Intestinal Atresia

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Intestinal atresia is a common cause of congenital intestinal obstruction and encompasses a spectrum of luminal defects that are categorized into stenosis and four different types of atresia. Stenosis is a narrowing of the bowel lumen without disruption. Type I atresia is occlusion of the lumen with a continuous outer wall, also referred to as a web. Although less common than in duodenal atresia, a type I jejunal atresia can manifest as a windsock deformity where there is a membrane that is stretched and telescopes inside the lumen. Type II atresia has discontinuity of the lumen with a persistent fibrous cord connecting the dilated proximal bowel with the decompressed distal bowel. Type III atresia is the most common type and has complete separation of the proximal and distal ends. In jejunoileal atresias, this represents two distinct subtypes. Type IIIa is characterized by a separation of the bowel and a V-shaped mesenteric deformity. Type IIIb results from loss of the entire superior mesenteric artery except for the middle and right colic arteries; this leaves only retrograde flow from the colonic arcades to supply the distal small bowel resulting in a short length of small bowel wrapped helically around its single arterial supply (Fig. 47.1). Type IIIb atresias are often referred to as a "Christmas tree" deformity, Maypole deformity, or apple-peel atresia. Type IV atresia is a combination of multiple type I–III atresias.

Jejunoileal atresia occurs in roughly 1 in 3000 live births and is most commonly sporadic. There are also familial forms that present with multiple atresias or apple-peel atresia; both of which are believed to be autosomal recessive. Jejunoileal atresia is associated with cystic fibrosis, malrotation, volvulus, and intrauterine intussusception.

Colonic atresia is a rare condition occurring in 1 in 20,000 live births and represents fewer than 10 % of all intestinal atresias. Colonic atresia is also believed to be primarily due to a vascular insult after organogenesis and is usually an isolated anomaly, but it can be associated with a number of other conditions including Hirschsprung's disease (HD), other gastrointestinal abnormalities, gastroschisis, abdominal wall defects, musculoskeletal abnormalities, and genitourinary abnormalities.

Diagnosis

Prenatal diagnosis can be suggested by dilated loops of bowel on ultrasonography or polyhydramnios. Polyhydramnios is less common than with duodenal atresia and becomes less frequent as the atresia becomes more distal due to the increased length of the fetal intestine to absorb fluid.

Clinically, jejunoileal and colonic atresias will present with emesis in the first 48 h of life. Jejunal atresias are classically associated with passage of a gray and mucoid meconium, but passage of normal meconium is possible as well. Patients with colonic atresia often do not pass meconium at all. Patients with more distal atresias are more likely to present later with a distended abdomen. Patients with colonic atresia may develop severe abdominal distension requiring mechanical ventilation. Delay in diagnosis of distal atresias can result in overdistension of the proximal end of the atresia, necrosis, perforation, septic peritonitis, and death. This is especially important in colonic atresia as the ileocecal valve can create high-pressure distension similar to a closed loop obstruction.

The initial diagnostic test for intestinal atresia is the abdominal radiograph. These patients exhibit multiple distended loops of bowel with air-fluid levels indicating intestinal obstruction (Fig. 47.2). The extent and number of distended loops will suggest the level of the atresia. A radiograph with only a few distended loops in the epigastrum is usually indicative of a proximal jejunal atresia, whereas diffuse distended loops are suggestive of a distal ileal or even

