CHAPTER 62
Duodenal Atresia and Stenosis

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Introduction

Congenital duodenal obstruction may be due to intrinsic or extrinsic lesions. Intrinsic duodenal obstruction may be caused by duodenal atresia, stenosis, diaphragm with or without perforation, or by a wind-sock web or membrane that balloons distally. Extrinsic duodenal obstruction may be caused by malrotation with Ladd's bands or a preduodenal portal vein or annular pancreas. The annular pancreas itself is not believed to be the cause of obstruction, as there is usually an associated atresia or stenosis in these patients.

Duodenal obstructions usually occur in the second part of the duodenum. They are believed to result from a developmental error during early foetal life within the area of intense embryological activity involved in the creation of the biliary and pancreatic structures. Thus, the obstruction usually occurs at or below the ampulla of Vater.

Duodenal obstruction is associated with prematurity (46%) and maternal polyhydramnios (33%).[1] In addition, there is a high incidence of specific associated anomalies, including Down syndrome (>30%), malrotation (>20%), congenital heart diseases (20%), and other gastrointestinal tract (GIT) and renal anomalies. Along with prematurity and low birth weight, these associated anomalies are known to be significant risk factors contributing to mortality in patients with duodenal atresia. Of note, the presence of Down syndrome itself does not influence the outcome of these babies.

Demographics

Although detailed statistics are not available in much of Africa, the incidence of duodenal obstruction is reported to be 1 in 5,000–10,000 births in most reports.[2] Duodenal obstruction and jejunoileal atresia rank among the two most common causes of intestinal obstruction in large series in the African population.[3]

Aetiology

It has been demonstrated that from gestational weeks 5 to 10, the duodenum is a solid chord. Intrinsic obstructions result from failure of vacuolization and recanalization. An annular pancreas results from fusion of the anterior and posterior anlage, forming a ring of pancreatic tissue that surrounds the second part of the duodenum. Extrinsic obstructions result from a variety of disorders of embryologic development specific to the pathology.

Clinical Presentation

Prenatal

Duodenal obstruction is readily diagnosed by prenatal ultrasound. Antenatal care with prenatal ultrasonography should therefore be offered to pregnant women in all circumstances. Duodenal obstruction presents up to gestational week 20 with a double-bubble phenomenon due to the simultaneous distention of the stomach and the first part of the duodenum. In more than 30% of cases, maternal polyhydramnios is present, and in some cases, serial amniotic aspiration has been reported as necessary. In facilities where ultrasound is not available, a high index of suspicion must be maintained in cases of maternal polyhydramnios. Pregnancy can last near to maturity, and spontaneous delivery is usually the case.

Postnatal Symptoms and Signs

Most women in Africa do not avail themselves of prenatal care, so the majority of duodenal obstructions present only after birth. Furthermore, because up to 60% of births occur outside health institutions, these cases often present very late. The most common presenting features are bilious vomiting and feeding intolerance. Dehydration and electrolyte depletion rapidly ensue if the condition is not recognized and intravenous therapy is not begun. Aspiration and respiratory failure may follow. Repeated nonbilious vomiting is seen in cases of supra-ampullary obstruction (20%). Patients with a web or partial stenosis can survive to present in a much delayed fashion.[2]

Physical signs are nonspecific but can include upper-abdominal distention with scaphoid lower abdomen. Additionally, in the appropriate clinical context, observation of typical Down syndrome features should raise suspicion towards duodenal obstruction as the cause for neonatal intestinal obstruction. Finally, a careful physical exam should concentrate on recognizing signs of significant congenital heart disease (e.g., cyanosis, murmurs), which could complicate perioperative management

Investigation

In tertiary perinatal centres where a prenatal diagnosis has already been established, no further diagnostic work-up is typically necessary.

In doubtful cases or in other settings, a plain abdominal x-ray is the key method for diagnosis. An x-ray showing double-bubble gas shadows is essentially pathognomonic for duodenal obstruction (Figure 62.1). If no double bubble is seen, instillation of 10–15 ml of air immediately prior to a plain abdominal radiograph may help to demonstrate these findings. In cases of stenosis or perforated membranes, air may be seen in the distal GIT. Water-soluble contrast radiography is confirmatory, but it is generally needed only in cases of incomplete obstruction. Radiographic findings of annular pancreas are usually indistinguishable from other forms of duodenal obstruction.

