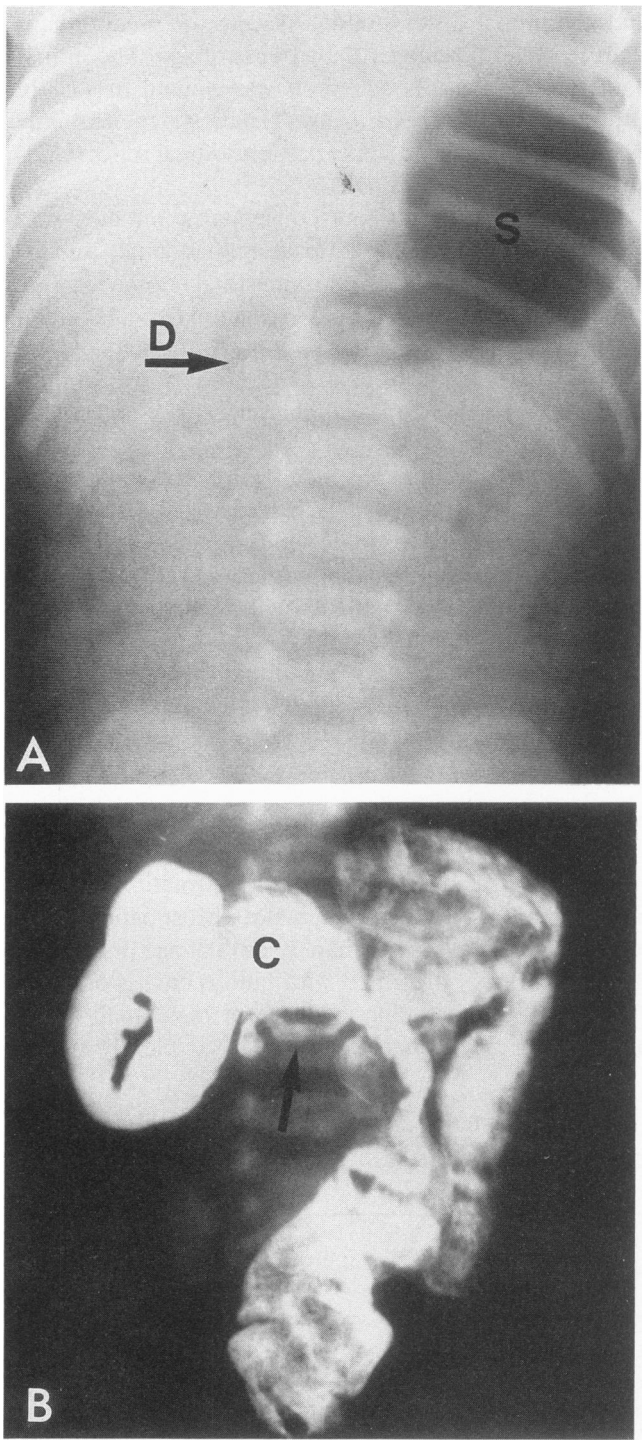


FIG. 1. (A) Plain film of the abdomen showing a dilated stomach with mild duodenal dilatation. Note the paucity of gas distal to the duodenum. S, stomach; D, duodenum. (B) Barium enema study showing malposition of the cecum. C, cecum; *arrow,* appendix.

denal obstruction, then contrast studies are generally not necessary. If duodenal obstruction is incomplete or the precise site of obstruction is unclear, then contrast studies are indicated.

In our institution, UGI is the investigation of choice for malrotation with volvulus. The BE was historically recommended and is still used by many centers. Barium enemas are quick, relatively safe, and identify malrotation by demonstrating an abnormally positioned cecum (Fig. 1B). There are three problems inherent to use of the BE alone to (indirectly) diagnose malrotation: (1) the cecum is mobile in 16% of all age groups, so an enema may not differentiate between a normal child with a high-positioned cecum, a child with malrotation but symptomatic for reasons other than volvulus, or a child with volvulus due to reasons other than malrotation[3]; (2) fluoroscopic and spot-film identification of the cecum may be difficult and confusing even to the experienced pediatric radiologist (Fig. 2), and (3) duodenal obstruction secondary to malrotation and volvulus may rarely occur in the presence of a normally positioned cecum.[5]

The advantages and safety of the UGI series for evaluation of malrotation are well recognized.[4,6] Barium is the preferred contrast material. The major UGI finding in malrotation is an absence of the ligament of Treitz with the proximal jejunum right of midline. Radiographic findings of malrotation with volvulus are: (1) absence of the ligament of Treitz, (2) dilation of the proximal duodenum terminating in a distinctive conical or beak shape, (3) a spiral or corkscrew appearance of the duodenum, and (4) thickening of small bowel folds, signifying vascular obstruction and lymphatic engorgement (Fig. 3).

