Yechiel Sweed and Arcady Vachyan

Congenital duodenal obstruction is the result of several embryologic defects in foregut development, canalization, or rotation. In addition, abnormal embryologic relationships between the duodenum and other structures in close anatomic proximity, such as the pancreas and portal vein, may also lead to congenital duodenal obstruction. Ladd classified these lesions as either intrinsic or extrinsic. Intrinsic lesions include duodenal atresia, duodenal stenosis, or duodenal web, whereas annular pancreas, malrotation, peritoneal bands, and anterior portal vein are classified as extrinsic.

The incidence of duodenal obstruction is reported to be 1 in 5000–10,000 births. In patients with intrinsic duodenal obstruction, there is a high incidence of associated anomalies, especially Down syndrome, which occurs in about 30% of these patients. Other associated anomalies include congenital heart disease, malrotation, annular pancreas, oesophageal atresia, urinary tract malformations, anorectal anomalies, and other bowel atresias.

25.1 Diagnosis

The diagnosis of duodenal obstruction may be suspected prior to the child's birth from prenatal ultrasonography, which may identify maternal polyhydramnios and demonstrate distension of the stomach and the first portion of the duodenum with swallowed amniotic fluid. Although prenatal ultrasonography is an accurate predictor of duodenal obstruction and allows preparation of parents, physicians, and institutions for the anticipated arrival of the patient needing prompt care at birth, it has neither influenced the incidence

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of associated life-threatening anomalies nor changed the survival rate.

The clinical presentation of duodenal obstruction is usually characterized by feeding intolerance and by onset of vomiting in the first 24–48 h of life. Because 80% of obstructions are located in the postampullary region of the duodenum, the vomitus in most cases is bile-stained. A careful physical evaluation for associated anomalies is performed. Cardiac and renal ultrasonographic examinations are also indicated because of the high incidence of associated malformations in other organ systems.

Diagnosis is achieved in most cases by plain abdominal radiographs, which demonstrate dilated stomach and duodenum, giving the characteristic appearance of a "doublebubble" sign. No gas is observed beyond the second bubble in instances of complete obstruction. In this setting, the plain film is sufficiently diagnostic, and no further imaging of the gastrointestinal tract is necessary. In partial duodenal obstruction, a plain film of the abdomen also will show a "double-bubble" appearance, but there is usually some air in the more distal intestine. Early upper gastrointestinal contrast radiography is indicated in these patients in order to establish the cause of incomplete duodenal obstruction.

Incomplete duodenal obstruction may lead to delayed onset of symptoms, and the diagnosis of duodenal diaphragm with a central aperture is sometimes delayed for months or years.

25.2 Preoperative Management

Although duodenal atresia is a relative emergency, the infant should not be rushed to the operating room until his or her haemodynamic and fluid and electrolyte status is stable. If the clinical history and findings on physical examination indicate that the baby is in no distress, and the radiograph is consistent with the usual presentation of duodenal atresia with no air beyond the second bubble, surgery should be per-

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formed within the first 2 days of life. Immediate surgical exploration should be performed, however, in patients with duodenal obstruction caused by malrotation resulting in extrinsic compression related to Ladd's bands across the duodenum or acute volvulus of the midgut.

Preoperative management consists of nasogastric decompression and fluid and electrolyte replacement. Care is taken to preserve body heat and avoid hypoglycaemia, as most of these newborn patients are premature or small for date. Preoperative systemic antibiotics are administered at least 30 min before the start of the operation.

25.3 Surgical Management

For most causes of congenital duodenal obstruction, duodenoduodenostomy via an open approach is the traditional surgical procedure. Over the past decade, however, the application of minimally invasive surgical (MIS) techniques and the advent of smaller laparoscopic instruments have expanded the potential of laparoscopy for repair of this obstruction.

