Jejuno-Ileal Atresia

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64.1 Introduction

Mural defects causing anatomical discontinuity or narrowing of the small bowel can morphologically be divided into either atresia or stenosis and cause about a third of all cases of neonatal intestinal obstruction. Atresia refers to the complete occlusion of the intestinal lumen and accounts for more than 90% of cases, while stenosis is defined as a partial intra-luminal occlusion with incomplete obstruction often resulting in delay in diagnosis until outside the neonatal period. There is a tendency to be born prematurely or small for dates. Other congenital anomalies outside the gastrointestinal tract are uncommon. In a small number of cases, a genetic cause has been identified. Antenatal and early postnatal diagnoses, along with improvements in neonatal care, have been important in the current overall survival of over 90% of newborns with jejunoileal atresia in most centres. However, the change in surgical technique from an anastomosis of the blind-ending bulbous proximal atresia to the distal bowel, to resection of the dilated bowel and primary end to end anastomosis, has been the prime factor in the dramatic improvement in survival. It was Louw, Nixon and Benson who led the way, all changing their technique more or less over the same time period but for different reasons, which led to one of the great success stories of paediatric surgery (Benson 1955; Louw 1959; Nixon 1960). Additional innovative surgical techniques to maximize the utilization of short residual bowel length have further improved long-term outcomes (Bianchi 1980; Kimura et al. 1996; Kim et al. 2003).

64.2 Historical Overview

Significant changes in surgical techniques and outcome of intestinal atresia and stenosis have occurred since Spriggs, in 1912, postulated that strangulation of a segment of foetal gut was most likely the causative factor producing intestinal atresia (Spriggs 1912). Clinical and morbid anatomical data were presented by Louw 1952 and subsequently proven by Barnard and Louw in canine experiments, which produced anomalies identical to congenital intestinal atresia in humans (Louw 1952; Louw and Barnard 1955; Louw et al. 1981a, b). The clarification of the vascular hypothesis and realization that the proximal dilated bowel remained with dysfunctional peristalsis directly influenced and brought the change to the surgical approach from previous attempts at surgical management, which included anastomosis of the bulbous blind end to the distally col-

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lapsed bowel, exteriorization of both ends as an initial stomas, Bishop-Koop, Santulli, or side-to-side anastomosis, to liberal back resection of the proximal blind-ending bulbous atretic bowel to the bowel of near normal diameter and primary end-to-end anastomosis of proximal and distal bowel with more equal lumen diameter (Benson 1955; Louw 1959; Nixon 1960; Nygaard 1967; Grosfeld et al. 1979). This change in technique during the mid-1950s resulted in the elimination of blind-loop syndromes and anastomotic dysfunction with an immediate reduction in mortality rates from 69% to 33% at Great Ormond Street, London, and 90–28% at the Red Cross War Memorial Children's Hospital, Cape Town (Louw 1966). Subsequent technical advances, improvement in neonatal care, anaesthesiology and nutritional support have further impacted upon and improved current day survival to over 90% (Tables 64.1 and 64.2) (Cywes et al. 1982).

Table 64.1 Types of 351 cases with intestinal atresia seen from 1959 to 2017 at Red Cross War Memorial Children's Hospital, Cape Town, South Africa

Type	Jejunum	Ileum	Total (%) (%)(%)(5)(%)
Stenosis	21	13	34 (9)
Type I	64	20	84 (24)
Type II	20	13	33 (9)
Type III(a)	28	24	52 (15)
Type III(b)	65	0	65 (19)
Type IV	69	14	83 (24)
Total	267	84	351

Table 64.2 Chronological improvement in outcomes with changes in surgical techniques, as well asimproved anaesthetic and postoperative care of neonates

Authors	Years of study	*N*	Survival (%)
Evans	1950	1498	9.3
Gross	1940–1952	71	51
Benson et al.	1945–1959	38	55
De Lorimer	1957–1966	587	65
Nixon and Tawes	1956–1967	62	62
Louw	1959–1967	33	94
Martin and Zerella	1957–1975	59	64
Cywes et al.	1959–1978	84	88
Danismead et al.	1967–1981	101	77
Smith and Glasson	1961–1986	84	61
Vecchia et al.	1972–1997	128	84
Rode et al.	1959–2007	318	92
	1990–2007	115	94

64.3 Incidence

Jejunoileal atresia has a prevalence rate of approximately 0.54–1.1 per 10,000 live births, with a third of infants either born prematurely or small for dates (Best et al. 2012). The male-to-female ratio is equal in most reports. Hereditary forms and familial patterns of atresia are exceptionally rare and may be on a basis of autosomal recessive or dominant transmission. A genetic basis, however, has been established for types III (b) and IV multiple atresias (Mishalany and Najjar 1968; Puri and Fujimoto 1988; Lambrecht and Kluth 1998). Associated chromosomal and extra-abdominal anomalies (7%) are well documented but not as common as in duodenal atresias.