If the cause of intestinal obstruction remains equivocal after the UGI, a contrast enema may be performed for evaluation of cecal position or for evidence of other entities, such as Hirschsprung's disease, which may mimic midgut volvulus. A contrast enema performed as the initial study generally cannot be followed by an UGI for several hours.

Successful management of midgut volvulus mandates expedient celiotomy, detorsion of the volvulus, and restoration of intestinal perfusion. Immediately after reduction, the intestine is usually edematous, congested, and some areas may appear necrotic. Covering the bowel with warm sponges while correcting the malrotation will usually be rewarded with remarkable vascular recovery of areas initially thought to be hopelessly compromised. Those areas that remain necrotic should be resected, with efforts to preserve intestinal length. After resection of obviously necrotic areas, we routinely replace long segments of intestine with questionable vascular integrity back into the abdominal cavity and return for a "second-look" celiotomy in 36 hours. Questionable areas will declare themselves in the interim, then stomas or a primary anastomosis may be constructed with confidence in tissue integrity.

Surgical correction of malrotation is straightforward, but must be complete to prevent recurrent volvulus (Fig. 4).[7] Loose retroperitoneal tissues attach the ascending colon to the right lateral abdominal wall, as well as connect the colon and its mesentery to the duodenum. "Ladds" bands, from the colon to the abdominal wall, pass over and often obstruct the duodenum, and are usually clearly evident as the dissection begins. Division of this first set of bands exposes the duodenum and clears the relative duodenal obstruction (Fig. 4B). At this point, however, the right colon is still attached to the duodenum by the second set of bands, and the original "omega" shape, with a relatively narrow mesenteric vascular pedicle, is retained (Fig. 4C). Division of the coloduodenal bands allows the mesentery to unfold, much as the opening of a book (Fig. 4D). This opening allows the mesentery to spread out to a broad-based attachment, moves the duodenum and small bowel clearly right of midline, and moves the cecum and right colon left of midline. Because of the possibility of associated duodenal stenosis, atresias, and webs, a Fogerty embolectomy catheter is routinely passed into the proximal jejunum by orogastric passage. Withdrawing the inflated balloon allows identification of intraluminal sites of duodenal obstruction. Several authors suggested coloduodenopexy to help prevent recurrent volvulus[8]; subsequent long-term reports have failed to show an advantage.[9] We will "Pex" the colon and duodenum only in the very rare patient who has an extremely narrow pedicle despite complete lysis of the retroperitoneal attachments. An appendectomy is always performed.

Rotational intestinal anomalies are usually incidental to other major congenital anomalies. Specifically, incomplete body wall fusions such as omphalocele, gastroschisis, or congenital diaphragmatic hernia are almost uniformly associated with malrotation. When possible, the malrotation is corrected when the abdominal wall defect is repaired. This is usually easily accomplished in patients with congenital diaphragmatic hernia or omphalocele. Thickening and foreshortening of the bowel in patients with gastroschisis may make identification and correction of the rotational anomaly difficult.

When evaluating a patient with a history of multiple anomalies who presents with recurrent abdominal pain, distention, or difficult feeding, one should consider malrotation in the differential diagnosis. Normal or nonspecific plain film radiographs are commonly seen in these patients and should not dissuade one from the diagnosis of malrotation or midgut volvulus. We recommend a UGI as the initial contrast study, which may be followed by a contrast enema, if needed. Malrotation may not be evident on initial contrast examinations. If symptomatology persists, one should not hesitate to repeat the contrast studies.