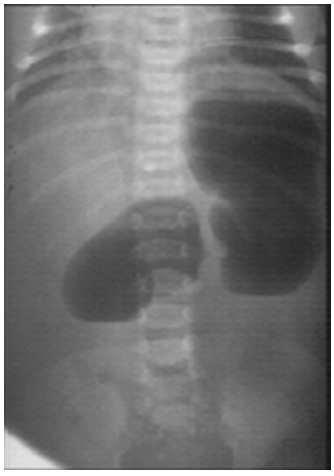

Figure 62.1: Double-bubble sign on plain x-ray. Note the lack of distal gas.

The most important differential diagnosis is duodenal obstruction due to malrotation, resulting in volvulus of the midgut loop or extrinsic compression related to Ladd´s bands across the duodenum. When no prenatal diagnosis is available, contrast radiography may be helpful to differentiate between these entities and can demonstrate the absence of the normal C-shaped curve of the duodenum or a classic "bird's-beak" shape secondary to a volvulus. When the diagnosis still remains in doubt, prompt laparotomy is warranted because undiagnosed volvulus can result in gangrene of the entire midgut within hours.

If available, in cases of incomplete obstruction, oesophagogastroduodenoscopy (EGD) can be done to prove the existence of an intrinsic obstructing membrane. An endoscopic approach to membrane resection can be utilized.

Management

Preoperative Care

The intensity of preoperative care is typically proportionate to the time from birth until hospital presentation. Initial therapy consists of nasogastric decompression and appropriate replacement of fluid and electrolytes. Most of these newborn patients are premature and small for their gestational age, so special care must be taken to preserve body heat and to avoid hypoglycaemia, especially in cases of very low birth weight, congenital heart disease, and respiratory distress syndrome. When incubators are unavailable, the "kangaroo" method of nursing these children offers the best hope for survival.

General Intraoperative Considerations

General anaesthesia with endotracheal intubation is required. The most commonly utilized incision is a muscle-cutting, transverse, right upper quadrant incision. However, some centres are now employing minimal access laparoscopic methods for repair of duodenal obstruction.

A side-to-side duodenoduodenostomy is the standard repair for duodenal stenosis, atresia, or obstruction due to a preduodenal portal vein. In 1977, Kimura and colleagues described a modification of this procedure, known as the diamond-shaped duodenoduodenostomy.[4] In this technique, a horizontal incision is made across the distal aspect of the proximal, dilated bowel, and a lengthwise incision is made along the proximal aspect of the distal, small-calibre bowel. This can achieve a greater diameter of the anastomosis for better emptying of the upper duodenum. In some cases, duodenojejunostomy can be an alternative and may afford an easier repair with minimal dissection. The choice of surgical procedure is largely based on the preference of the surgeon.

When an annular pancreas associated with duodenal obstruction is encountered (Figure 62.2), the treatment of choice is performance of a duodenoduodenostomy between the segments of duodenum above and below the area of the ring of pancreas. One should never consider division of the pancreatic ring because that could result in a pancreatic fistula while the underlying stenosis or atresia of the duodenum would remain unchanged.

In the case of an endoluminal membrane, duodenotomy and resection of the membrane can be done after localisation of the ampulla of Vater. Alternatively, bypass of the membrane can be performed via a duodenoduodenostomy, if desired. As seen in Figure 62.3, fenestrated membranes may be amenable to an endoscopic approach to resection in centres where this facility is available.[5]

Operative Details

Once the abdomen is entered, the hepatic flexure of the colon is mobilised. The duodenum is adequately mobilized by a Kocher manoeuvre. The ligament of Treitz is divided as needed. A transpyloric tube passed via the nose or mouth is helpful at this stage. Air or saline can be passed into the second part of duodenum to assess the nature and level of obstruction.

The dilated proximal duodenum and collapsed distal duodenum are approximated by using stay sutures.