64.4 Etiopathogenesis

A localized intrauterine vascular accident can cause ischaemic necrosis, liquefaction of tissues and subsequent resorption of the affected devitalized segment(s). The ischaemia hypothesis is supported by evidence of incarceration or snaring of bowel in an exomphalos or gastroschisis and foetal events such as intussusception, midgut volvulus, trans-mesenteric internal herniation and thromboembolic occlusion resulting in atresia. Meconium ileus and Hirschsprung's disease should also be considered as possible underlying etiological factors in ileal atresia (Santulli and Blanc 1961).

Additional hypotheses based on careful clinical observations, morphological studies and experimentation include Tandler's concept of failure of recanalization of the solid cord stage of intestinal development, obliterative embryological events at Meckel's point with excessive resorption of the vitelline duct with adjacent ileum, epithelial occlusions and foetal inflammatory diseases. The localized nature of the defect would explain the low incidence of coexisting abnormalities of the extra-abdominal organs. Jejunoileal atresias have rarely been described in identical twins. A rare autosomal recessive pattern of transmission has been documented, and

pathological findings could support the concept that a developmental process early on could have affected the whole bowel. This occurrence is termed hereditary multiple intestinal atresia (HMIA) and is associated with combined immunodeficiency and morphological alterations in other organs such as the thymus, lung, spleen and liver, leading to a poor prognosis and is quite different to the previously described atresias (Lambrecht and Kluth 1998).

64.5 Pathophysiology

With the development of the obstruction in the developing foetus, the proximal bowel becomes dilated as a result of onward peristalsis and pressure of luminal contents (Nixon and Tawes 1971). This dilatation may extend proximally for a variable length and in jejunal atresias may extend into the duodenum as well. The distal bowel is usually collapsed and wormlike in appearance. The histopathology of the bowel is usually normal apart from some attenuation of muscle and nerves in the proximal dilated segment. Secondary ischaemia, necrosis and bowel perforation may occur at the atresia either from raised intraluminal pressure from postnatal swallowed air and liquids or from volvulus of the bulbous blind end. Thus, a delay in diagnosis may contribute to morbidity and was a frequent cause of mortality in historical series (Louw et al. 1981a, b).

64.6 Pathology

The morphological classification of jejunoileal atresia into types I–IV has significant prognostic and therapeutic implications (Martin and Zerella 1976; Grosfeld et al. 1979). The level of the most proximal atresia determines whether it is classified as jejunal or ileal. Although single atresias are most commonly encountered, 6–12% of infants will have multiple atretic segments, and up to 5% may have an additional colonic atresia. The appearance of the atretic segment is determined by the type of occlusion, but in all cases a maximum dilatation of the proximal bowel occurs at the site of the obstruction where the bowel is often dysperistaltic and of questionable viability, especially when treatment is delayed. In rare instances, jejunoileal atresias have been found to coexist with oesophageal, duodenal, colonic or rectal atresias. The incidence of the different types seen in 351 at our centre over a 58-year period is shown in Table 64.1.

- *Stenosis* (<9%) is characterized by a short localized narrowing of the bowel without discontinuity or a mesenteric defect. Although more frequently seen with duodenal fenestrated webs a 'wind-sock' effect can be created when increased intraluminal pressure in the proximal bowel bulges the membrane into the distally collapsed bowel creating a conical transition zone. The bowel is of normal length.
- *Atresia type I* (24%) is represented by a transluminal septum or short atretic segment. The dilated proximal bowel remains in continuity with the collapsed distal bowel, there is no mesenteric defect, and the bowel is of normal length (Fig. 64.1).

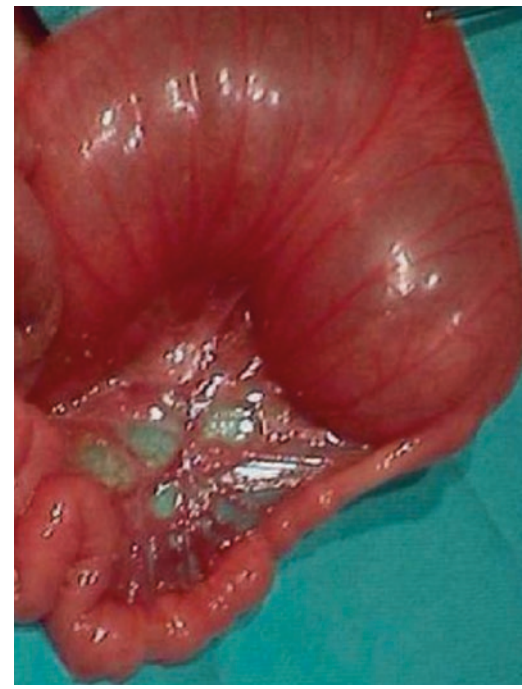

Fig. 64.1 Typical clinical picture of jejunal atresia type I

- *Atresia type II* (9%) has two blind-ending atretic ends connected by a fibrous cord along the edge of the mesentery. There is no mesenteric defect and the bowel length is not foreshortened.
- *Atresia type IIIa* (15%) is similar to type II except that the fibrous connecting cord is absent and there is a V-shaped mesenteric defect. The bowel length may be foreshortened. Cystic fibrosis is commonly associated with this variety.
- *Atresia type IIIb* (19%) (apple peel or Christmas tree) consists of a proximal jejunal atresia often with associated malrotation, absence of most of the superior mesenteric artery and a large mesenteric defect. The distal bowel is coiled in a helical configuration around a single perfusing artery arising from the right colic arcades(Fig. 64.2). Additional type I or type II atresias may be found in the proximal and distal bowel. There is always a significant reduction in intestinal length. A familial incidence and atresias amongst siblings and identical twins point to a more complex genetic transmission with an overall recurrence rate of 18% in subsequent siblings born to the same parents.