25.3.1 Open Approach to Duodenal Obstruction

The baby is placed supine on the table, on a warming blanket, with a small roll under the upper abdomen. Endotracheal anaesthesia is used. A nasogastric tube is passed to decompress the stomach. An intravenous infusion is set up. The abdominal skin is prepared by cleaning with prewarmed povidone iodine.

A transverse supraumbilical abdominal incision is made 2 cm above the umbilicus, starting in the midline and extending laterally into the right upper quadrant (Fig. 25.1). A small incision is made in the posterior fascia and peritoneum after these are drawn up with forceps. To enlarge this initial incision, two fingers are inserted and the fascia and peritoneum are cut along the length of the wound. The underlying structures are retracted.

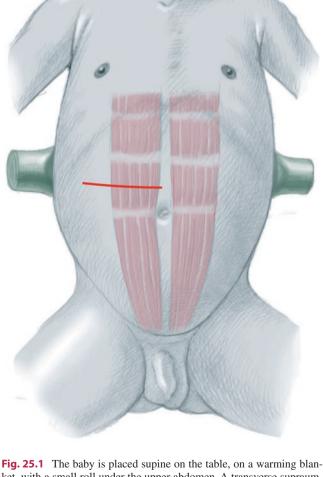


Fig. 25.1 The baby is placed supine on the table, on a warming blanket, with a small roll under the upper abdomen. A transverse supraumbilical abdominal incision is made 2 cm above the umbilicus, starting in the midline and extending laterally into the right upper quadrant

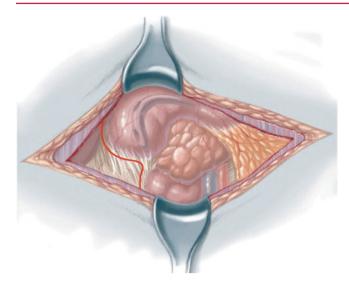


Fig. 25.2 After exposing the peritoneal cavity, the surgeon inspects the entire bowel for the presence of other anomalies. The ascending colon and the hepatic flexure of the colon are mobilized medially and downwards to expose the dilated duodenum. The duodenum is then adequately mobilized and freed from its retroperitoneal attachments (Kocher manoeuvre) (*red line*). Great care must be exercised not to dissect or manipulate either segment of the duodenum medially, to avoid injury to the ampulla of Vater or the common bile duct. The duodenum distal to the site of obstruction is small and decompressed. The requirements for distal mobilization vary according to the location of the atresia and to the gap between the two segments. If necessary, the ligament of Treitz is divided, and mobilization and displacement of the distal duodenum is performed behind the superior mesenteric vessels, thus allowing a satisfactory anastomosis to be performed without any tension

After exposing the peritoneal cavity (Fig. 25.2), the surgeon inspects the entire bowel for the presence of other anomalies. There may be an associated annular pancreas or malrotation in one third of the patients. If the colon is in a normal position, malrotation is probably not a coexisting factor. The stomach and first portion of the duodenum are usually thickened and dilated. The liver is carefully retracted superiorly. The ascending colon and the hepatic flexure of the colon are mobilized medially and downwards to expose the dilated duodenum.

The duodenum is then adequately mobilized and freed from its retroperitoneal attachments (Kocher manoeuvre). Great care must be exercised not to dissect or manipulate either segment of the duodenum medially, to avoid injury to the ampulla of Vater or the common bile duct. The tube in the stomach is then passed distally into the dilated duodenum and helps to locate the point of obstruction and determine if a "windsock" abnormality is present.

The type of atresia as well as any pancreatic abnormality (annular pancreas) or the presence of a rare preduodenal portal vein are noted. In patients with an annular pancreas, the pancreatic tissue should never be divided and should be bypassed. The duodenum distal to the site of obstruction is small and decompressed. The requirements for distal mobilization vary according to the location of the atresia and to the gap between the two segments. If necessary, the ligament of Treitz is divided, and mobilization and displacement of the distal duodenum is performed behind the superior mesenteric vessels, thus allowing a satisfactory anastomosis to be performed without any tension.