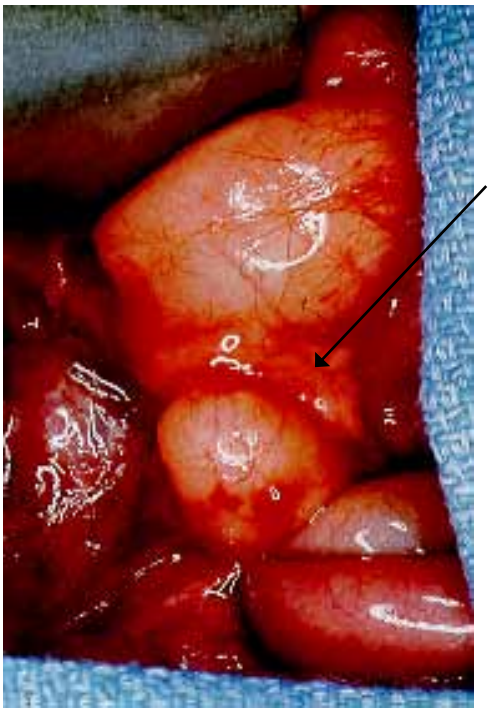

Figure 62.2: Annular pancreas with underlying duodenal stenosis.

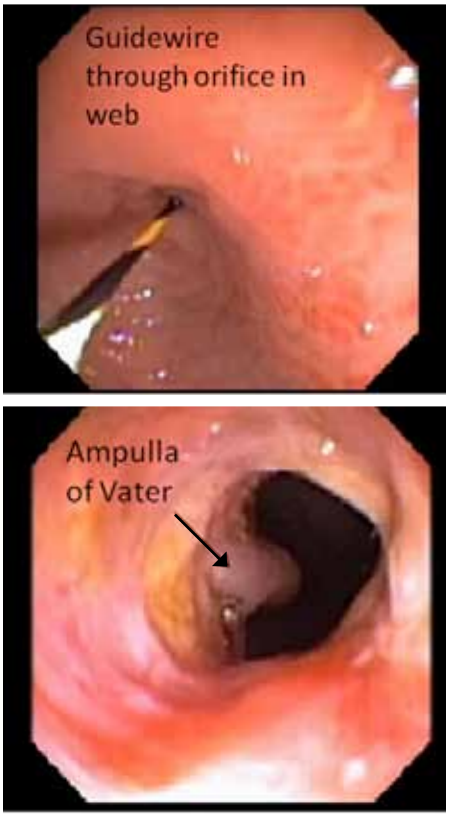

Figure 62.3: Endoscopic view of duodenal web before and after endoscopic dilatation and fenestration of the membrane.

For side-to-side anastomosis, interrupted Lembert sutures (4-0 or 5-0 vicryl or monocryl) start the dorsal part of anastomosis if a two-layer closure is desired. A transverse duodenotomy is made in the proximal segment, 1 cm above the stenosis, to avoid injury of the pancreatico-biliary system. Easy retrograde passage of a tube into the stomach rules out a duodenal web proximal to the duodenotomy. A parallel incision is made in the distal duodenum. The posterior layer of anastomosis is completed by inverting interrupted or continuous sutures of 4-0 or 5-0 vicryl or monocryl. A transanastomotic nasoduodenal silicone tube can be inserted to allow very early enteral feeding beginning at day one or two after surgery. The anterior layer of the anastomosis is completed in the same way. A few Lembert sutures may be used to complete the anastomosis.

For diamond-shaped duodenoduodenostomy, a little more mobilization is needed to bring the redundant proximal duodenal wall down to overlie the proximal portion of the distal segment. Then a transverse incision in the proximal and a longitudinal incision in the distal duodenum are made. The papilla of Vater is located by gentle pressure on the gallbladder. Stay sutures approximate the parts in corresponding points, as shown in Figure 62.4, and the remainder of the anastomosis and placement of a transanastomotic tube is carried out as previously described. Figure 62.5 shows intraoperative photos before and after duodenoduodenostomy.

For a duodenal web, the membrane is usually located in the second part of the duodenum. Localization of the membrane can be assisted by passage of a nasogastric tube (NGT) into the duodenum down to the level of the membrane. Care is to be taken to identify the so-called wind-sock phenomenon, which refers to a proximally attached, lax membrane that bulges into the distal duodenum, making the obstruction point appear more distal than it actually is. This can be identified by looking for a dimpling of the duodenal wall at the attachment point of the membrane more proximal than the distal tip of the NGT.