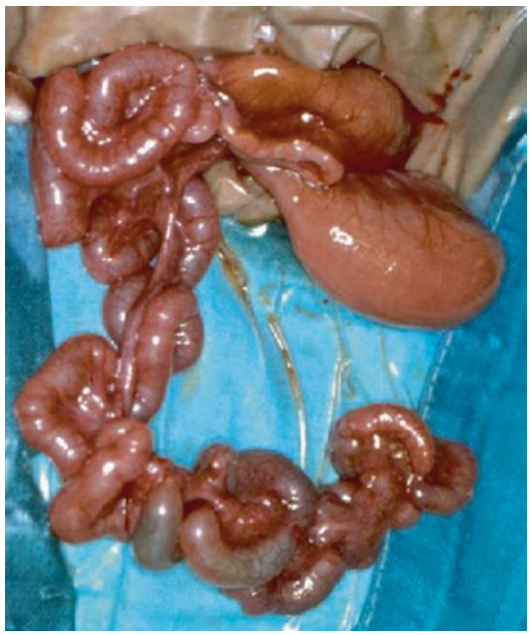

Fig. 64.2 Type IIIb (apple peel) atresia

- *Atresia type IV* (24%) represents multiple segmental atresias like a string of sausages or a combination of types I–III. Bowel length is always reduced. The terminal ileum is usually spared; up to 25 atretic sites have been encountered in a single patient.

64.7 Management

64.7.1 Clinical Presentation and Diagnosis

A delay in diagnosis may lead to impairment of bowel viability or frank necrosis and perforation, fluid and electrolyte abnormalities and an increase incidence of sepsis (Figs. 64.3 and 64.4). Antenatal perforation of atretic bowel may present at birth with meconium peritonitis. The differentiation between atresia and other forms of intrinsic and extrinsic bowel obstruction due to

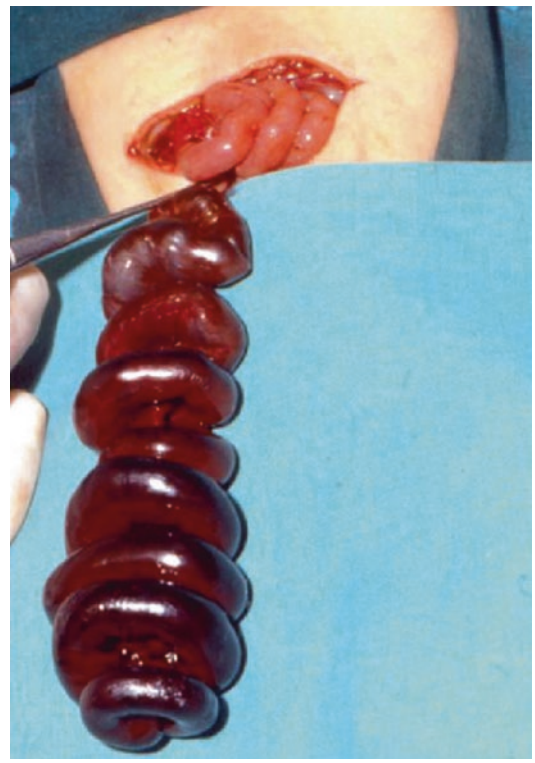

Fig. 64.3 A gangrenous type IIIb (apple peel) jejunal atresia due to volvulus at its base where the single marginal artery supplies ileum, which is coiled in an apple peel configuration

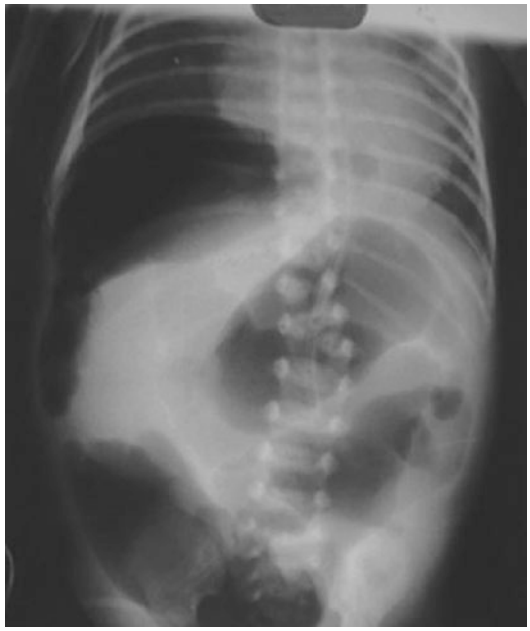

Fig. 64.4 Abdominal radiograph of a newborn showing a pneumoperitoneum due to proximal bowel perforation