Duodenoduodenostomy is the procedure of choice for patients with duodenal atresia, stenosis, and annular pancreas. Two surgical techniques—side-to-side duodenoduodenostomy or proximal transverse-to-distal longitudinal ("diamond-shaped") anastomosis—may be performed.

25.3.2 "Diamond-Shaped" Anastomosis

Diamond-shaped duodenoduodenostomy has been reported to allow earlier feeding, earlier discharge, and good longterm results. With two traction sutures, the redundant wall of the proximal duodenum is pulled downward to overlie the proximal portion of the distal duodenal segment (Fig. 25.3). A transverse incision in the distal end of the proximal duodenum and a longitudinal incision in the smaller limb of the duodenum distal to the occlusion are made so as to allow good approximation of the openings without tension (Fig. 25.4) and distal atresias are ruled out. The papilla of Vater is located by observing bile flow from gentle compression of the gall bladder.

A single-layer anastomosis is created using Vicryl sutures (Fig. 25.5). Before completion of the anastomosis, a transanastomotic feeding tube may be passed down into the upper jejunum for an early postoperative enteral feeding.

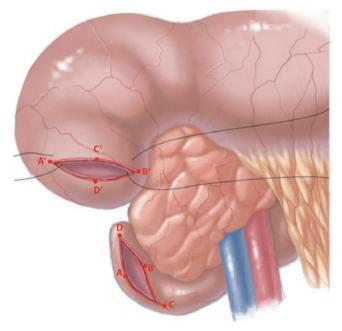


Fig. 25.3 To perform a diamond-shaped anastomosis, two traction sutures in the redundant wall of the proximal duodenum are used to pull it downward to overlie the proximal portion of the distal duodenal segment. A transverse incision is made in the distal end of the proximal duodenum and a longitudinal incision is made in the smaller limb of the duodenum distal to the occlusion. These are made in such a position as to allow good approximation of the openings without tension

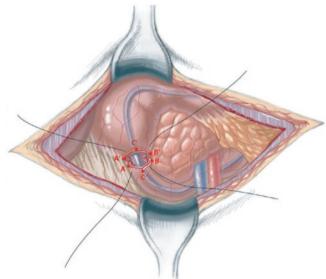


Fig. 25.4 The orientation of the sutures in the diamond-shaped anastomosis and the overlapping between the incisions. At this stage, a small Nélaton catheter is passed distally through the opening made in the distal segment, and 20–30 mL of warm saline is injected to rule out atresias distally. The catheter is then removed

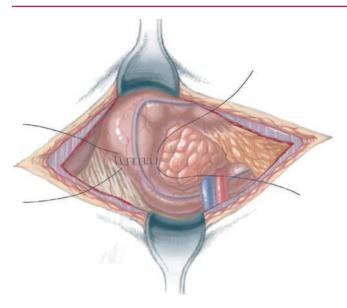


Fig. 25.5 A single-layer anastomosis using interrupted 5/0 or 6/0 Vicryl sutures with knots tied inside the posterior wall of the anastomosis and interrupted sutures with anterior knots tied outside the anterior wall. Before completion of the anterior part of the anastomosis, a transanastomotic feeding tube (5F silicone) may be passed down into the upper jejunum for an early postoperative enteral feeding

25.3.3 Repair of Duodenal Web

When abdominal exploration yields a diagnosis of duodenal web (identified by the advancement of the gastric tube into the proximal dilated duodenum), the dilated duodenal wall is incised (Fig. 25.6) and the duodenum is opened. The windsock duodenal web must be clearly identified because the visible transition from the distended proximal duodenum to the small downstream duodenum may be several centimetres distal to the base of the web. Traction applied at the apex of the web deforms the duodenum at its point of attachment and allows excision at the base (Fig. 25.7).

The duodenal membrane is usually localized in the second part of the duodenum, and occasionally in the third portion. It can be complete or may have a central hole. Anatomically, the ampulla of Vater may open directly into the medial portion of the web itself—anteriorly, posteriorly, or with dual openings into the membrane—or it may open close to it.