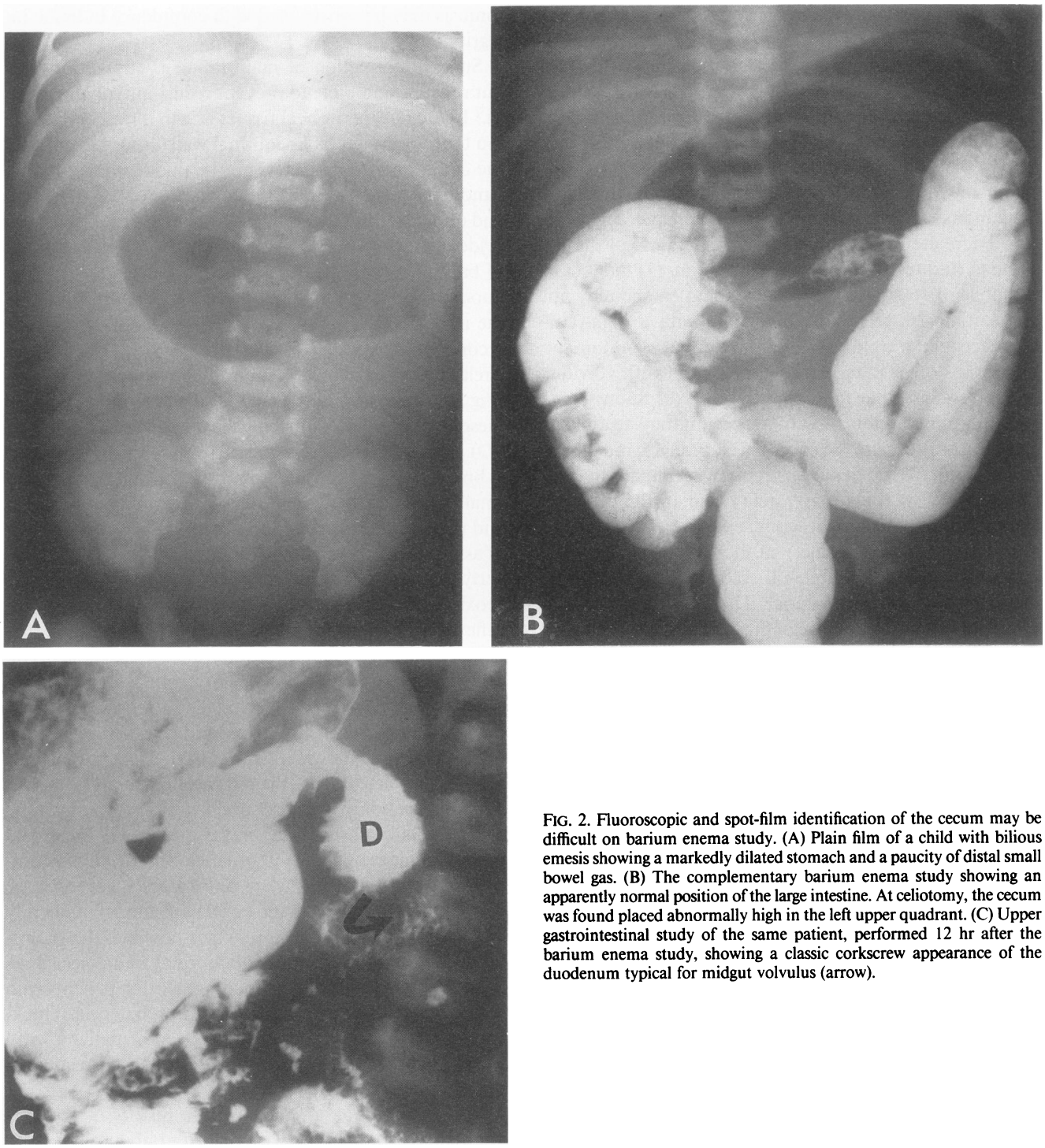
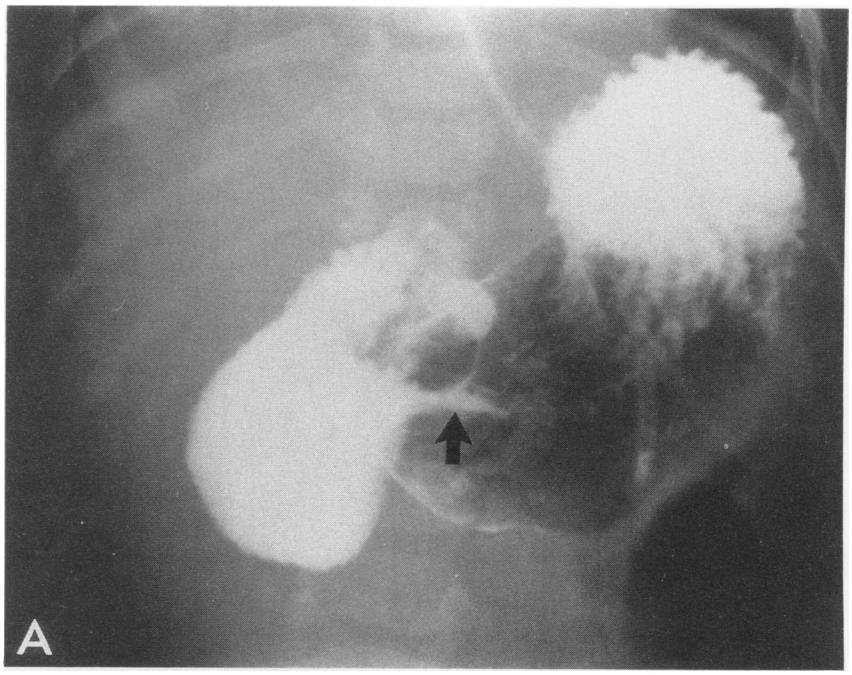