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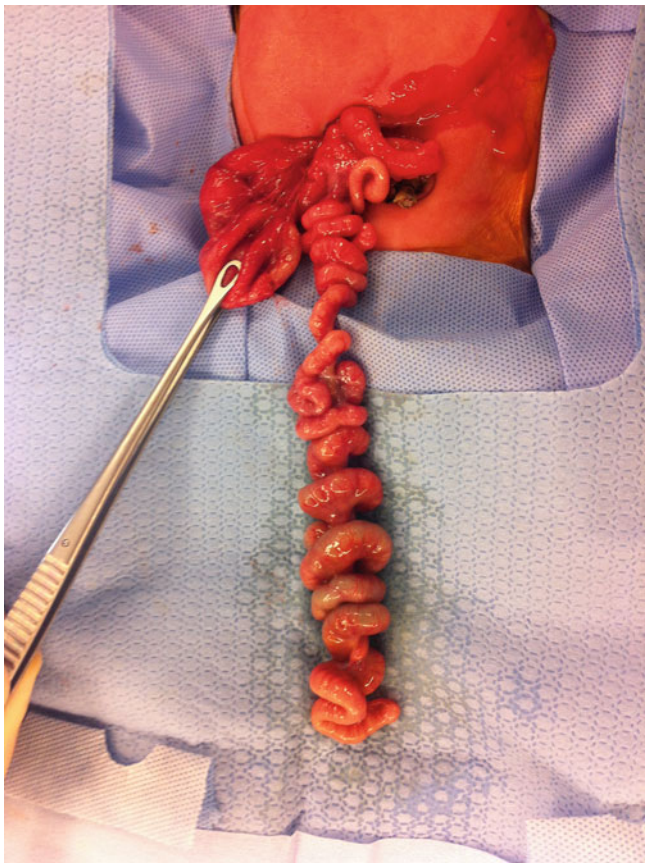

Fig. 47.1 Intraoperative image of newborn with a type IIIB atresia

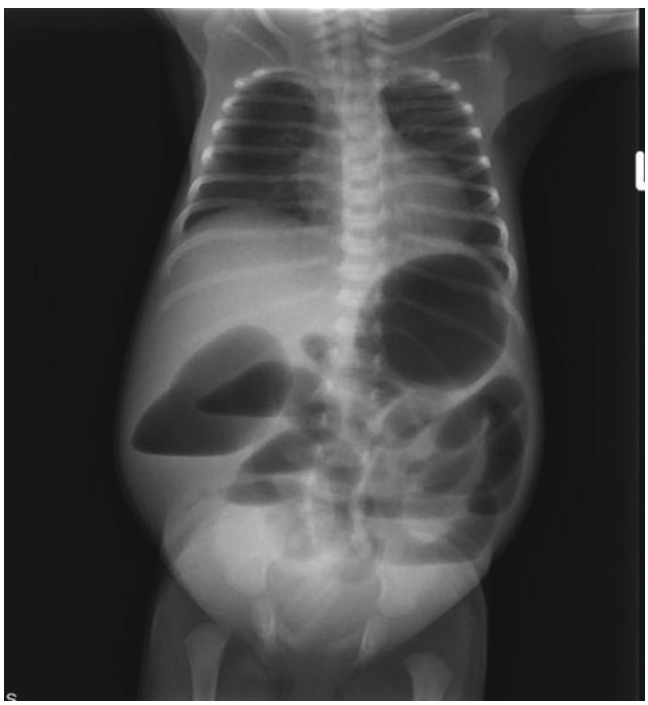

Fig. 47.2 Abdominal radiograph in a patient with ileal atresia (Courtesy of Dr. Paresh K. Desai, Radiopaedia.org)

colonic atresia. In general, colonic atresia will exhibit more severe dilation, which can be misread as pneumoperitoneum. However, it is important to remember that the neonatal colon does not reliably exhibit haustra as it does in older children or adults; therefore, it can be difficult to distinguish a distal ileal atresia and colonic atresia on a plain radiograph.

Contrast radiography can also be useful to define the anatomy of the atresia and associated anomalies. An upper GI contrast study is usually not necessary but might be needed to distinguish a proximal jejunal atresia from malrotation with volvulus in a neonate with bilious emesis. They may also be helpful for distinguishing a stenosis from a complete obstruction when the plain radiographs are equivocal or for diagnosing a windsock deformity. In cases of distal obstruction, contrast enemas are extremely useful because of the difficulty discerning distal small bowel from colon on plain radiograph. The contrast enema will show the passage of retrograde contrast to the level of the atresia. It can also aid in evaluating for total colonic aganglionosis or concomitant HD. Whenever possible, we would recommend obtaining a contrast enema to assess for additional distal atresias prior to entering the operating room; this then allows the intraoperative evaluation for distal atresia to be limited to flushing the distal small bowel into the cecum. In HD, the enema study will show contrast passing retrograde through a narrowed area of distal colon and into the dilated proximal colon usually with a reversal of the rectosigmoid ratio. In colonic atresia, the contrast enema will often show a microcolon of the distal segment from disuse, and contrast will fail to reach proximal dilated bowel.

