A Comprehensive Review of Intestinal Atresias

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Introduction: Intestinal atresia is an uncommon but challenging disease that requires a high level of suspicion for timely diagnosis and expeditious treatment. Its incidence ranges anywhere from 3.4 per 10,000 to 1 per 66,000 live births. Herein we present a comprehensive review of the various presentations of Intestinal Atresia and provide an algorithm for its evaluation.

Methods: A review of modern English Language in Index databases (PubMed, SCOPUS, EMBASE, MEDLINE, etc) in the English language was preformed. The etiology, pathophysiology, and management of the intestinal atresia at different levels of the intestinal tract were studied.

Results: Atresia is often diagnosed soon after birth and requires a multidisciplinary approach for optimal outcomes. Duodenal atresia is associated with many congenital anomalies and requires a multisystem workup once diagnosed. Jejuno-ileal atresia is the most common intestinal atresia and has fewer associated congenital defects than duodenal atresia. Colonic atresia is the rarest of the intestinal atresias but often has the best outcome as long as it is promptly diagnosed and treatment is not delayed.

Prognosis of neonates with intestinal atresia is overall dependent on co-morbidities, timing of intervention, and length of bowel remaining status-post surgical repair.

Intestinal Atresia
Duodenal Atresia
Pathophysiology

Duodenal atresia is one of the many causes of neonatal intestinal obstructions with an incidence of 1 in 5000-10000
The most widely accepted etiology of duodenal atresia was proposed by Tandler in 1900 and is a lack of recanalization, or excessive endodermal proliferation, of the developing intestine around 8-10 weeks gestation, which differs from the vascular infarct etiology of other intestinal atresias(2). The duodenum has a dual vascular supply via the Celiac artery and the Superior Mesenteric artery which makes it less susceptible to ischemia. Duodenal atresia can range from complete separation of the intestines (Type III), to lumen separation while connected via a fibrous cord (Type II), or simple obstruction due to a mucosal web (Type I) such as the infamous “windsock deformity.”

Clinical Presentation & Diagnosis

Diagnosis can often be obtained prenatally via ultrasound which will demonstrate polyhydramnios&/or a dilated stomach. Post-natal clinical presentation is typically failure to tolerate feeds within the first 24-48 hours of life. The infant may have non-bilious or bilious emesis depending on whether the atresia is prior to the ampulla of vater or distal (80%) to it. If the atresia is present in the form of a mucosal web or stenosis then post prandial emesis may present later in life. Abdominal film will demonstrate the “double bubble” sign with gastric and duodenal gas separated by the pylorus. If there is gas distally a STAT upper GI study should be performed to rule out midgut volvulus which requires emergent repair. A rare cause of distal gas on plain film could be an annular pancreas, which has been found in 23-33% of duodenal atresias(3,4). In the case of an annular pancreas if there is complete duodenal atresia but a patent duct of Santorini above and patent duct of Wirsung below then gas may pass distal to the atretic segment. However, the more common and more emergent diagnosis to exclude is that of malrotation with volvulus.

Approximately 30% patients with duodenal atresia will have trisomy 21, but only 3% Trisomy 21 patients will have duodenal atresia(5). Other commonly associated anomalies include cardiac, malrotation, and annular pancreas. It is recommended that patients with duodenal atresia routinely have a cardiac and renal US, and if associated with Down’s Syndrome a rectal suction biopsy to rule out Hirschsprung’s(5).

Treatment

Management of duodenal atresia is via surgical bypass or resection. Prior to surgery the patient should be medically optimized as best as possible which includes correction of electrolyte abnormalities, return to euovolemia, and proximal decompression via nasogastric tube. Given the association with trisomy 21 and cardiac anomalies it is recommended to have a pre-op echo and consultation with cardiology if timing permits. Perioperative antibiotics should consist of a second generation cephalosporin. If the neonate has had time to colonize the gut, >24 hours of life, anaerobic coverage should be added. Surgical repair consists of a duodenoduodenostomy via either diamond-shaped anastomosis (transverse proximal incision with a longitudinal distal incision), side-to-side anastomosis, or even a duodenonojenostomy(6,7). If there is a significant size discrepancy a tapering enteroplasty may be necessary. Traditionally this repair has been performed via laparotomy but with modern laparoscopic techniques minimally invasive surgery is an option for appropriate candidates at some centers (8,9). However, at the time of the operation the length of the bowel should be interrogated to rule out additional anomalies which is difficult to undertake laparoscopically. Dissection and anastomosis can be challenging due to the retroperitoneal position of the distal limb, therefore some surgeons will use surgical clips instead of sutures. During correction of the duodenal atresia it is important to exclude distal atresia of the small or large bowel, or correct malrotation which has been found 19-28% of patients with duodenal atresia(3,4).

Management of a duodenal web has classically been resection via duodenotomy. Some authors propose also performing a duodenostomy for a mucosal web in order to decrease the risk of a stricture in the future. It was previously recommended to leave trans-anastomotic feeding tubes; modern data has shown this will prolong the time to PO intake and lengthen hospital stay. As such, trans-anastomotic feeding has been abandoned(10).