volvulus or internal hernia is the most important consideration that requires early diagnosis. Many cases of intestinal atresia are now diagnosed prenatally by ultrasonographic investigation of the foetus, showing dilated intestine with vigorous peristalsis, suggesting obstruction, particularly so in pregnancies complicated by third trimester polyhydramnios (Wax et al. 2006). Intestinal atresia is also suspected in foetuses with gastroschisis and evidence of intestinal dilatation. However, prenatal ultrasound has a relatively poor predictive value for bowel abnormalities (31–42%). MRI imaging may prove to be more accurate in the prenatal diagnosis of bowel atresia (Veyrac et al. 2004). A positive family history will help identify hereditary forms. As the prognosis of intestinal atresia is excellent, there is no need for intrauterine intervention or early induction of labour before 38 weeks of gestation.

After birth, intestinal atresia or stenosis can present early with large intragastric volumes at (>25 ml gastric aspirate) followed by persistent bile stained vomiting, although in 20% of children, symptoms may be delayed for more than 24 h. Abdominal distension is frequently present at or soon after birth; the more distal the obstruction, the more generalized the abdominal distension. Visible loops of bowel may be observed, but peristaltic activity is an inconsistent finding. Proximal jejunal atresia often presents with gastric distension, one or two loops of visible bowel in the upper abdomen decompressable by nasogastric tube aspiration in an otherwise gasless abdomen. Although the classic first stools passed by these patients are small, grey in colour and mucoid, normal meconium can occasionally be passed, which would suggest a late gestation insult leading to the development of atresia. Delay in detection, increasing intra-luminal pressure and/or secondary torsion of the proximal atretic distended bowel can lead to ischemia, perforation and peritonitis, precipitating abdominal tenderness, with oedema and erythema of the abdominal wall. One-third of infants can have biochemically determined nonhaemolytic jaundice.

The diagnosis is confirmed by radiological examination of the abdomen and chest (Fig. 64.5).

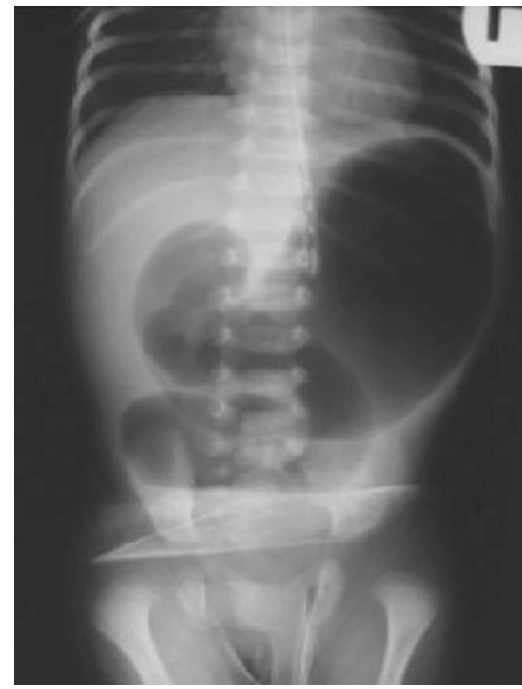

Fig. 64.5 Abdominal radiograph of a newborn with jejunal atresia showing a few dilated proximal small bowel loops

Erect and supine whole-body chest/abdominal radiographs done after birth will reveal distended air-filled small intestinal loops proximal to an obstruction with no gas visible in the rectum. In some instances, the first abdominal radiograph can reveal a completely opaque abdomen due to fluid-filled obstructed bowel. Emptying the stomach via a nasogastric tube and the injection of a bolus of air will demonstrate the obstruction without resorting to a contrast radiography.

The more distal the obstruction, the greater are the number of air- and fluid-filled, distended loops of bowel (Fig. 64.6). The bowel immediately proximal to the site of obstruction may have the appearance of a very large air- and fluid-filled loop. A prone lateral view may distinguish between distal small bowel and colonic obstructions. Occasionally scattered intra-peritoneal or scrotal calcification or a large meconium pseudocyst or hydrocele may be encountered or seen radiographically, signifying intrauterine bowel perforation, meconium spill with dystrophic calcification.

In the presence of a radiologically determined complete obstruction, a contrast enema can be performed to confirm the level of obstruction (small or large bowel), document the calibre of the colon, exclude an associated colonic atresia and locate the position of the caecum as an indication of malrotation (Fig. 64.7). Care must be taken not to rupture the microcolon, as the latter signifies an unused colon with the obstruction located in the small bowel. Where the atresia is formed late in intrauterine life, the bowel distal to the atresia has a more normal calibre. With incomplete proximal small bowel obstruction, an upper gastrointestinal contrast study is indicated to demonstrate the site and nature of the obstruction and to exclude midgut volvulus.

