Intestinal Atresia and Stenosis

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Contents

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63.1 Introduction

Atresias of the jejunum and ileum account for about one third of all cases of neonatal intestinal obstruction with a third of infants born prematurely or small for dates. Stenoses are much less common, accounting for about 5% of cases, and seldom present in the newborn period because of delay in diagnosis. While these conditions are associated with excellent prognoses in developed countries, delayed presentation and limitations in resources to support patients with delayed return of intestinal function contribute to overall lower survival rates in many African countries. Early recognition and proper surgical management are vitally important to improving the patient's chance of survival in countries with limited access to health-care resources.

63.2 Demographics

The incidence of intestinal atresia in the United States is approximately 1 in 3000 live births [1] but may be more frequent in Africa with a reported incidence as frequent as under 1:1000 live births with types III and IV comprising 35% of the total number. Reports from Nigeria [2–4] have shown that intestinal atresias are less common than imperforate anus, but occur with similar frequency to Hirschsprung's disease. There also appears to be an increased incidence in twin pregnancies at 7.3 per 10,000 live births. While published reports suggest a higher prevalence of jejunoileal atresia among African-American children in the United States, no racial predilection has been identified by authors from African countries. The male-to-female ratio is equal in most reports. There have been other familial associations reported, frequently type IV lesions, but to date, a specific gene mutation has not been discovered.

63.3 Etiology and Pathophysiology

Our present understanding of the etiology of intestinal atresias is based upon the classic experimental work of Louw and Barnard reported in 1955 [5]. These investigators observed that ligating mesenteric vessels and causing strangulated obstruction in fetal dogs resulted in atretic lesions of the small intestine that were similar to those observed clinically in human neonates. Thus, atresias and stenoses of the small intestine are believed to be due to an ischemic insult. This etiologic mechanism explains the frequent association of atresias with mesenteric defects and with other conditions that may cause strangulated obstruction of the intestinal tract (e.g., volvulus, intussusception, internal hernias, and gastroschisis). An ischemic etiology may also explain why intestinal atresia is associated with maternal smoking and vasoconstrictor drug exposure during pregnancy. More recently, some of the established concepts of bowel development and rotation have been questioned.

The morphological classification into four types has both prognostic and therapeutic implications. Classification is as follows (Fig. 63.1) [6]:

- Stenoses (11%)
- Atresia type I (23%)
- A transluminal septum with proximal dilated bowel in continuity with collapsed distal bowel. The bowel is usually of normal length.
- Atresia type II (10%)
- Two blind-ending atretic ends separated by a fibrous cord along the edge of the mesentery with mesentery intact
- Atresia type IIIa (15%)
- Similar to type II, but there is a mesenteric defect and the bowel length may be foreshortened
- Atresia type IIIb ("apple peel" or "Christmas tree" deformity) (19%)
- Consists of a proximal jejuna atresia, often with malrotation with absence of most of the mesentery and a varying length of ileum surviving on perfusion from retrograde flow along a single artery of supply
- Atresia type IV (22%)
- Multiple atresias of types I, II, and III like a string of sausages. Bowel length is always reduced. The terminal ileum as in type III is usually spared

The immediate consequence of an atresia is dilatation of bowel proximal to the first occlusion encountered for a variable distance due to increased pressure as a result of ongoing fetal ingestion of amniotic fluid. This may lead to a cyanotic appearance of the dilated segment as well as some areas of necrosis. This dilated bowel, even when the obstruction is relieved by resection and anastomosis or stoma formation, remains dilated having inefficient prograde peristalsis – with histological and histochemical abnormalities being observed up to 20 cm proximal to the obstruction [7]. Surgical strategies to overcome this problem of dilatation and dysmotility include back resection of dilated bowel to normal caliber intestine or reduction in the diameter by various tapering maneuvers. Bowel distal to the atresia is disused and narrow, but essentially should have normal peristaltic function once continuity has been restored.