Prognosis

The overall prognosis of duodenal atresia depends on the concominate congenital anomalies. Isolated duodenal atresia has a good prognosis with the most likely complications being from the surgery itself, such as delayed gastric emptying or stricture. Most infants will resume enteral feeding within a few days post-op. If there is prolonged delayed gastric emptying (> 2 weeks) an upper GI with small bowel follow through should be obtained to rule out a distal stricture.

Jejunal-Ileal Atresia

Pathophysiology

Jejuno-ileal atresia is the most common type of atresia and ranges from 1-5000 to 1-15000 births. There is an association with nearly 30% of infants having a low birth weight. While jejuno-ileal atresia is not generally associated with chromosomal anomalies, there is a suspected autosomal recessive pattern of inheritance to some type IV atresias that can present anywhere from the stomach to rectum (11). This is contrasted to duodenal atresia which has documented association with trisomy 21. Jejuno-ileal atresia is believed to be the result of a mesenteric arterial infarct during the 2nd or 3rd trimester that results in a lack of small bowel development. This theory has been supported since initial experiments were carried out by Louw& Barnard in Cape Town in the 1950s (12). Additional canine studies also supported this theory when ligation of canine mesenteric arteries in-utero led to the development of atresia(13).

Although there are not associated chromosomal anomalies there are associated congenital anomalies. Approximately 10-
16% of infants with gastroschisis will have some sort of jejuno-ileal atresia(14,15). It is also associated with volvulus, omphalocele, intussusception, or internal hernia (16-19). All of these can impede blood supply to the developing bowel and cause strangulation, thereby causing the vascular infarct necessary for the atresia to form.

The severity and timing of the infarct during development determine the extent of atresia, which is categorized according to the Louw Classification that was modified by Grosfeld (see table 1). The frequency and severity of each type is inversely related, and the morbidity/mortality expectedly increases with severity (15,20) Type I is the least severe and intestinal transit is simply blocked by an intraluminal membrane, such as a mucosal web. The serosa remains in continuity and there is no mesenteric defect. Type II atresias also lack a mesenteric defect but the bowel is connected via a fibrous cord. Type III atresias are further subdivided by Grosfeld. Type IIIa consists of a mesenteric gap and no connection between bowel segments. Type IIIb typically occurs just distal to the duodenojejunal flexure with the remaining small bowel coiled around the ileocolic artery giving it the classically described “apple peel” appearance. Type IV atresia also has a mesenteric defect but has multiple atresias and segments of bowel in discontinuity and has often been described as a “string of sausages”.

**Clinical Presentation & Diagnosis**

Jejuno-ileal atresia is associated with fewer congenital anomalies than duodenal atresia due to its occurrence later in development. It will present as abdominal distention and bilious emesis of the neonate and should be treated as an emergency given the similar presentation of malrotation or a volvulus. Occasionally it can present as failure to pass meconium. Initial imaging should be an abdominal film which will demonstrate multiple loops of proximal bowel with air fluid levels and lack of gas in the colon and rectum. A contrast enema is often next to help differentiate a distal atresia from hirschsprung’s disease, small left colon syndrome, or meconiumileus. It also evaluates the colon for atresia which is useful as the large bowel is difficult to evaluate intraoperatively. Colonic atresia can be definitively excluded if contrast is seen refluxing from colon into dilated loops of bowel. In the neonate colonic haustra are not prominent and may not be seen on plain film. Colon is best identified on place film by tracking the predicted location. Additional testing should be performed for cystic fibrosis given its high correlation (10%) in patients with jejuno-ileal atresia(14,21).

**Treatment**

Once the diagnosis of atresia has been established the patient should have a nasogastric tube for proximal bowel decompression and be adequately resuscitated before proceeding to the OR. Intra-op findings will dictate the final procedure to be performed but generally consists of resection of the atretic segments with primary anastomosis. While maintaining bowel length is critical, it is also imperative to minimize the amount of atretic or massively dilated bowel lest it cause a functional obstruction. As long as bowel length will not be excessively shortened it is recommended to resect the proximally atretic bowel (18,22,23). However, in patients where short gut is of concern a tapering enteroplasty may be necessary due to size mismatch and is preferred to formal resection of the dilated bowel(20). In the setting of Type IV atresia multiple anastomoses are preformed and often protected best as possible with a proximal stoma. In patients with gastroschisis and midgut atresia there remains controversy regarding optimum management. Current options include initial exteriorization of the bowel, primary anastomosis as previously discussed, or a multi-stage approach by placing the atretic segments into the abdominal cavity and closing the abdominal wall with a delayed definitive repair several weeks later (24-26).