The clinical and radiological presentation of jejunoileal stenosis will be determined by the level and degree of stenosis. The diagnosis is often delayed for several weeks to months, and investigations may be inconclusive due to subclinical symptoms and findings.

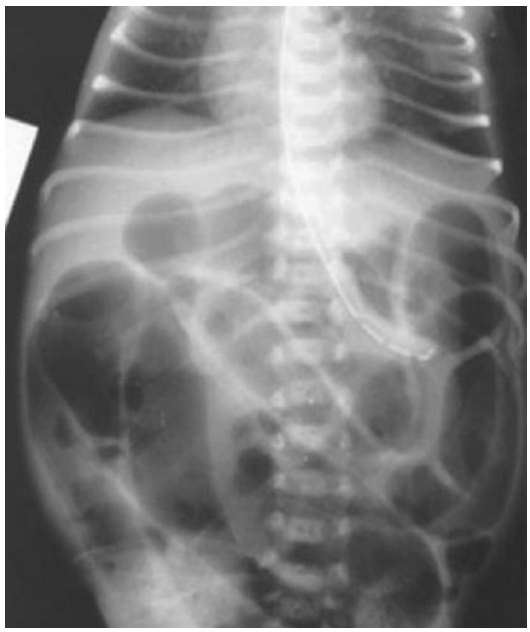

Fig. 64.6 Abdominal radiograph of a newborn with distal small bowel stenosis showing large air-filled intestinal loops

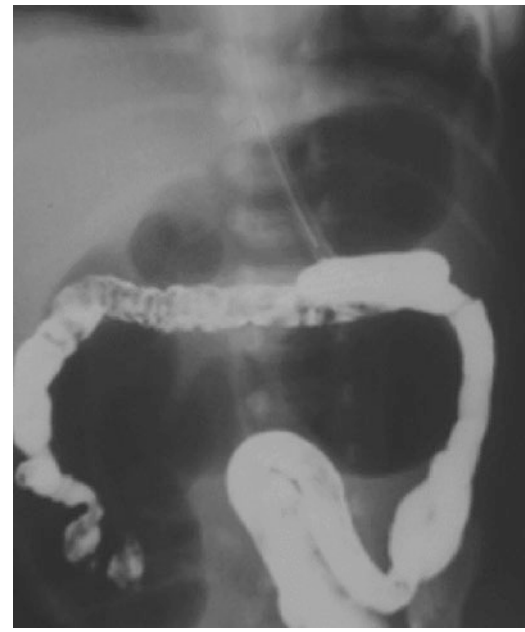

Fig. 64.7 Contrast enema confirming a normally rotated and patent colon with evidence of proximally dilated small bowel loops

64.7.2 Differential Diagnosis

A spectrum of other diseases can present with symptoms and signs of neonatal intestinal obstruction closely mimicking jejunoileal atresia. These include atresia of the distal duodenum, colonic atresia and long-segment Hirschsprung's disease as well as those with intraluminal contents causing obstruction such as the inspissated mucous pellets found with meconium ileus and hypoperistalsis as in small left colon syndrome and meconium plug syndrome. Extrinsic factors like midgut volvulus and incarcerated hernia and nonmechanical (functional) causes such as ileus from sepsis or early necrotizing enterocolitis should also be considered.

64.7.3 Surgical Management: Making the Infant 'Safe for Surgery'

These babies must be resuscitated and haemodynamically stabilized before transfer. Preoperative management is directed at optimizing the status of the infant, which apart from the atresia could be compromised by delayed diagnosis, hypothermia, fluid and electrolyte imbalance, associated abnormalities, prematurity, obstetrical-related complications, systemic sepsis and compromised bowel. Basic factors required include a warm humidified environment, gastric decompression to prevent aspiration, fluid management (maintenance, replacement of deficits, and ongoing losses), correction of haematological and biochemical abnormalities and prophylactic antibiotics.

64.7.4 Anaesthesia

Neonates tolerate surgery well provided their special needs, physiological limitations and disease processes are duly taken into account. The major anaesthetic considerations are related to prematurity, fluid and electrolyte homeostasis, abdominal distension, the risk of aspiration and associated life-threatening congenital anomalies. Invasive monitoring is indicated in sick or unstable infants, and a central line may be required for intravenous feeding postoperatively. The anaesthetic management is dictated by the condition of the infant and the available facilities. Light general and epidural anaesthesia may avoid the need for postoperative ventilation.

64.7.5 Surgical Strategy

The operative management of intestinal atresia is individualized and determined by the pathologic findings, associated abnormalities, the length of the undamaged bowel and the general condition of the infant (Louw 1966; Millar and Cox 2020).