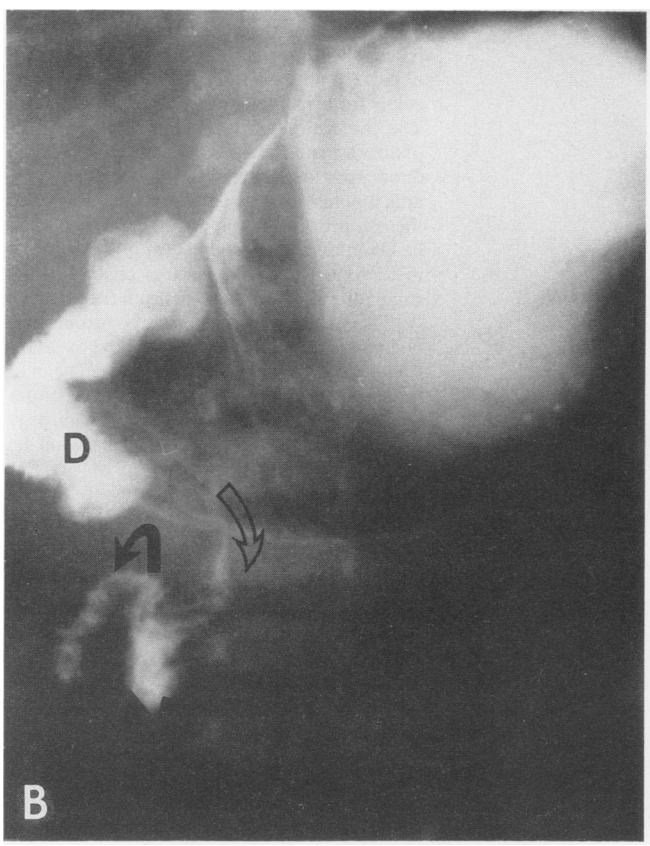
FIG. 2. Fluoroscopic and spot-film identification of the cecum may be difficult on barium enema study. (A) Plain film of a child with bilious emesis showing a markedly dilated stomach and a paucity of distal small bowel gas. (B) The complementary barium enema study showing an apparently normal position of the large intestine. At celiotomy, the cecum was found placed abnormally high in the left upper quadrant. (C) Upper gastrointestinal study of the same patient, performed 12 hr after the barium enema study, showing a classic corkscrew appearance of the duodenum typical for midgut volvulus (arrow).

The technique of limited upper intestinal series followed by barium enema is usually rewarding.

Nine patients in our series died. Two expired with multiple congenital anomalies associated with Trisomy 18 and 13, one with TPN-related hepatic failure, one with multiple difficulties associated with his prematurity, and two with overwhelming septic episodes after several months' hospitalization. Three patients died as a direct result of intestinal necrosis from midgut volvulus associated with their malrotations. The overall mortality rate of volvulus has not changed significantly since the last report from this institution (2.9% *versus* 4%).