For membrane resection, a longitudinal incision is made, bridging between the wide and the narrow segments, or at the level of duodenal attachment of the membrane in the case of a wind-sock deformity. It is important to note that the ampulla of Vater may open directly into any membrane or close to it in its posterior-medial part. Therefore, identification of the ampulla is mandatory before excision of the membrane. Excision begins with a radial incision starting in the central ostium and leaving a rim of 1–2 mm of tissue at the duodenal wall. Once again, great care is to be taken to avoid damage to the ampulla of Vater. The resection line is oversewn with continuous suture vicryl 5-0. Before closing the duodenum transversely, patency of the distal duodenum is to be proven with a small silicon catheter and saline.

Postoperative Considerations and Complications

Intravenous infusions are continued for the postoperative period. Using a transanastomotic tube laying deep in the jejunum, feeding can be started as early as 48 hours postoperatively. Where available, parenteral nutrition via a central or peripherally inserted catheter can be very effective for longer-term nutritional support if transanastomotic enteral feeding is inadequate, not feasible, or not tolerated by the patient. All patients have a prolonged period of bile-stained gastric aspirate. This is mainly due to the ineffective peristalsis of the distended upper duodenum. The commencement of oral feeding is dependent upon a decrease in the volume of gastric aspirate and is often delayed for up to several weeks. Patients who have a severely prolonged return of duodenal function and have exceptionally marked dilatation of the proximal duodenum may benefit from reoperation and tapering of the proximal segment, although this is rare.

Anastomotic leak, intraabdominal sepsis, and wound complications also are rare.

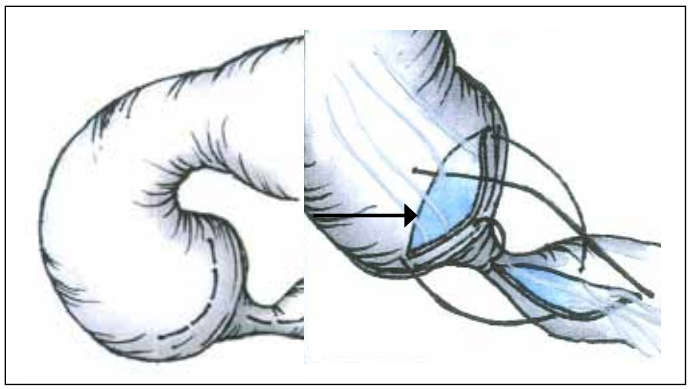

Figure 62.4: Diamond-shaped duodenoduodenostomy.

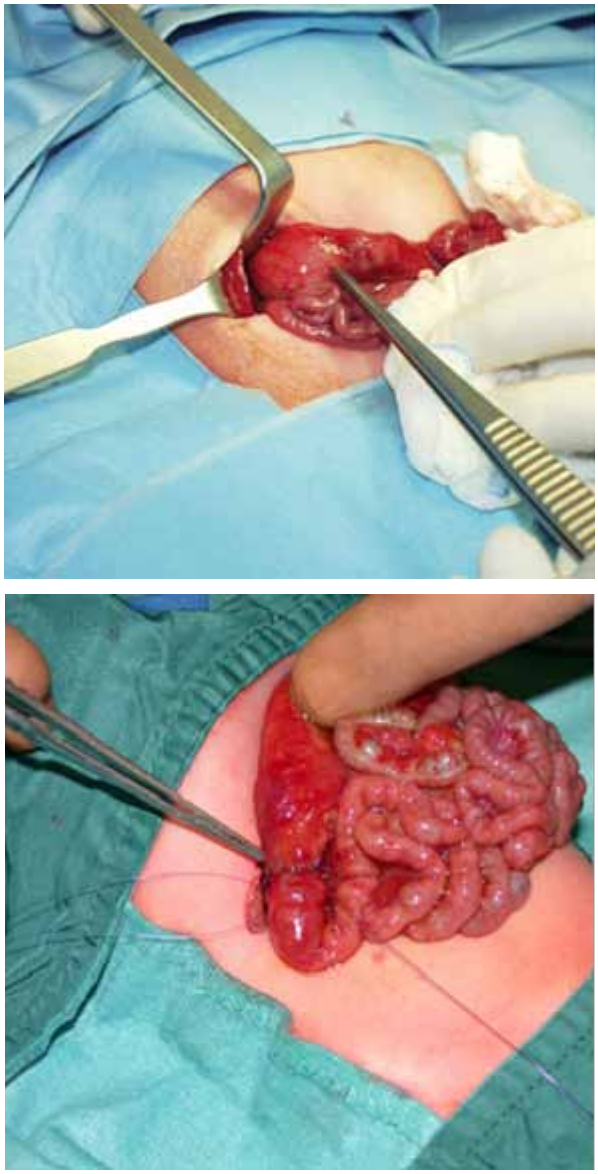

Figure 62.5: Intraoperative photo of duodenal atresia before (top) and after (bottom) duodenoduodenostomy.