64.7.5.1 Standard Surgical Procedure

A supra-umbilical transverse incision provides excellent exposure of the entire gastrointestinal tract. A laparoscopic or periumbilical approach may also be used with exteriorization of the atretic segment through a small umbilical incision (Banieghbal and Beale 2007; Lima et al. 2009; Murphy et al. 2009). The bowel is exteriorized to determine the site and type of atresia and to exclude further downstream atresias or stenoses and associated lesions such as incomplete rotation or meconium ileus. The appearance of the atretic segment depends upon the type of occlusion, but in all cases, maximum dilatation and hypertrophy of the proximal bowel occurs at the point of the first obstruction. This segment often has reduced peristaltic activity and is of questionable viability, whilst the bowel distal to the obstruction is collapsed, tiny and wormlike in appearance. The intestinal content in more proximal atresias is milked backwards into the stomach from where it is aspirated. In the more distal atresias, the intestinal contents can be milked into the bulbous segment to be resected. If a volvulus is present, it should be derotated. It is essential to exclude other distal small and large bowel atresias by injecting normal saline down the lumen of the distal bowel. More than one atresia can occur in up to 20% of cases. The total length of small bowel is then accurately measured along the antimesenteric border because residual bowel length has prognostic significance and will deter-

mine the method of reconstruction. If the total usable bowel length is deemed adequate (>80 cm + ileocaecal valve), 5–15 cm bulbous hypertrophied proximal bowel is resected back to the near normal calibre of the bowel. In performing the resection, one should preserve as much of the mesentery as possible for later use to fill in any gaps in the small bowel mesentery left after end-to-end anastomosis. Having decided on the site of transection, the bowel is then divided at right angles leaving an opening of approximately 0.5–1.5 cm in diameter. The blood supply should be adequate to ensure a safe anastomosis. If, however, an extensive cutback resection is contraindicated because of insufficient residual bowel length, the bulbous portion alone or any compromised bowel should be resected. The proximal bowel should then be tapered obliquely hand-sewn or using a GIA stapler leaving the proximal bowel opening a similar size to the distal bowel lumen to facilitate an end-to- end primary anastomosis.

Proximal bowel resection is followed by very limited distal small bowel resection over a length of 2–3 cm. The resection line should be slightly oblique towards the anti-mesenteric border (fish-mouth) to ensure that the openings of the proximal and distal bowels are of approximately equal size to facilitate easy end-to-end or rarely an end-to-back (Denis-Browne) single-layer anastomosis; 5/0 or 6/0 absorbable suture material is used (Fig. 64.8). Alternatively, an extra-mucosal end-to-end anastomosis can be performed, placing sutures on the proximal bowel further apart so as to accommodate the discrepancy in the bowel lumen diameters. The mesentery is approximated with interrupted sutures, which may be difficult with large mesenteric defects. A side-to-side anastomosis should not be performed as it can lead to a blind loop syndrome. There is little place for routine gastrostomy or transanastomotic feeding tubes where facilities for parenteral nutrition are available.

64.7.5.2 Special Considerations

Atresia type I and stenosis are best dealt with by primary resection and end-to-end anastomosis. Procedures such as simple transverse enteroplasties, excision of membranes, bypass techniques or side-to-side anastomosis are no

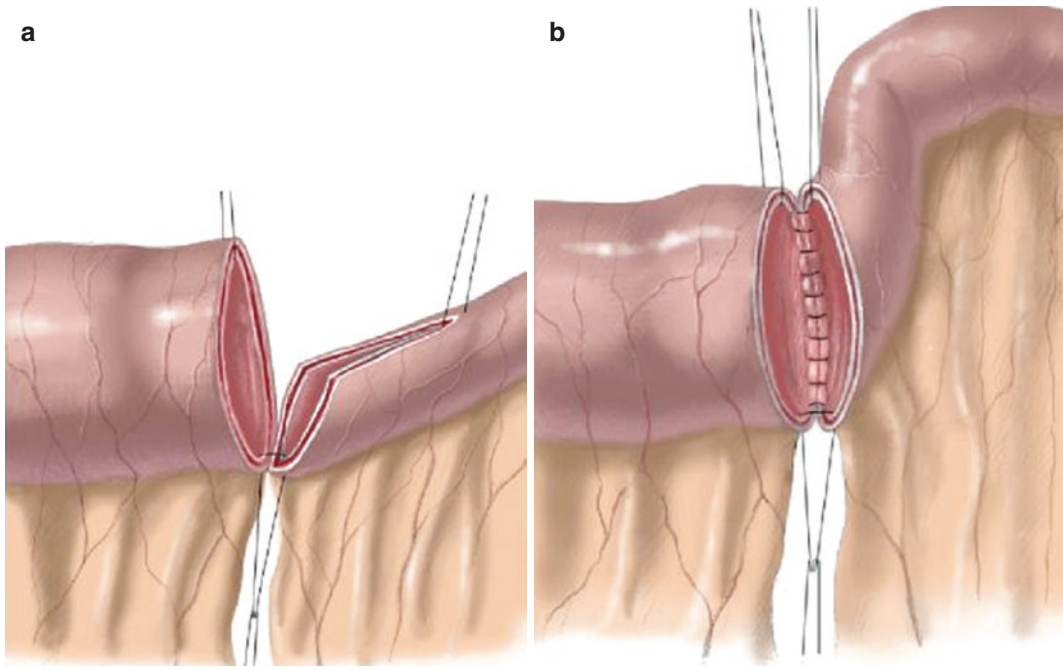

Fig. 64.8 End-to-end (**a**) or end-to-back (**b**) single-layer bowel anastomosis

longer utilized. They fail to remove the abnormal dysfunctional segments of intestine, thus increasing the risk of the blind loop syndrome and dysmotility.