63.4 Clinical Presentation

Intestinal atresias in Africa are usually not diagnosed prenatally. However, atresias of the proximal jejunum are frequently associated with polyhydramnios. Therefore, many of these patients are born prematurely and often

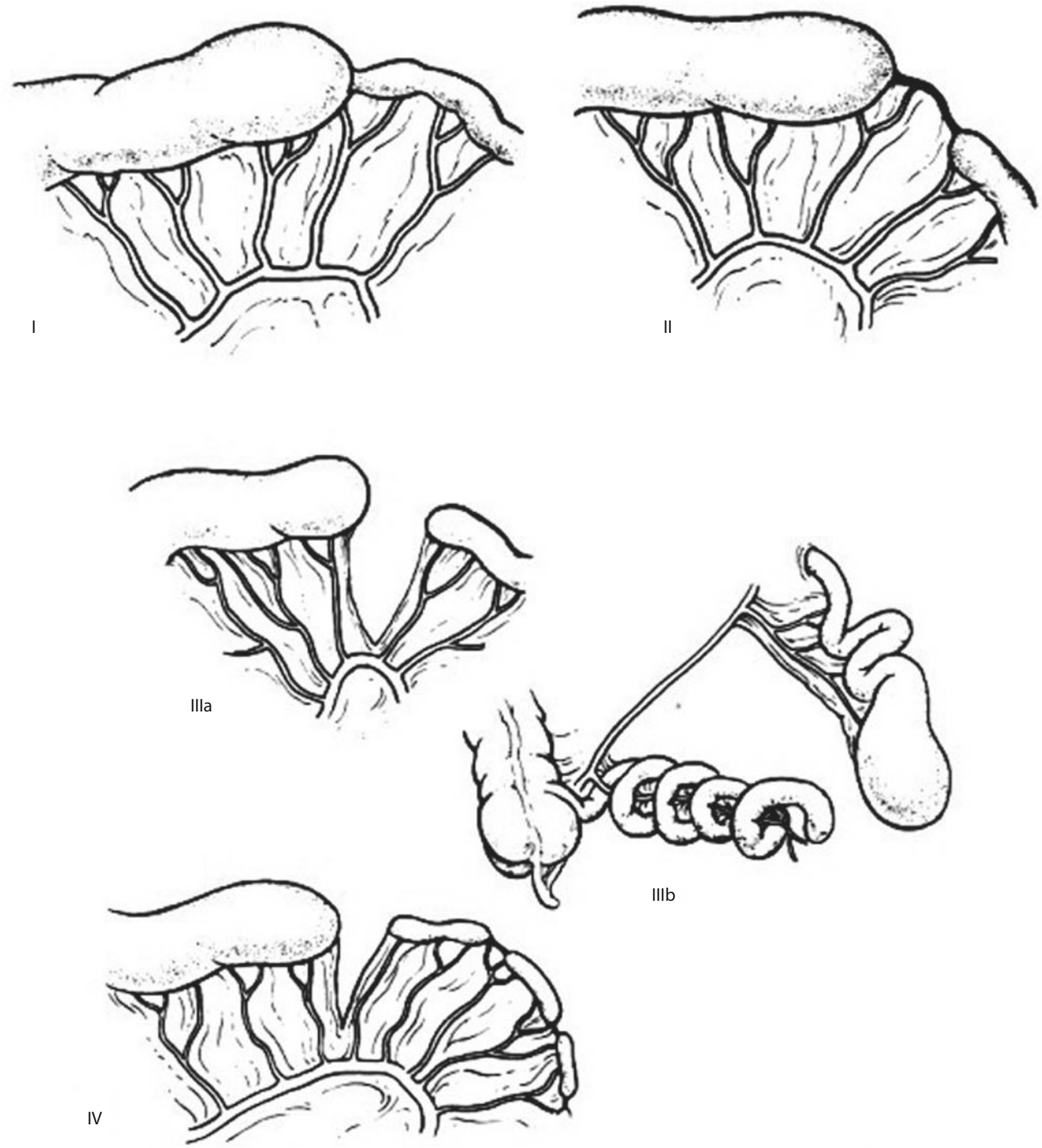

Fig. 63.1 Classification of intestinal atresia. (Source Grosfeld et al. [9])

are small for gestational age, probably resulting from inability to absorb nutrients from ingested amniotic fluid. Nearly, all infants with intestinal atresias develop symptoms within the hours after birth. However, several publications have documented that neonates in African countries often do not reach definitive medical care for several days. Unlike in patients with atresias, those with intestinal stenoses are not usually diagnosed until well beyond the neonatal period.

