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Minimally Invasive Management of Duodenal and Jejunal Atresia

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28.1 Introduction

Duodenal atresia occurs due to a failure of the lumen to recanalize during the 11th week of gestation. Over half of patients have an associated congenital anomaly with congenital heart disease, trisomy 21, malrotation, annular pancreas, and tracheoesophageal fistula among the most common. The classic presentation is a newborn with early bilious emesis. This finding is dependent on a post-ampullary obstruction, and a small percentage of cases may present with non-bilious emesis due to a pre-ampullary lesion [1]. Prenatal ultrasound often suggests obstruction due to dilated proximal small bowel and polyhydramnios. After birth, the classic imaging finding is the "double bubble" of stomach and duodenal bulb with an absence of distal gas; however, the presence of distal gas does not exclude atresia [2]. Gray and Skandalakis classified the types of duodenal atresia in 1972. Type I defects are the most common and contain a thin membranous separation between the two portions of bowel. In type II, an atretic, fibrous cord connects the two halves, and in type III the segments are entirely separated, and there is an adjacent mesenteric defect [3].

Jejunal atresia is a separate entity and is thought to occur due to a late intrauterine vascular event that compromises the development of one or more sections of the midgut. Prenatal diagnosis is less common, though proximal lesions may present similar to duodenal atresia. Presentation may vary slightly based on location. Bilious emesis is a hallmark. Abdominal distension may not be present in proximal lesions due to the inability to sequester fluid throughout the intestines, but it is common in distal atresias. As such, distal atresias tend to present later as the child may tolerate the first few feedings. Associated congenital anomalies are far less common than in duodenal atresia, though defects related to the midgut are more common and severe. These include mesenteric defects with potential for internal hernias, volvulus, and multiple lesions accounting for significant bowel length. Supine and decubitus radiographs are generally adequate to confirm diagnosis and often display classic obstructive findings. The most commonly used classification system for distal intestinal atresias is the 1979 Grosfeld modification of the system originally described by Louw and Barnard (Fig.28.1) [4, 5]. Type I is a mucosal web or atresia with an intact bowel wall and mesentery. Type II is atretic bowel segments connected by a fibrous cord. Type IIIa is atretic bowel segments with a corresponding mesenteric defect, and IIIb is described as the apple peel atresia or Christmas tree defect. Type IV refers to multiple atretic defects.

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We describe in this chapter the minimally invasive approaches to duodenal and jejunal atresia.

Laparoscopic repairs have been shown to be at least as safe and efficacious as open repair [6, 7]. Some series demonstrate shorter hospital stays, time to initial feeding, and time to goal oral intake with the laparoscopic approach [8].

28.2 Preoperative Preparation

Neither of these conditions alone are surgical emergencies such that preoperative resuscitation and foregut decompression are the first objectives. Associated volvulus or internal hernias with strangulation are the only emergent indications for operation which can occur with more distal atresias.

With suspected duodenal atresia without clear radiographic evidence, we perform a limited upper gastrointestinal study by instilling just enough contrast volume to evaluate for malrotation and volvulus and evacuating any residual contrast. An echocardiogram should be obtained preoperatively in all cases, with additional workup electively and as clinically indicated.

Conversely, these measures may be selectively pursued as clinically indicated in jejunal atresia given the low incidence of associate anomalies. Unlike duodenal atresia, a water-soluble contrast enema is useful first to demonstrate microcolon from a small bowel obstruction, second to evaluate for concurrent distal atresia, and finally to evaluate associated Hirschsprung's disease or meconium ileus/plug. It is important to council families preoperatively that up to 15% of these cases may result in short bowel syndrome [9].

28.3 Positioning

The patient is positioned supine on the operating table with the arms tucked at their sides. Monitors for the surgeon and assistant are placed at the head of the bed. We usually accomplish this by turning the baby perpendicular to the table at the head of the bed and placing monitors opposite the surgeons. This requires passing the cords off the table over the opposite side of the baby from the head of the bed to avoid having the cords over the endotracheal tubing.

28.4 Instrumentation

A 5 mm trocar is used for the camera, and a 30°, 5 mm laparoscope is used for most cases with only 3 mm instruments. These can be place directly through the abdominal wall without ports using #11 blade to make the stab incisions.

28.5 Technique

The operation of choice for duodenal stenosis is a duodenoduodenostomy. Port placement may vary, and two options are shown in Fig. 28.2. In the upper abdomen, either a port or a transabdominal stitch through the falciform ligament can be used for liver retraction and exposure. The duodenum is mobilized sufficiently to identify the obstructing lesion and to create a tension-free anastomosis. A transverse enterotomy is made in the anterior wall of the dilated, proximal duodenum, and a longitudinal enterotomy is made on the antimesenteric border of the duodenum distal to the lesion (Fig. 28.3). Stay sutures placed at the corners can better align the bowel, and the back wall is sutured before the front in a single layer to create a diamond-shaped anastomosis, as seen in Fig. 28.4 [10]. Suture choice and interrupted vs.



Fig. 28.2 Classic "double bubble" seen in duodenal atresia [14]





Fig. 28.4 Options for laparoscopic port placement [14]



Fig. 28.5 Radiographic and intraoperative findings of jejunal atresia [14]

continuous technique do not seem to impact complication rates [7]. A tapering duodenoplasty is useful to accommodate a dramatic size mismatch.