Preoperative Management

Prior to operative correction of these anomalies, these patients should have an NG or OG tube placed for gastric decompression and IV fluid resuscitation to correct underlying dehydration and electrolyte abnormalities. Any patient with suspicion of intestinal perforation (free air or significant free fluid on radiograph, peritonitis, sepsis) should receive broad-spectrum antibiotics and undergo emergent abdominal exploration. All patients not receiving empiric antibiotics receive appropriate perioperative antibiotics beginning with a preoperative dose and continuing for 24 h.

Jejunoileal Atresia

Operative management of intestinal atresias should be based on the type and location of the lesion, the appearance of the bowel in the OR, and the presence of other anomalies or comorbidities. We prefer a supraumbilical transverse laparotomy. First, we eviscerate and examine the entire

small bowel, measure the small bowel length, assess intestinal rotation status, and examine for multiple atresias. Before performing the anastomosis, we ensure that there are no distal atresias by placing a rubber catheter into the cut open end of the distal bowel and flushing fluid into the cecum. If we did not obtain a contrast enema preoperatively, we will place a rubber catheter with a syringe of fluid attached in the rectum prior to prepping and draping; subsequently, we will flush fluid retrograde and be sure it reaches the cecum during the procedure. Alternatively, the antegrade fluid flushed into the cecum can be followed around the colon until it exits the anus.

The next step is to evaluate the size discrepancy between the ends prior to performing the anastomosis. A dilated proximal segment will have poor motility and can contribute to delayed bowel function or functional obstruction. If the dilated proximal segment is short (up to 10 cm) and the patient has a normal length of small bowel (a healthy full-term infant has an average bowel length of 250 cm, a preterm infant around 150 cm), then the dilated segment can be resected to decrease the size mismatch between the two ends. However, resection of the dilated segment may not be appropriate for patients with a long dilated segment or significantly foreshortened bowel (<100 cm). These patients should receive a tapering or plication enteroplasty. We prefer the tapering enteroplasty, which is performed by decompressing the proximal limb, placing a large rubber catheter in the lumen (20–30 Fr), and then performing a stapled resection on the antimesenteric border. We do not oversew the staple line, but others might do so usually with interrupted sutures.

After resection or enteroplasty, if there is still a size discrepancy, the distal end can be better matched to the proximal end by trimming its antimesenteric border obliquely to provide an elliptical opening. If the circumference of the edge of the distal bowel is still too small, a longitudinal incision can be made on the antimesenteric border to effectively lengthen the edge.

A special management challenge can arise with type IV atresia. While this can represent any combination of the other three types, it commonly presents with short segments of bowel separated by atretic segments. With this defect, the two available options are to resect atretic segments with the intervening bowel or to perform multiple anastomoses. This decision is made by evaluating the total small bowel length. If the patient's total bowel length is significantly less than average, then each island of bowel should be preserved to maximize length, and multiple anastomoses should be performed.

After the two ends are appropriately sized, we then perform the hand-sewn end-to-oblique anastomosis using interrupted 5-0 or 6-0 absorbable sutures in a single layer. We close the mesenteric defect with interrupted absorbable sutures taking care not to damage the vascular supply. The remaining length of small bowel is then measured and documented in the operative report; this is an important step for all atresia patients as they are at risk for short bowel syndrome and intestinal failure.