Intestinal atresia should be suspected in any newborn showing evidence of bowel obstruction (bilious vomiting, abdominal distension, and failure to pass meconium). Aspiration of >25 ml of fluid from the stomach via a nasogastric tube is very suggestive of

obstruction. Antenatal ultrasound scanning may show dilated loops of bowel with vigorous peristalsis, which is diagnostic of obstruction. Polyhydramnios may develop but is more commonly seen in duodenal and esophageal obstructions. The more distal the atresia, the more generalized the abdominal distension. After aspiration of gastric contents, the abdomen will be less distended and visible peristalsis may be observed. There is a failure to pass meconium, and typically small volume gray mucoid stools are passed. Abdominal tenderness or peritonitis only develops with complications of ischemia or perforation. This commonly occurs with delay in diagnosis and is due to increased intraluminal pressure from swallowed air and secondary volvulus of the bulbous blind-ending bowel at the level of the first obstruction.

63.5 Physical Examination

Findings on physical examination are frequently not very revealing. Most patients will have some degree of abdominal distension. The amount of distension will vary depending on the level of obstruction. Patients generally do not have abdominal tenderness or an abdominal mass. Therefore, the presence of these findings suggests a complicated obstruction associated with ischemia or prenatal perforation, or that the cause of obstruction is malrotation with midgut volvulus.

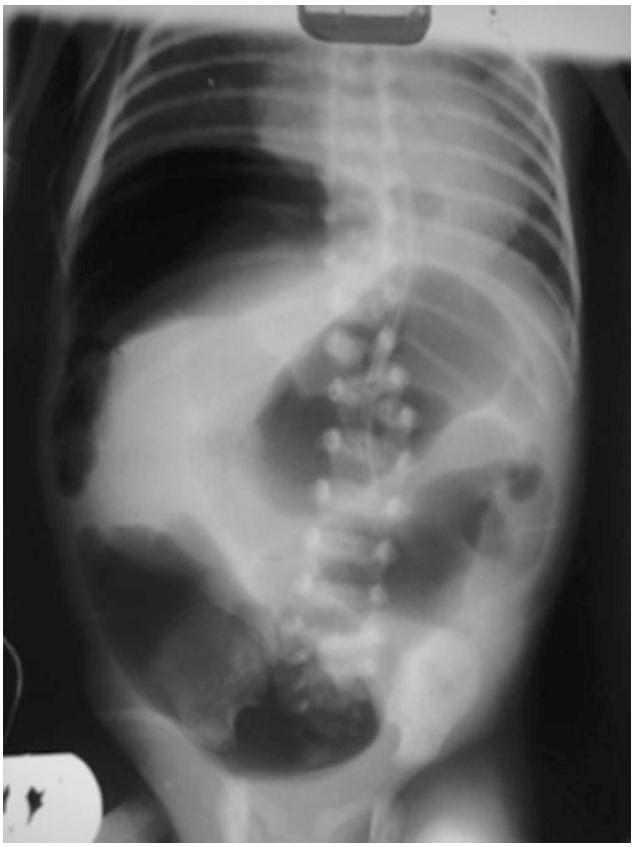

Fig. 63.2 Abdominal radiograph showing several dilated gas-filled loops in a jejunal atresia

63.6 Investigations

In most patients, a simple abdominal x-ray with anteroposterior and either cross-table or left lateral decubitus projection are adequate to make the diagnosis based on the presence of dilated air-filled intestinal loops and air–fluid levels (Fig. 63.2). In addition, plain abdominal x-rays will suggest the level of obstruction based on the number of dilated bowel loops. The presence of multiple dilated bowel loops without air–fluid levels suggests the possibility of meconium ileus, particularly if the intestinal content has a "ground glass" appearance. Presence of a single and very dilated loop with a large fluid level is often indicative of colonic atresia.

The differential diagnosis includes other causes of intestinal obstruction in the neonate. In patients with evidence of a proximal complete obstruction, the differential diagnosis is limited, and no additional diagnostic studies are required. In patients with multiple dilated bowel loops suggesting a distal obstruction, the differential diagnosis includes several conditions for which surgical intervention may not be required. Therefore, in these patients, a contrast enema may be helpful to look for evidence of meconium plug or meconium ileus which may respond to nonoperative managements. In addition, a contrast enema may demonstrate findings suggestive of Hirschsprung's disease, which would direct initial management toward obtaining confirmatory tests for this disease. Contrast enema showing a patent colon is helpful in that demonstration of colonic patency by injection of saline at operation, a sometimes tedious procedure, is not required (Fig. 63.3).