Jejunal atresia is approached similarly, beginning with a broad survey of the abdomen contents. Any associated volvulus or internal hernia is reduced and bowel viability is assessed. The lesion is found by identifying the transition point between dilated proximal and normal or small-caliber distal bowels, and any adhesions are lysed to mobilize this segment (Fig. 28.5). Laparoscopy is often limited by intestinal distension in a small abdomen, and eviscerating via the umbilical incision, with or without an extension of the incision, is a useful adjunct. We frequently approach these by separating the cord vessels individually and eviscerating the bowel for an extracorporeal operation, which can be done without a cosmetic defect and avoiding the laparotomy. The overall goals are to maintain bowel length and establish continuity for enteral feeding. Size mismatch is routinely encountered and can be addressed by techniques such as tapering, elliptical anastomosis, or antimesenteric cutback anastomosis. Elliptical anastomoses are end-toend reconstructions and are created by either cutting the distal bowel at an angle resecting more off the antimesenteric side or by cutting a slit in the antimesenteric border. Tapering is achieved by making an enterotomy in the distal end of the proximal dilated bowel and resecting antimesenteric bowel retrograde until more normal-caliber bowel. A 20-24 F rubber catheter can be placed in the lumen of the bowel to prevent narrowing. Grossly dilated small bowel is at high risk for dysfunctional motility and may need to be resected to prevent pseudo-obstruction and bacterial overgrowth. Type IIIb lesions place the patient at high risk for short bowel syndrome and have unique considerations. In these situations, absorptive surface area must be maximized, and therefore dilated, dysfunctional segments are tolerated. They may then be used in a taper or serial transverse enteroplasty.

28.6 Postoperative Care

In both conditions activity and bathing are not restricted after surgery. Acetaminophen is the primary analgesic with narcotics used for breakthrough pain.

In duodenal atresia, patients are kept NPO and on TPN, which is often initiated preoperatively, for 5 days. An orogastric tube is left in place to suction and may be transitioned to dependent drainage. On day 5, we perform an upper gastrointestinal contrast study. If no leak is identified and contrast empties beyond the anastomosis, the gastric tube is removed, and feeds are initiated. One series has suggested that a transanastomotic feeding tube can expedite time to initiation of feeds and time to goal feeds [11]. Traditional teaching suggested more proximal lesions resulted in longer time to return of bowel function. This was because it took weeks for the nasogastric tube to diminish the amount of bilious output. Early contrast may empty from the stomach, even with frank bilious output from teh gastric tube. This taught us that bilious output continues for so long with proximal lesions because of an incompetent pylorus allowing for suction of the duodenum, not because of inadequate bowel function.

Jejunal atresias follow standard postoperative advancement pathways based on return of bowel function, and these patients will likewise remain on TPN until then. High nasogastric output may require replacement.

28.7 Results

The average length of surgery is approximately 90–120 min. Complications include anastomotic leak, anastomotic stricture, missed obstruction, delayed gastric emptying, short bowel syndrome, dysfunctional bowel motility, and bacterial overgrowth. Operative mortality is low at $\leq 4\%$ [1].

Surgical follow-up is not required in straightforward cases with patients on goal enteral feeds, particularly with duodenal atresia. Complicated cases and short bowel syndrome require specialized, multidisciplinary follow-up with the potential for surgical revision.

28.8 Tips and Tricks

- Be aware of the windsock deformity of a duodenal web or a diaphragm. If unrecognized an anastomosis may be created distal to the obstruction. Passage of a catheter proximally and distally can help exclude luminal obstruction.
- Identify the head of the pancreas and look for pancreatic tissue near the transition point, as a partial annulus can still the source of obstruction.
- In the case of premature infants, it is still worth mobilizing the duodenum completely and performing the anastomosis transumbili-

cally; this is also true when a tapering enteroplasty is used. We have done this for patients as small as 1 kg.

- Awaiting transition of orogastric tube output from bilious to clear and expecting traditionally normal volumes of output will delay progress after duodenoduodenoplasty. Patients will tolerate feeds well before this time.
- If a contrast enema is not obtained prior to operation for a distal small bowel atresia, ensure patency of the distal bowel by instilling saline antegrade through an enterotomy at the site of resection.
- Calcifications may be seen on radiographs and are concerning for in utero perforation, while displacement of the bowel loops by a gasless mass may indicate a meconium pseudocyst.

28.9 Discussion

Duodenal and jejunal atresias are intrinsic congenital intestinal obstructions and must be differentiated from both other intrinsic causes such as web or stenosis and extrinsic causes such as an annular pancreas. They are clinically distinct entities with different etiologies, associated congenital anomalies, and management strategies.

The presentation and management of duodenal atresia are generally more straightforward than jejunal atresias, and it is usually the associated disorders that require more involved evaluation. The operation is readily achieved laparoscopically, though a hybrid approach of laparoscopic mobilization with extracorporeal suture is a viable alternative. A criticism of the laparoscopic technique has been the inability to adequately evaluate for a concurrent distal atresia; however the rate of simultaneous lesions is less than 1% [12]. Thus, full inspection beyond what is capable laparoscopically is not necessary. Outcomes for duodenal atresia have been studied up to 30 years out, with a 9% revision rate. The late mortality rate was 6%, and the vast majority are due to comorbid conditions [13].

The complexity of jejunal atresias is in the operative decisionmaking, and the same principles may be applied to ileal atresia. The multitude of techniques can be used to achieve the primary goals of preserving bowel length and establishing enteral continuity for feeds. If both of these conditions are met, then minimizing the impact of dilated, dysfunctional bowel via tapering or resection can be entertained.

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