Prognosis

Although prognosis of intestinal atresia in general is good, an overall mortality of 7% for duodenal obstruction is shown in large series.[6] Associated congenital anomalies are identified as an independent risk factor for an impaired clinical course. Low birth weight and the problems of prematurity further increase mortality risk.

The morbidity and mortality of neonatal intestinal obstruction is higher in Africa (40%) than in developed countries and is most likely due to late patient presentation and poor neonatal intensive care facilities available in many countries in the continent.[3]

Conclusion

The morbidity and mortality of intestinal obstruction can be improved with earlier referral to specialty centres and with meticulous resuscitation before surgery. Duodenoduodenostomy or duodenotomy with membrane resection in the appropriate circumstance are the typical operations of choice and produce good results with minimal short- or long-term operative-related morbidity. Problems of late presentation and poor neonatal intensive care facilities constitute the basis for the variance in outcomes in Africa when compared to those in developed countries.[7]

Nevertheless, even in Europe and North America, the outcome for children with duodenal obstruction is basically influenced by the degree of prematurity and the presence of associated anomalies.

Evidence-Based Research

Table 62.1 is an observational 15-year retrospective study of the use of diamond-shaped anastomosis for duodenal atresia.

Table 62.1: Evidence-based research.

Title	Diamond-shaped anastomosis for duodenal atresia: an experience with 44 patients over 15 years
Authors	Kimura K, Mukohara N, Nishijima E, Muraji T, Tsugawa C, Matsumoto Y
Institution	Department of Surgery, Kobe Children's Hospital, Kobe, Japan
Reference	J Pediatr Surg 1990; 25(9):977–979
Problem	Role of diamond-shaped anastomosis in the treatment of duodenal atresia and stenosis.
Intervention	Duodenoduodenostomy via diamond-shaped anastomosis.
Comparison/ control (quality of evidence)	In this retrospective observational study, 44 patients over a 15-year period were examined for outcome after diamond shaped anastomosis. All patients underwent this method of repair, so there was no control group.
Outcome/ effect	In all patients, oral feedings were commenced 3.66 ± 1.4 days postoperatively (range, 2 to 6). There was no operative-related mortality. Twenty-one patients had long-term follow-up from 6 months to 15 years. All patients had normal body weight for their age at last record, and current upper GI contrast study (done in 19 of 21 patients) revealed normal calibre of duodenum and anastomosis in all studied cases.
Historical significance/ comments	This study, reported by the originator of the diamond-shaped anastomosis, states the efficacy of this technique in duodenal atresia. Given the relative rarity off the disorder, this report offers a substantial collection of patients with a prolonged follow-up period. Although a comparison group who underwent traditional side-to-side anastomosis was not included here, the results compare favorably to previously published reports of side-to-side anastomotic techniques. Because of its technical ease and its potential to allow early recovery of enteral function without excessive late complications, this technique may be of particular use in undeveloped regions where opportunity for follow-up care is limited.

Key Summary Points

1. Obstructions of duodenum can be intrinsic or extrinsic.
2. There is a high incidence of prematurity and associated anomalies, including cardiac and renal defects as well as Down syndrome.
3. Prenatal ultrasound can be very helpful and may reveal maternal polyhydramnios or a "double-bubble".
4. Physical signs are nonspecific but can include upper-abdominal distention with scaphoid lower abdomen.
5. The most important differential diagnosis to consider is duodenal obstruction due to malrotation, resulting in volvulus of the midgut loop
6. Postnatal plain radiograph revealing a double bubble (distended stomach and proximal duodenum) without evidence of distal gas in the appropriate clinical setting is essentially pathognomic for duodenal atresia.
7. Repair for all forms of duodenal obstruction can be accomplished through side-to-side or diamond-shaped anastomosis proximal and distal to the obstruction. Additionally, duodenal webs can be approached through partial resection of the membrane itself.

References

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