The remarkable improvement in perioperative mortality rate of children with midgut volvulus is a result of





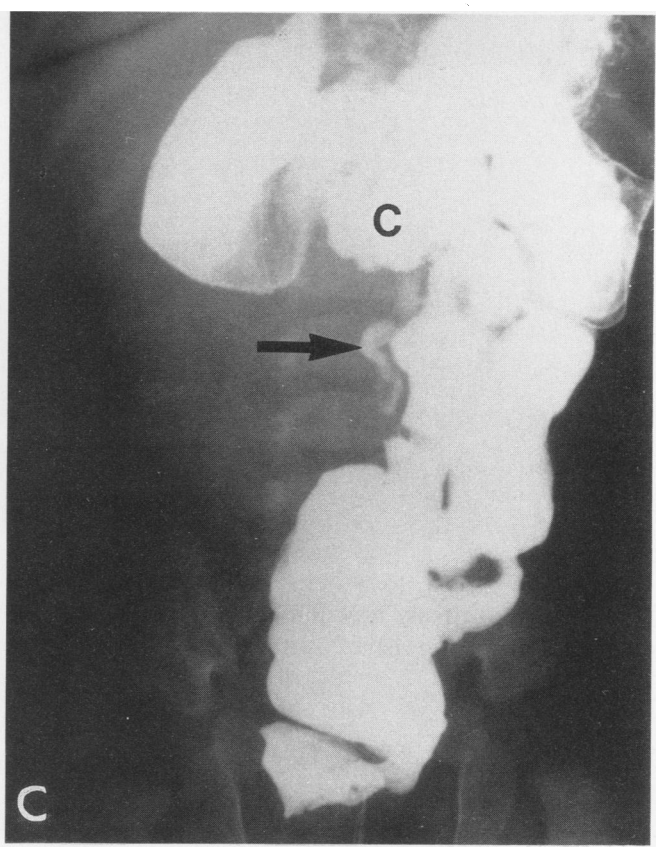


FIG. 3. Classic roentgenographic findings in patients with malrotation. (A) Contrast in the duodenum shows complete obstruction with the distinctive conical or beak shape (arrow). (B) The spiral or corkscrew appearance of the duodenum with a low or absent ligament of Treitz. D, duodenum; arrows indicate course of contrast through the corkscrew-shaped bowel. (C) Barium enema study of the patient in Fig. 3B showing typical malrotation, with large bowel predominately to the left of midline and the cecum riding high in the left upper quadrant. C, cecum; arrow, appendix.

developments in techniques of aggressive nutritional support. Parenteral nutrition is instituted early and continued until the patient is able to support himself or herself on full volitional feedings.

Malrotation of the intestines with midgut volvulus is a complicated and potentially lethal congenital anomaly. Unlike other complex congenital anomalies, once the embryogenesis and resulting anatomic abnormalities of malrotation are understood, the surgical repair is relatively straightforward. Repair of the anomaly must be complete