In patients with intestinal stenoses, plain abdominal x-rays may demonstrate proximal bowel dilation; however, in most patients, a gastrointestinal contrast meal or enema is required to confirm and locate the site of partial obstruction.

63.7 Management

All patients should receive judicious fluid hydration prior to operative intervention. In addition, a nasogastric or orogastric tube should be passed to empty the stomach and decrease the risk of vomiting with aspiration. In general, patients with intestinal atresias have a low risk of associated cardiac anomalies, so that preoperative special investigation is not required unless the patient has clinical evidence of a serious cardiac defect.

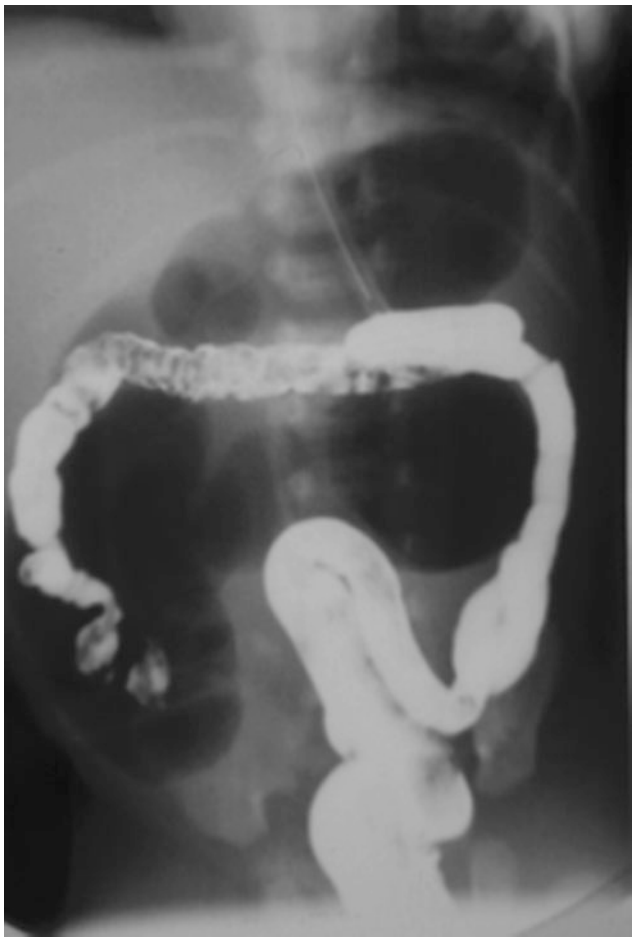

Fig. 63.3 Contrast enema showing normal colon with dilated proximal small bowel in an infant with jejunal atresia

At exploration, usually via a transverse supraumbilical incision or periumbilical approach, the site of the most proximal atresia is readily identified as the site of marked change in intestinal caliber. The outer wall of the intestine at the site of obstruction may appear intact, or there may be an associated defect in continuity of the intestine and the mesentery (Fig. 63.4). Generally, surgical treatment requires excision of the ends of the intestine involved in the atresia. It is also important to look for distal sites of obstruction which can occur in up to 20% of patients and may not be immediately obvious due to lack of caliber change beyond the proximal atresia. These distal points of obstruction can be identified by flushing the distal intestinal lumen with saline to confirm intestinal continuity to the level of the rectum.

After resection of the atretic segment, the surgeon is faced with the difficult task of re-establishing continuity between intestinal segments with marked size discrepancies. Another consideration is the potential dysmotility of the proximal markedly dilated segment, which may result in delayed intestinal function and problems with bacterial overgrowth. Therefore, in patients with a relatively short segment of severely dilated proximal intestine, resection of the dilated segment with re-establishment of continuity by end-to-end anastomosis is a good option. However, in patients with long segments of proximal intestine that are significantly dilated, resection of the whole involved segment may result in inadequate remaining intestinal length to allow absorption of enteric nutrients (i.e., short-bowel syndrome). Therefore, these patients frequently are treated by either imbrication or tapering enteroplasty of the proximal dilated segment. To date, no randomized studies have compared the outcomes for patients with intestinal atresias with or without the addition of an enteroplasty or plication, but imbrication tends to result in recurrence of dilatation and dysmotility. In patients where the atresia is just distal to the duodenojejunal flexure, it may be advantageous to resect the dilated bowel, de-rotate, and taper the duodenum with primary anastomosis. This facilitates passage of a trans-anastomotic feeding tube and early restoration of foregut function. The total residual length of bowel should be measured with a tape and recorded, as this gives some guidance as to prognosis.

