CHAPTER 63
INTESTINAL ATRESIA AND STENOSIS

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Introduction

Atresias of the jejunum and ileum are common causes of bowel obstruction in the neonate, with a third of infants born prematurely or small for their gestational age. Stenoses are much less common and seldom present in the newborn period due to delay in diagnosis. Whereas 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 contributes to overall lower survival rates in many African countries.[1–4] Early recognition and proper surgical management are vitally important to improving survival in countries with limited access to health care resources.

Demographics

The incidence of intestinal atresia in the United States is approximately 1 in 3,000 live births, but may be more frequent in Africa, with a reported incidence of less than 1 in 1,000 live births, with types III and IV (see next section) comprising 35% of the total number. Two reports from Nigeria[1,2] have shown that intestinal atresias are less common than imperforate anus, but occur with similar frequency to Hirschsprung's disease. Published reports suggest a higher prevalence of jejunoileal atresia among African American children in the United States,[5] but no racial predilection has been identified by authors from African countries. Nearly all infants with intestinal atresias develop symptoms within hours after birth. However, several publications have documented that neonates in African countries often do not reach definitive medical care for several days.[2,4] Unlike atresias, many patients with intestinal stenoses are not diagnosed until well beyond the neonatal period.

Aetiology/Pathophysiology

Our present understanding of the aetiology of intestinal atresias is based upon the classic experimental work of Louw and Barnard reported in 1955.[6] These investigators observed that ligating mesenteric vessels and causing strangulated obstruction in foetal 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 ischaemic insult. This aetiologic 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 ischaemic aetiology may also explain why intestinal atresia is associated with maternal smoking and vasoconstrictor drug exposure during pregnancy.

The morphological classification into four types has both prognostic and therapeutic implications (Figure 63.1):

- Stenoses occurs in 11%.

- Type I atresia (23%) is a transluminal septum with proximal dilated bowel in continuity with collapsed distal bowel. The bowel is usually of normal length.

- Type II atresia (10%) involves two blind-ending atretic ends separated by a fibrous cord along the edge of the mesentery with mesentery intact.

- Type IIIa atresia (15%) is similar to type II, but there is a mesenteric defect and the bowel length may be foreshortened.

- Type IIIb atresia (19%) ("apple peel" or "Christmas tree" deformity) consists of a proximal jejunal 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.

- Type IV atresia is a multiple atresia 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 the bowel for a variable distance proximal to the first occlusion encountered. This dilated bowel, even when the obstruction is relieved by resection and anastomosis or stoma formation, remains dilated, having inefficient prograde peristalsis. Surgical strategies to overcome this include back resection of this bowel to a normal-calibre intestine or reduction in diameter by various tapering manoeuvres.
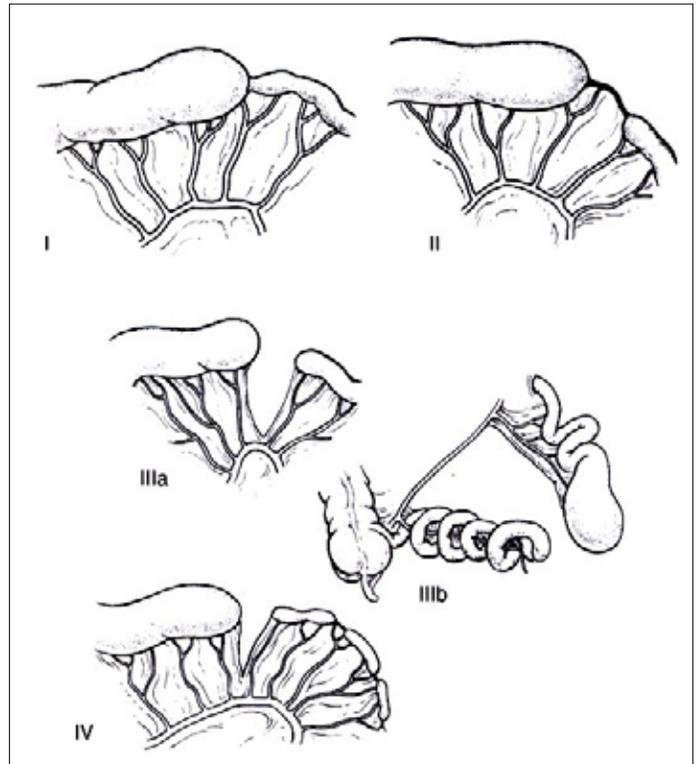
Source: Grosfeld JL, et al. Operative management of intestinal atresia and stenosis based on pathological findings. J Pediatr Surg 1979; 14:368.

Figure 63.1: Classification of intestinal atresia (see text for explanation of types I–IV).