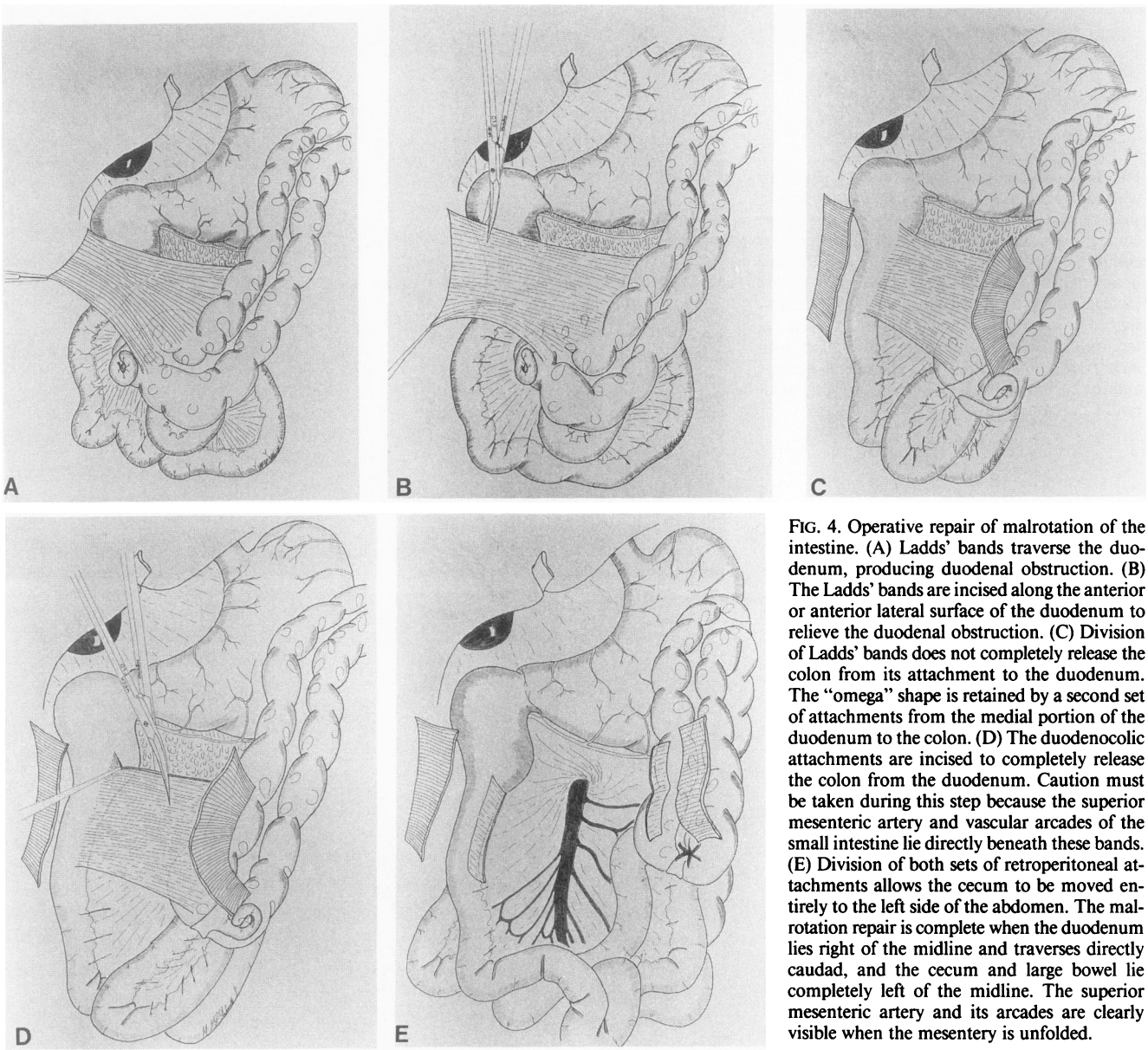


FIG. 4. Operative repair of malrotation of the intestine. (A) Ladds' bands traverse the duodenum, producing duodenal obstruction. (B) The Ladds' bands are incised along the anterior or anterior lateral surface of the duodenum to relieve the duodenal obstruction. (C) Division of Ladds' bands does not completely release the colon from its attachment to the duodenum. The "omega" shape is retained by a second set of attachments from the medial portion of the duodenum to the colon. (D) The duodenocolic attachments are incised to completely release the colon from the duodenum. Caution must be taken during this step because the superior mesenteric artery and vascular arcades of the small intestine lie directly beneath these bands. (E) Division of both sets of retroperitoneal attachments allows the cecum to be moved entirely to the left side of the abdomen. The malrotation repair is complete when the duodenum lies right of the midline and traverses directly caudad, and the cecum and large bowel lie completely left of the midline. The superior mesenteric artery and its arcades are clearly visible when the mesentery is unfolded.

to provide a normally functioning gastrointestinal tract and to minimize the risk of continued obstruction or recurrent volvulus.

References

1. Powell DM, Othersen HB, Smith CD. Malrotation of the intestine in children: The effect of age on presentation and therapy. J Pediatr Surg 1989; 14(8):777–780.
2. Snyder WH, Chaffin L. Embryology and pathology of the intestinal tract: presentation of 40 cases of malrotation. Ann Surg 1954; 140(3):368–380.
3. Andrassy RJ, Mahour GH. Malrotation of the midgut in infants and children. Arch Surg 1981; 116:158–160.
4. Stringer DA. Congenital and developmental anomalies of the small bowel. *In* Stringer DA, ed. Pediatric Gastrointestinal Imaging. Toronto: BC Decker, 1989, pp 235–249.
5. Berdon WE, Baker DH, Bull S, et al. Midgut malrotation and volvulus. Radiology 1970; 96:375–383.
6. Simpson AJ, Konidas JC, Kerasna IH, et al. Roentgen diagnosis of midgut malrotation: value of upper gastrointestinal radiographic study. J Pediatr Surg 1972; 7:243–252.
7. Weitzman JJ. The surgical treatment of malrotation of the intestine, motion picture, ACS-625, American College of Surgeons Surgical Film Library, Davis and Geck, Danbury, CT.
8. Brennom WS, Bill AH. Prophylactic fixation of the intestine for midgut nonrotation. Surg Gynecol Obstet 1974; 138:181–184.
9. Stauffer UG, Hermann P. Comparison of the late results in patients with corrected intestinal malrotation with and without fixation of the mesentery. J Pediatr Surg 1981; 15(1):9–12.