Patients who have multiple atresias (type IV) or an apple-peel deformity (type IIIb) (Figs. 63.5 and 63.6) are particularly challenging management problems. These patients may require multiple anastomoses and frequently will experience long-term delays in return of intestinal function. In addition, many of these patients will have short-bowel syndrome due to inadequate residual intestinal length. In general, the formation of stomas is unnecessary and should be avoided as dilated bowel does not reduce in caliber and fluid and electrolyte losses may be severe.

While most centers still perform surgery via a laparotomy, circumumbilical incision as well as laparoscopic-assisted surgical techniques are gaining popularity, and in centers where there is adequate experience and equipment, the cosmetic outcomes are improved with little effect on the functional outcomes as long as the basic principles discussed above are adhered to.

Where short-bowel syndrome is evident, it is advised to perform a primary end-to-end anastomosis and allow for adaptation to progress before intervening surgically at a later stage outside the neonatal period rather than embark on primary autologous intestinal reconstructive surgery (AIRS) such as the serial transverse enteroplasty procedure (STEP) or lengthening and tailoring procedures (LILT, Bianchi) as these have no defined place at the initial operation. In the African setting, however, this approach may not be feasible due to the fact that TPN is not universally available.

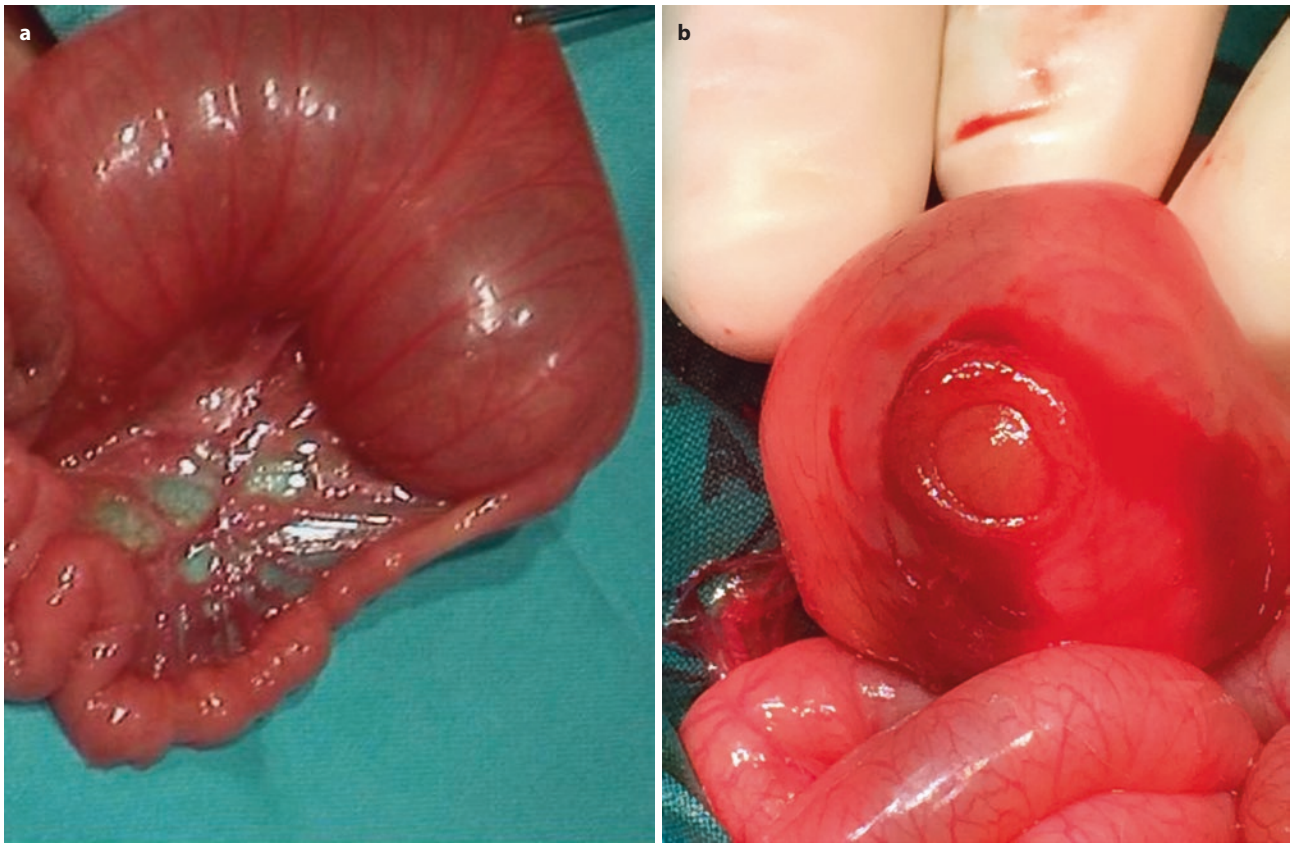

Fig. 63.4 Type I jejunal atresia. Membrane occlusion without mesenteric defect or loss of intestinal length **a** and the cut surface at surgery showing the occluding membrane **b**