This close relationship mandates the identification of the papilla of Vater before excision of the web. The web is opened along the lateral side of the membrane and excision from the duodenal wall takes place (Fig. 25.8). An intermittent bile flow is usually seen via the papilla of Vater,

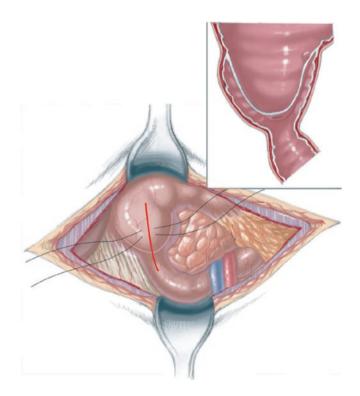
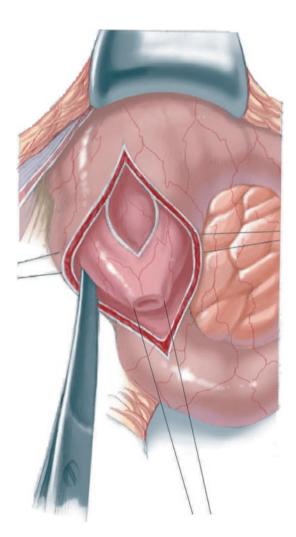


Fig. 25.6 To repair a duodenal web (identified by the advancement of the gastric tube into the proximal dilated duodenum), two stay sutures are placed at the anterior dilated duodenal wall. A longitudinal incision of 2.5–3 cm (*red line*) is performed above the "transitional zone" between the wide and the narrow segments of the duodenum, and the duodenum is opened

indicating to the surgeon the exact line of excision. The resection line is then oversewn and the duodenum is closed transversely (Fig. 25.9). Because of the pitfalls in cases of lax membrane that may bulge downwards distally into the distended duodenum (the so-called windsock phenomenon), and in order to avoid missing the anomaly, the patency of the distal duodenum must be identified by inserting a catheter through the duodenotomy before its closure.

Following completion of the web resection and closure of the duodenum, the abdominal cavity is irrigated with 50 mL of sterile warm saline. The wound is closed in layers: the peritoneum and the posterior fascia and the anterior fascia by two layers using continuous 4/0 Dexon or Vicryl sutures. The skin is closed with a running intracuticular suture using 5/0 Vicryl or Dexon suture.



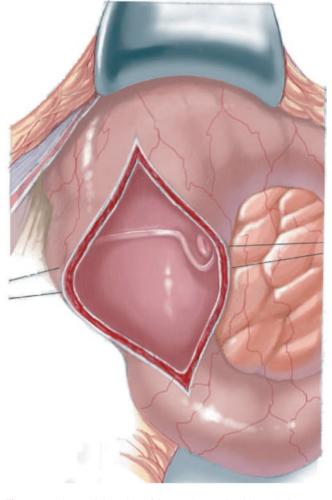


Fig. 25.8 The medial portion of the membrane should remain intact, to avoid damage to the ampulla of Vater

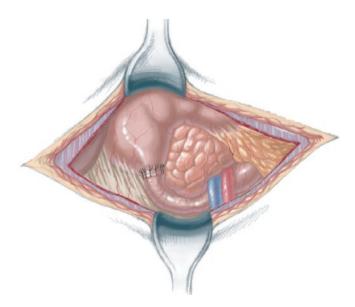


Fig. 25.7 Two other stay sutures are placed at the margins of the duodenal incision. Traction applied at the apex of the web deforms the duodenum at its point of attachment and allows excision at the base. A single 4/0 Vicryl stay suture is placed at the centre of the membrane. The web is opened along the lateral side of the membrane and excision from the duodenal wall takes place, leaving a rim of tissue of 2–3 mm