Atresia types II and IIIa are managed in the same manner as type I with back resection and primary end-to-end anastomosis. The conservation of bowel length is mandatory.

Multiple membranous diaphragms (*type I atresias*) can be successfully perforated by transluminal bouginage done along the entire length of the affected small bowel (Romao et al. 2011).

High jejunal atresia: With type IIIb or high jejunal atresia, the proximal bowel should be derotated, the ligament of Treitz, if present, should be taken down, and resection of the bulbous portion may be extended into the second part of the duodenum taking care to stay well clear of the ampulla of Vater. This is followed by an *antimesenteric tapering duodenojejunoplasty* (Kling et al. 2000). Bowel tapering can safely be done over a length of between 20 and 35 cm. This is done to conserve bowel length, to reduce disparity in anastomotic size and to improve duodenal and proximal jejunal prograde peristaltic function. At completion, the bowel is left in a position of derotation with the duodenum-jejunum positioned on the infant's right side, the mesentery broad-based and the caecum lying anterior, to the left of the midline in the upper abdomen. These additional maneuvers induce rapid return of prograde intestinal function, and the neonates are usually able to tolerate graded to full oral intake within 14 days.

With type IIIb atresias, the distal 'apple peel' component of the bowel is gently laid out so that the inner margin of mesentery is clearly visible from its right colon origin to the atretic blind end. The division of avascular restricting mesenteric bands along the free edge of the distally coiled narrow mesentery may be required, thereby releasing kinking and interference with the bowel blood supply. The large mesenteric defect may be left open, but where possible the preserved mesentery can be used to fill in the defect. Furthermore, to prevent kinking of the marginal artery after the completion of the anastomosis, the bowel is replaced carefully into the peritoneal cavity in the position of non-rotation. There is no need to remove the appendix.

Multiple type IV atresias, present in 20% of cases, are often localized to a segment facilitating an en-bloc resection with a single anastomosis in preference to multiple anastomosis. If multiple anastomoses are deemed necessary because of insufficient bowel length, it is useful to railroad each bowel segment to be anastomosed onto a silastic feeding tube before completing the anastomoses to avoid torsion and ensure correct orientation and continuity of the bowel. This tube may be left in situ for a time to serve as a stent until bowel function has returned. It is always important to ensure the preservation of the maximum bowel length to avoid the short bowel syndrome; for this reason the following techniques may be necessary.

Tapering enteroplasty is indicated when the ischemic insult has resulted in an atresia with markedly reduced intestinal length (<80 cm); where long-length resection of abnormal or multiple atretic segments are required; to equalize disparity in anastomotic lumen size; for the correction of a previous failed inversion plication procedure and to improve function in a persistently dilated non-functioning megaduodenum following surgery for upper jejunal atresia (Thomas Jr. and Carter 1974).

The *inversion plication enteroplasty method* has the theoretical advantages of reducing the risk of leakage from a long antimesenteric suture line, conserving mucosal surface area, and may even facilitate the return of bowel peristalsis (Fig. 64.9). More than half of the antimesenteric bowel circumference may be enfolded into the lumen over an extended length without causing an obstruction. The drawback of this method is that the holding suture line can unravel within a few months, precipitating persistent motor dysfunction. As an alternative, antimesenteric seromuscular