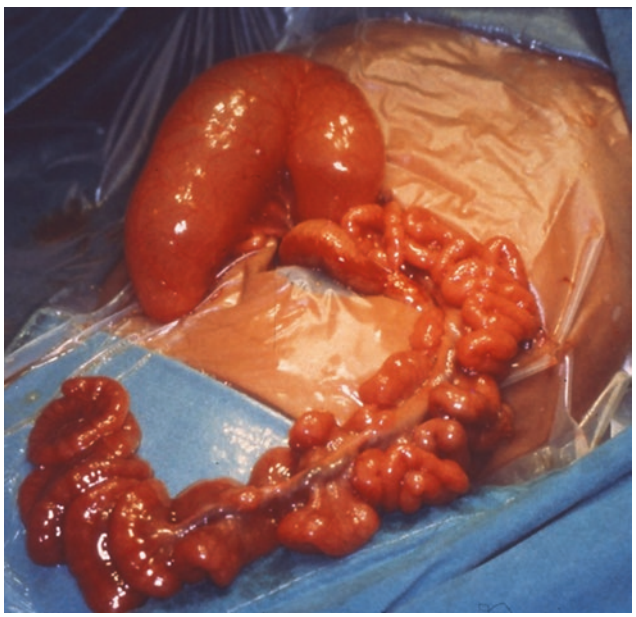

Fig. 63.5 Type III b atresia. Note the proximal jejunal atresia, malrotation, and mesenteric defect with a single artery of supply from the middle and right colic vessels with significant loss of intestinal length

63.8 Postoperative Complications

The most common postoperative complication is a functional obstruction at the site of anastomosis [8]. Unfortunately, this complication may be due to the underlying intestinal dysmotility associated with this anomaly and may not be preventable by changes in surgical technique. Other less commonly observed complications include anastomotic leak and adhesive obstructions. Obstructions due to missed distal unrecognized atresias should not occur and can be prevented by proper evaluation at the time of the initial operation.

63.9 Prognosis and Outcomes

Most patients with intestinal atresia do not have associated life-threatening anomalies. Therefore, the primary factor that impacts mortality is ability to support the nutritional needs of the patient during the postoperative period while awaiting adequate bowel function to allow enteral alimentation. [9] In centers where parenteral nutritional support is feasible, these patients can be

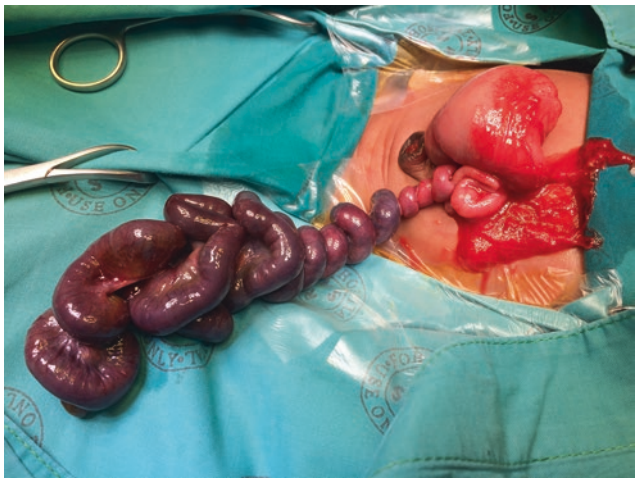

Fig. 63.6 Type III b atresia with antenatal volvulus of the "apple peel"

supported for prolonged periods of time while awaiting gastrointestinal function. However, in centers without these resources, patient mortality will be higher and primarily attributable to malnutrition. The judicious use of nasojejunal or gastrostomy trans-anastomotic feeding tubes for enteral feeding may be life saving.