Fig. 25.9 The resection line is oversewn using interrupted 5–0 absorbable sutures. The duodenum is then closed transversely with interrupted sutures

A nasogastric tube is left in place for postoperative gastric drainage. A gastrostomy may be performed if the need is anticipated. Intravenous therapy and antibiotics are continued postoperatively. The patient is kept without oral intake until stool is passed and limited clear or pale-green gastric drainage is noted (<1 mL/kg per h). The commencement of oral feeding may be delayed for several days and occasionally for 2 weeks or more. Postoperatively, patients may have a prolonged period of bile-stained aspirate, mainly because of the inability of the markedly dilated duodenum to produce effective peristalsis. Many surgeons therefore use transanastomotic tubes for feeding in the early postoperative period.

25.3.4 Laparoscopic Management of Duodenal Obstruction

For the laparoscopic approach, neonatal (3-mm) laparoscopic instruments and trocars are used. The patient is positioned supine at the end of the operating table and the surgeon stands at the patient's feet. The abdomen is insufflated and then two additional trocars are inserted (Fig 25.10). A 3-mm grasping forceps for lifting the liver can be also introduced in the left upper quadrant without a trocar. A better view of the dilated duodenum can be also achieved by using a suture to lift up the falciform ligament. The suture is inserted through the abdominal wall in the right upper quadrant, lifts the ligament, and then is passed back through the abdominal wall and tied.

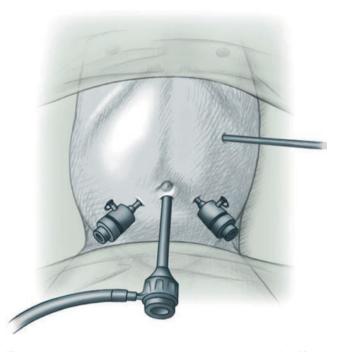
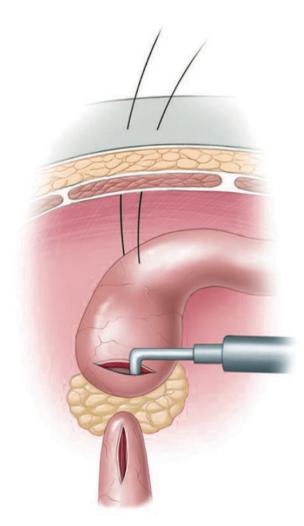


Fig. 25.10 For the laparoscopic approach, the abdomen is insufflated with CO_2 (6 mmHg, 2 L/min) through a 5-mm umbilical port for a 30° laparoscope. Then two additional 3-mm trocars are inserted under direct vision in the right lower quadrant and left middle abdomen. A 3-mm grasping forceps for lifting the liver can be also introduced in the left upper quadrant without a trocar

The first surgical step is to mobilize the colon and the duodenum. A stay suture is inserted through the abdominal wall to move the bulky part of the bulbus duodeni out of the way, allowing a view of the distal duodenum and a more convenient approach to the anastomosis. A transverse incision is made in the distal wall of the dilated duodenum (Fig. 25.11), followed by a longitudinal incision in the distal collapsed duodenum for the "diamond-shaped" anastomosis of Kimura. As with the open repair, stay sutures are placed at each corner to facilitate the anastomosis (Fig. 25.12).

Before suturing the anterior wall of the anastomosis, the anesthesiologist advances a Nélaton catheter, which is grabbed laparoscopically and inserted into the distal collapsed segment. Patency of the proximal small bowel loops is cleared by washing saline solution via the Nélaton catheter. Once the anastomosis is completed, the ports are removed and the sites are closed with absorbable sutures.

When a duodenal membrane is suspected, a longitudinal incision is made on the anterior wall of the duodenum, crossing from the distended duodenum to the distal collapsed duodenum (Fig. 25.13). A urinary catheter is inserted through the abdominal wall directly into the distal duodenal segment, the balloon is filled, and the catheter is gradually pulled back. A membrane with an aperture will stretch itself on top of the balloon. The membrane is incised carefully in its lateral aspect and the longitudinal incision is closed.