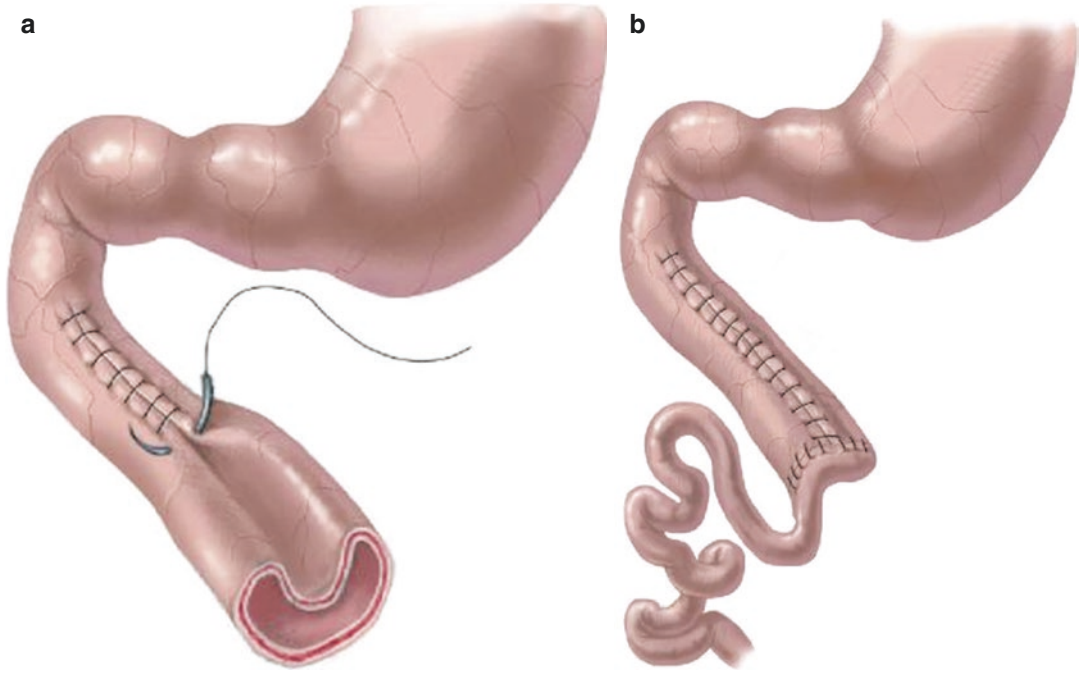

Fig. 64.9 (**a**–**b**) Inversion plication duodenojejunoplasty for conserving length in high small bowel atresia

stripping combined with inversion plication may prevent this complication and preserves maximal mucosal surface for absorption (Kimura et al. 1996).

Intestinal atresia and gastroschisis: The intestinal peel, often encountered in gastroschisis, hinders the detection and surgical management of an associated atresia. Two options are available, namely, the primary resection of the atretic segment with immediate anastomosis, which is only indicated if this can be done when there is little or no peel. If the identification of the atretic segment is difficult and the bowel very oedematous, the best approach is to reduce the eviscerated bowel with the atresia left undisturbed, awaiting resorption of the peel and oedema, when a safe primary resection and anastomosis can be performed, which is usually 14 to 21 days later (van Hoorn et al. 1985).

Exteriorization or stomas: Although primary anastomosis is always preferred, a temporizing proximal stoma or chimnied anastomosis (Bishop-Koop method) or a second-look exploration 48–72 h later may be advisable when the vascular integrity of the intestine is questionable. This may be relevant in cases with late diagnosis, type III (b) atresias or with gross intraperitoneal faecal contamination.

64.8 The Short Bowel Syndrome

Insufficient bowel length either as a result of the primary injury, excessive removal of residual bowel or an ischaemic insult to the remaining bowel as a postoperative complication can lead to the short bowel syndrome with long-term sequelae for growth and development (Wales 2004).

Bowel lengthening procedures are best not performed during the initial surgery. Four currently applicable techniques are longitudinal intestinal lengthening and tapering method or LILT (Bianchi 1980), serial transverse enteroplasty or STEP technique (Kim et al. 2003), spiraling intestinal lengthening and tailoring (SILT) (Cserni 2011, van Praagh 2022) and the myoenteropexy method of Kimura. Although they theoretically increase intestinal length and reduce lumen diameter,

thereby improving both peristalsis and maintaining absorptive capacity, the outcome ultimately depends on residual small bowel length, the presence of the ileocaecal valve and any complicating associated liver disease (Frongia et al. 2013). Maximal bowel adaptation should first be attained utilizing both TPN and enteral feeding as adjunctive methods of therapy before these additional surgical techniques are contemplated.

64.9 Postoperative Care

Standard methods are used. Nasogastric decompression is usually required for 4–6 days after the operation (longer for high jejunal atresias). Therapeutic antibiotics are continued for 5–7 days or longer, and an oral antifungal agent is given prophylactically. Graduated oral intake is commenced when the neonate is alert, sucks well, and there is evidence of prograde gastrointestinal function, i.e. clear gastric effluent of low volume, a soft abdomen and when flatus or faeces have been passed per rectum.

Surveillance for alimentary dysfunction should continue until the infant has established and stabilized normal gastrointestinal function. If at any time there is a suspicion of a leak at the anastomosis (suggested by vomiting, abdominal distention, tenderness and evidence of sepsis), a plain erect or decubitus radiograph of the abdomen should be taken. If this reveals free air in the abdomen more than 24 h after the operation, laparotomy should be performed immediately and the leaking site sutured or the anastomosis redone. Other complications encountered include wound sepsis and dehiscence, anastomotic kinking, ischaemia with late-onset anastomotic stenosis, adhesive obstruction and the short bowel syndrome.

After massive intestinal length loss, TPN or other forms of enteral support should be continued with until maximum bowel adaptation is reached. It is imperative that graduated enteral, preferably breast milk feeding, should be introduced as soon as possible to stimulate and enhance this process, which can take many months (Sigalet 2001).

64.10 Conclusion

Following surgical correction of the anomaly, the majority of children grow and develop normally, the end result being influenced by the residual intestinal length and absorptive and peristaltic function of the residual bowel, associated anomalies and the successful management of the short bowel syndrome.

The quality of life and functional status are usually very good and are not affected by intestinal atresia, although cystic fibrosis, the short bowel syndrome and TPN-induced liver failure may alter prognosis. Factors contributing to the <10% mortality rate include delay in presentation, the type of atresia, proximal bowel infarction with peritonitis, anastomotic dysfunction, missed distal atresias, the short bowel syndrome, pneumonia, sepsis and associated abnormalities.

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