63.10 Prevention

Unfortunately, at present, there are no options for prevention since these anomalies are usually not recognized prior to birth.

63.11 Ethical Issues

In resource-poor regions without recourse to intensive care and parenteral nutrition infants with ultra-short-bowel resulting from congenital atresia may have to be managed conservatively. Discussion around parental expectations and center outcomes should be part of the informed consent. Nursing staff and other care givers should also be party to the decision-making process. Withdrawal of treatment which is thought futile is often difficult to institute. If there are choices to be made based on allocation of limited resources, then infants with the potential for good outcomes may be given preference for meager resources. However, it is only the infrequent case of intestinal atresia that develops intestinal failure and with prompt operation and preservation of as much functioning bowel as possible prognosis should be excellent.

63.12 Evidence-Based Surgery

Title	What is our development progress for the treatment outcome of newborn with intestinal atresia and stenosis in a period of 28 years?
Authors	İbrahim Akkoyun, Derya Erdoğan,[1] Yusuf Hakan Çavuşoğlu,[1] and Özden Tütün[1]
Institution	Department of Pediatric Surgery, Dr. Faruk Sükan Maternity and Children Hospital Konya, Ankara, Turkey. Department of Pediatric Surgery, Dr. Sami Ulus Maternity and Children Hospital, Ankara, Turkey
Reference	N Am J Med Sci. 2013 Feb; 5(2): 145–148
Problem	The aim of this study was to examine our series of intestinal atresia and stenosis patients in a period of 28 years in a developing country and to display our progress in treatment and survival rates today.
Intervention	In this study, a total of 141 intestinal atresia and stenosis cases were retrospectively evaluated.
Comparison/control (quality of evidence)	The cases were categorized in two groups as 45 cases before the 1990 (group 1) when it was impossible for total parenteral nutrition (TPN) solutions to be used regularly, without complication and for a long time and 96 cases after 1990 (group 2) when this was possible. While the survival rate before 1990 was 55%, after 1990, it was 94%.
Outcome/effect	As a result, long-term regular TPN usage significantly improved survival in newborns with intestinal atresia and stenosis in a developing country.

Title	Prognostic factors related to mortality in newborns with jejunoileal atresia
Authors	Bracho-Blanchet E[1], González-Chávez A, Dávila-Pérez R, Zalles Vidal C, Fernández-Portilla E, Nieto-Zermeño J
Institution	Departamento de Cirugía General, Hospital Infantil de México Federico Gómez, Secretaría de Salud, México, D.F., Mexico
Reference	Cir Cir. 2012 Jul-Aug;80(4):345–351
Problem	To assess the factors related to mortality in neonates with jejunoileal atresia.
Intervention	Case–control nested in a cohort design, comparative study during 10 years, between deceased and survivors analyzing factors related to mortality before surgery, during surgery, and in the postoperative period.

Comparison/control (quality of evidence)	We analyzed 70 patients in 10 years, and there were 10 deaths (14.2%). None had a prenatal diagnosis. Factors related to mortality were intestinal perforation with a relative risk (RR) of 4.4, peritonitis (RR: 5.6), the need of stomas (RR: 4.9), the presence of sepsis (RR: 4.6), and a residual small bowel length below 1 meter (RR: 7.4).
Outcome/effect	Delay in diagnosis causes late intervention and increased mortality and results in late transport of the neonate which increases mortality.
Historical significance/comments	It is necessary to spread awareness of this disease in the medical community to improve prenatal detection and in utero transfer to centers able to deal with this condition.

Key Summary Points

1. Intestinal atresia may occur at any level of the gastrointestinal tract.
2. Small bowel atresia in most cases is due to an antenatal ischemic insult to a segment of intestine. Resorption of the infarcted segment leads to occlusion of the lumen with a varying degree of dilatation of the proximal blind end.
3. A third of infants with intestinal atresia are born prematurely.
4. Differential diagnoses include midgut volvulus, meconium ileus, extensive aganglionosis, and intussusception.
5. Primary operation consists of a generous back resection of the bulbous blind end and an end-to-end anastomosis.
6. Outcomes are generally good if sufficient bowel length remains.
7. Stomas should be avoided.
8. Mortality rate depends on birth weight, residual bowel length, the degree of dysmotility, associated anomalies, and septic complications.

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