Some surgeons utilize less invasive techniques such as a periumbilical or laparoscopic approach to improve cosmesis and potentially speed postoperative recovery. With the circumumbilical approach, an incision is made either three-quarters of the way around the umbilicus or vertically through the umbilicus, and the abdomen is entered through the midline fascia. With the laparoscopic approach, an initial trocar is placed at the umbilicus and the abdomen is insufflated to 8–10 mmHg. Two additional trocars are then inserted, usually to the left and right of the umbilicus. Graspers are used to run the bowel, identify the proximal and distal ends, and evaluate for malrotation and additional atresias. Once the anatomy has been defined, the proximal and distal ends are exteriorized through the umbilicus, the proximal end is trimmed, and a standard anastomosis is performed.

These less invasive approaches may not be appropriate for certain patients. Laparoscopic visualization can be limited in patients with a large amount of bowel distension. In addition, we prefer to manage type IIIb atresias with an open procedure as the distal bowel is coiled around its sole blood supply and must be carefully uncoiled to maintain correct orientation of the vasculature.

A primary anastomosis is not always the best option. If the bowel ends show signs of vascular compromise or if the abdomen is grossly contaminated from a perforation, then an enterostomy and mucous fistula should be performed. We usually place these at opposite ends of the incision, but they can also be located next to each other in double-barrel fashion in one corner of the incision or performed at a different location from the incision.

Postoperatively, these patients should be managed with the expectation of delayed bowel function. The NG or OG tube should be confirmed intraoperatively and maintained on suction. When the tube output is decreased and no longer bilious and the patient is passing flatus or stool, the tube can be removed and the child slowly advanced on feedings. Parenteral nutrition should be started early and maintained until the patient is tolerating full feeds.

The most common postoperative complications are anastomotic leak, obstruction, and stricture. Long-term outcomes for these patients are good with survival >90 %. Some patients suffer from chronic gastrointestinal dysmotility. Others are at risk for developing short bowel syndrome including patients who underwent a large resection or who were born with foreshortened bowel or a type IIIb atresia.

Colonic Atresia

The two surgical options for colonic atresia are primary anastomosis or colostomy with delayed anastomosis. The chosen approach should take into consideration the patient's condition, the presence of perforation or peritonitis, the condition of remaining colon, and the presence of associated anomalies such as gastroschisis or HD.

In patients without comorbid conditions, a primary anastomosis is the preferred option if a distal atresia has been excluded. Because HD is not uncommon in patients with colonic atresia, an intraoperative rectal biopsy should be sent for frozen section prior to performing the anastomosis. Failure to adequately rule out HD could result in anastomotic leak or persistent bowel obstruction. The operation is performed through a supraumbilical transverse laparotomy. When planning the incision, always think ahead for potential enterostomy sites in the event that the anastomosis cannot be completed. Once the abdomen is open, the dilated proximal colon is exteriorized and then the distal colon. There can be a large size discrepancy between the two ends. The bulbous portion of the proximal end should be resected back to an area of more normal caliber to better approximate the diameter of the distal colon. Once the distal end is trimmed obliquely to more closely match the circumference of the resected proximal end, the anastomosis is completed in an end-to-oblique single-layer fashion using absorbable suture.

If the caliber of the proximal colon cannot approximate the distal segment without a long segment resection, then a primary anastomosis should not be attempted. In this instance, we will use a two-stage approach. During the initial stage, the proximal end is brought out as an end colostomy and the distal end as a mucous fistula. The resected ends of the bowel and a rectal biopsy should be sent to pathology to evaluate for ganglion cells. Subsequently, the child is brought back electively for anastomosis in 2–3 months. Some institutions have adopted the practice of feeding the colostomy output through the mucous fistula to stimulate dilation of the distal colon for better size approximation.

Postoperatively, colonic atresia patients should retain their gastric tube, and parenteral nutrition should be provided until return of bowel function. Once the NG output is both clearing and decreasing and the patient is passing flatus or stool, the tube can be removed and the diet can be slowly advanced. Survival for colonic atresia is >90 %; however, surgical repair is not without complication. The most common complications are anastomotic leak, stricture, postoperative bowel obstruction, and, rarely, short bowel syndrome.