History

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 are small for their gestational age, the latter due to inability to absorb nutrients from the amniotic fluid in patients with proximal intestinal obstructions.[7]

Intestinal atresia should be suspected in any newborn showing evidence of bowel obstruction (bilious vomiting, abdominal distention, and failure to pass meconium). Aspiration of >25 ml of fluid from the stomach via a nasogastric tube (NGT) 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 it is more commonly seen in duodenal and oesophageal obstructions. The more distal the atresia, the more generalized the abdominal distention. After aspiration of gastric contents, the abdomen will be less distended and visible peristalsis may be observed. There is usually a failure to pass meconium, and typically small-volume gray mucoid stools are passed. Abdominal tenderness or peritonitis develops only with complications of ischaemia or perforation. This commonly occurs with a 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.

Physical Examination

Findings on physical examination are frequently not very revealing. Most patients will have some degree of abdominal distention. The amount of distention will vary, depending upon 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 ischaemia or prenatal perforation, or that the cause of obstruction may be malrotation with midgut volvulus.

Investigations

In most patients, a simple abdominal x-ray with anteroposterior (AP) and either cross-table or left lateral decubitus projection are adequate to make the diagnosis based upon the presence of dilated, air-filled intestinal loops and air-fluid levels (Figure 63.2). In addition, plain abdominal x-rays will suggest the level of obstruction based upon 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. A single very dilated loop with a large fluid level is often indicative of 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 a meconium plug or meconium ileus, which may respond to nonoperative management. In addition, a contrast enema may demonstrate findings suggestive of Hirschsprung's disease, which would direct initial management towards obtaining confirmatory tests for this disease. A 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 (Figure 63.3).

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

Figure 63.2: Abdominal radiograph showing several dilated gas-filled bowel loops in a jejunal atresia.

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

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.

At exploration, the site of the most proximal atresia is readily identified as the site of marked change in intestinal calibre. 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 (Figure 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 calibre 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. In patients for whom the atresia is just distal to the duodenojejunal flexure, it may be advantageous to resect the dilated bowel, derotate, and taper the duodenum with primary anastomosis. This facilitates passage of a transanastomotic 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) present particularly challenging management problems (Figures 63.5 and 63.6). 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 because dilated bowel does not reduce in caliber, and fluid and electrolyte losses may be severe.

Postoperative Complications

The most common postoperative complication is a functional obstruction at the site of anastomosis. 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.

Prognosis and Outcomes

Most patients with intestinal atresia do not have associated life-threatening anomalies. Therefore, the primary factor that impacts mortality is the ability to support the nutritional needs of the patient during the postoperative period while awaiting adequate bowel function to allow enteral alimentation. In centres where parenteral nutritional support is feasible, these patients can be supported for prolonged periods of time while awaiting gastrointestinal function. However, in centres without these resources, patient mortality will be higher and primarily attributable to malnutrition. The judicious use of nasojejunal or gastrostomy transanastomotic feeding tubes for enteral feeding may be life saving.

Figure 63.4: Type I jejunal atresia. Membrane occlusion without mesenteric defect or loss of intestinal length.

Figure 63.5: Type IIIb atresia (apple peel deformity). Note 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.

Figure 63.6: Type IIIb atresia with antenatal volvulus of the "apple peel" leading to short bowel.

Prevention

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

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 centre outcomes should be part of the informed consent. Nursing staff and other caregivers should also be party to the decision-making process. Withdrawal of treatment that is thought to be 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 meagre 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.

Evidence-Based Research

Table 63.1 presents a study of the change in mortality rates from jejunoileal atresia and stenosis over a nearly 50-year period.

Table 63.1: Evidence-based research.

Title	Jejuno-ileal atresia and stenosis
Authors	Rode H, Millar AJW
Institution	Red Cross Children's Hospital, Cape Town, South Africa
Reference	In: Prem Puri, ed. Newborn Surgery, 2nd ed. Arnold, 2003, Pp 445–456
Problem	Late presentation of jejunoileal atresia and stenosis in Africa.
Intervention	Surgery is curative if presentation is early enough.
Comparison/control (quality of evidence)	Before 1952, the mortality for jejunoileal atresia alone was 90%. Between 1952 and 1955, the mortality was 80% when primary anastamosis was performed without bowel resection. Between 1955 and 1958, the mortality decreased to 22% due to liberal resection of the dilated loop and primary anastomosis. From 1959 to 2000, the mortality has decreased to 10%. Factors contributing to mortality were type III atresia, proximal bowel infarction, peritonitis, anastomotic leaks, missed distal atresias, short bowel syndrome, sepsis, and human immunodeficiency virus (HIV) infection.
Outcome/effect	The survival rate is more than 90% in well-resourced countries, but 40–50% in Africa, where no nutritional support is available and presentation is late.
Historical significance/comments	No advancement on vascular theory for aetiology. A variant of multiple intestinal atresias may have a familial/genetic cause.

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 ischaemic 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. One-third of infants with intestinal atresia are born prematurely.

4. Differential diagnoses include midgut volvulus, meconium ileus, extensive aganglionosis, and intussusception.

5. The primary surgery for intestinal atresia 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. The mortality rate depends on birth weight, residual bowel length, the degree of dysmotility, associated anomalies, and septic complications.

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