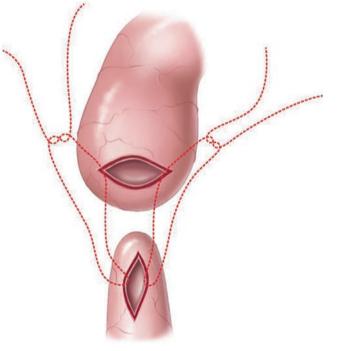


Fig. 25.12 Stay sutures are placed at each corner to facilitate the diamond-shaped anastomosis. The anastomosis is created with either a separate running suture for the posterior and then anterior walls, or single interrupted stitches of Vicryl. Intracorporeal knot tying is used

Fig. 25.11 The colon is mobilized to the left, and the duodenum is then adequately mobilized and freed from its retroperitoneal attachments (Kocher maneuver). A stay suture with a large needle is then inserted through the abdominal wall close to and below the right costal margin. The stay suture is introduced into the bulbus duodeni to move the bulky part of the bulbus out of the way, allowing a view of the distal duodenum and a more convenient approach to the anastomosis. A transverse incision using the hook is made in the distal wall of the dilated duodenum, followed by a longitudinal incision in the distal collapsed duodenum for the diamond-shaped anastomosis of Kimura

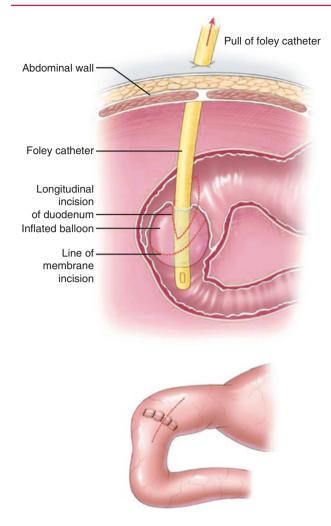


Fig. 25.13 When a duodenal membrane is suspected, a longitudinal incision is made on the anterior wall of the duodenum, crossing from the distended duodenum to the distal collapsed duodenum A urinary catheter is inserted through the abdominal wall directly into the distal duodenal segment without a trocar. The balloon is filled and the catheter is gradually pulled back. If a membrane with an aperture is present, it will stretch itself on top of the balloon. The membrane is incised carefully in its lateral aspect, leaving the medial part of the membrane intact, to prevent injury to the papilla Vater. The longitudinal incision is closed transversely with a running suture or several interrupted sutures

25.4 Outcome and Complications

Long-term outcome after repair of congenital duodenal obstruction is excellent, with contemporary operative survival exceeding 95% and with most patients reported to be asymptomatic, with normal growth.

The experience with laparoscopic duodenoduodenostomy demonstrates that it can be performed safely and successfully in the neonate with excellent short-term outcomes. The last retrospective studies comparing the surgical outcome of laparoscopic repair versus open repair found that laparoscopy is a safe and effective technique. The results, including operative time, length of stay, time to full feeding, and complication rate, were similar in both groups.

The main factors contributing to the mortality in patients with duodenal obstruction are a high incidence of associated anomalies, prematurity, and low birth weight. The associated complex cardiac defects continue to be the leading cause of death, particularly in infants with trisomy 21, but recent reviews document that advances in both paediatric cardiology and cardiac surgery in neonates and infants have reduced this mortality significantly.

Early postoperative complications include anastomotic obstruction, continuing obstruction due to missed anomalies, leakage at the anastomosis, and prolonged adynamic ileus. Upper intestinal radiographic studies are necessary to reveal the source of the problem.

The late gastrointestinal complications include megaduodenum, duodenogastric reflux, gastritis, peptic ulcer, and gastro-oesophageal reflux. Megaduodenum is a particularly troublesome problem, which may result either from anastomotic obstruction or from an inherent motility disorder of the proximal duodenum.

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