Summary

Intestinal atresia commonly presents as a stable neonate with emesis and gray mucoid meconium. Type I atresia is occlusion of the lumen with a continuous outer wall, type II atresia has discontinuity of the lumen with a persistent fibrous cord, type III atresia has complete separation of the proximal and distal ends with a mesenteric defect, and type IV atresia is a combination of multiple type I–III atresias. The abdominal radiograph will show multiple distended loops of bowel with air-fluid levels indicating intestinal obstruction. Primary anastomosis should only be considered when there is no abdominal contamination and distal obstruction has been ruled out. Evaluation for malrotation must be performed intraoperatively.

The dilated proximal segment should be resected in most cases, and tapering enteroplasty should be used for long dilated jejunal segments to improve the size mismatch of the two ends.

Feedings should be started once bowel function has returned, and TPN should be used to support patients until bowel function returns. Long-term outcomes for these patients are good with survival >90 %. The most common postoperative complications are anastomotic leak, obstruction, and stricture.

Editor's Comment

In general, the stable newborn with bilious emesis, abdominal distension, and dilated loops of bowel on plain radiograph has one of the four diagnoses – meconium plug syndrome, meconium ileus, Hirschsprung's disease, or intestinal atresia – and should therefore have a contrast enema rather than an UGI. This is one of the few exceptions to the "every child with bilious emesis needs either an urgent upper GI or a laparotomy" rule. Proximal obstruction due to duodenal or pyloric atresia does not present with abdominal distension or distal dilated loops, and although malrotation with volvulus can present with a comparable clinical picture, it quite rarely does so, and an experienced examiner can recognize the differences. A contrast enema is therapeutic for meconium plug syndrome and some cases of meconium ileus, allows identification of a transition zone in patients with HD, and confirms the diagnosis of intestinal atresia by demonstrating an unused "microcolon." With the confidence that comes with experience, one might eventually be able to recommend surgery for intestinal atresia based simply on the plain radiographic findings that clearly shows a single absurdly dilated loop of bowel, but there is no shame in requesting a contrast enema to be more certain.

Except with colonic atresia, one should consider a laparoscopic approach in most patients with intestinal atresia. The setup is the same as for a laparoscopic pyloromyotomy with the only port being through the umbilicus; 3-mm stab incisions can be used for the instruments. The ends of the atresia can usually be delivered through the widened umbilical incision and an extracorporeal anastomosis performed. For most type II and IIIa atresias, this approach works nicely. One of the challenges with intestinal atresias is the size mismatch between the two ends and then performing a hand-sewn anastomosis on the diminutive bowel. This is a particular problem with colonic atresias. Until recently a stapled anastomosis was not an option as the diameter of the distal limb was too small to admit the anvil of a 10-mm stapler. With the introduction of the 5-mm linear stapler, many of these atresias can now be repaired with a side-to-side stapled anastomosis. This could reduce the incidence of anastomotic stricture and shorten OR times.

Whether to resect or taper the bowel is a decision best made after assessing the length of viable intestine and how much bowel would be lost if the dilated portion were resected. In most cases, it is best to resect the most severely dilated bowel (often dysmotile anyway) and taper of the bowel just proximal to this segment. Imbrication is more difficult and tends to undo itself over time. Even after tapering, some infants will develop pseudo-obstruction and require resection of a dysmotile segment at a later date.

Management of the apple-peel lesion (type IIIb) can be tricky. One finds a proximal atresia, a large mesenteric defect with loss of the primary trunk of the SMA, and the entire ileum and distal jejunum remaining precariously viable on the basis of a tiny remnant of the distal SMA and the marginal artery. Some of these patients do quite well after simple primary repair, but many have long-term motility or absorption problems and need parenteral nutrition for a long time. It is important to untwist the bowel carefully, close the mesenteric defect without compromising the remaining blood supply, and inject saline through the distal bowel to rule out another stricture.

Suggested Reading

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