Patients with short gut will often benefit from a lengthening procedure. One of the first such procedures was described by Bianchi in the 1980s and consists of serial longitudinal division of the dilated bowel. This creates hemiloops of bowel that are then anastomosed to each other in an isoperistaltic fashion(27). In 2003 a new procedure was described by Kim et. al called serial transverse enteroplasty, or STEP (28). This procedure is technically less challenging than the Bianchi and consists of partial and equidistant bowel transections with a stapler to leave a 1-2cm lumen. Some of the advantages of STEP over the Bianchi include the ability to perform it when there is a foreshortened mesentry, and also to perform serial lengthening procedures once the bowel ages and increase in girth(29,30).

When bowel lengthening procedures fail and there is persistent short bowel syndrome the patient must be on lifelong TPN unless a bowel transplant can be performed.

**Prognosis**

The overall prognosis of jejuno-ileal atresia is better than that of duodenal because there are fewer associated genetic anomalies but still depends on the type of atresia and associated congenital anomalies. Short bowel syndrome is most likely to occur in patient with Type IIIa, IIIb, and IV atresias due to the initially short overall length of bowel that is further decreased with the required anastomoses(4). The incidence of short gut syndrome is varied and can range from as low as 3% to high as 43% in some series(20,31).

**Colonic Atresia**

**Pathophysiology**

Colonic atresia is the least common type atresia out of all of the intestinal atresias. The incidence of large bowel atresias estimated to be from approximately 4.2 per 10,000 live births when combined with rectal atresia/stenosis to 1 per 66,000 live births for only colonic atresia (32,33). The pathophysiology of colonic atresia is thought to be similar to jejunal-ileal atresia in that it is the result of anintraperitoneal mesenteric infarct. This
theory has been reproduced in canine studies (12). Theories have also been proposed as to the mechanism of infarct, the vast majority of which point to an ischemic etiology. One case report cited the presence of an internal hernia with a defect through the falciform ligament which subsequently leads to extrinsic mesenteric vascular obstruction (34). Strangulation secondary to gastrochisis has also been proposed as a mechanism to bowel injury resulting in colonic atresia (35–37). It has also been recently proposed that type II and IIIa right colonic atresias may form from constriction as the umbilical ring closes (38). Classification of colonic atresia parallels that of jejuno-ileal atresia.

**Clinical Presentation & Diagnosis**

Colonic atresia, like jejunal-ileal atresia, is not as commonly associated with congenital anomalies. However, there are a few anomalies that have been seen in neonates with colonic atresia including gastrochisis, Hirschsprung’s disease, and other intestinal atresias. It is thought that colonic atresia may be slightly more common in males than females (4:3) (21). The right colon is more commonly affected than the left colon. Colonic atresia on prenatal ultrasound has been described in a case report as dilation of the colon with prominent haustrae, but findings are usually more vague such as polyhydramnios (39). Again, this finding is nonspecific and postnatal diagnosis is the most reliable.

Neonates with colonic atresia will present with signs of obstruction such as bilious emesis and abdominal distention. They may also have failure to pass meconium (40). Often times in colonic atresia, diagnosis is delayed as abdominal distention and emesis typically occurs after several hours after onset of feeds (41). Work up typically begins with a plain radiograph of the abdomen in which one may see dilated loops of bowel, air fluid levels, and distal bowel gas. In order to further differentiate between colonic and ileal atresia, a contrast enema is the diagnostic test of choice. In colonic atresia, a contrast enema will reveal microcolon and absence of contrast in the proximal bowel. A suction biopsy may also be done to rule out Hirschsprung’s disease.

**Treatment**

Prior to surgical intervention, a neonate with a diagnosis of colonic atresia should have a nasogastric tube placed for gastrointestinal decompression. It may be necessary to replace nasogastric tube output with IV fluids. Dehydration can be a common complication in these neonates secondary to emesis and high NG output as the small bowel has limited resorbative capacity. Furthermore, colonic atresia can be overlooked until the infant suddenly stops tolerating feeds, develops abdominal distension & emesis, or has not passed meconium. The delay in diagnosis can result in worsened sepsis, electrolyte imbalance, and increased mortality (42–44). Therefore, care should be taken to maintain meticulous monitoring of electrolytes preoperatively. Surgical repair is usually achieved with resection of the proximal colon and creation of an ileo-colic anastomosis (40). An alternative approach is to create and initial stoma with delayed repair (21). Stoma creation is favored in left colon atresia whereas resection and primary anastomosis is more common with right colon atresia (21). Although colonic atresia may sound similar to simple distal bowel obstruction, urgent repair of colonic atresia is necessary as a literature review of over 200 cases found that delay of surgical repair after 72 hours of life is associated with higher mortality rates (41). Additionally, if the patient has a competent ileocecal valve colonic atresia can act as a closed loop obstruction.

**Prognosis**

Complications from colonic atresia tend to be rare as these patients are less likely to have other anomalies. However, the presence of other anomalies expectedly results in a worse prognosis. Some retrospective epidemiologic studies have quoted the mortality rate at >20% but unfortunately these data include colonic along with rectal atresia and have multiple concomitant conditions (32). Most studies, including the experience of the authors of this chapter, demonstrate an excellent prognosis when colonic atresia is appropriately treated and overall mortality is exceedingly low (4, 45). Typically, patients require TPN for just a few days post-procedure while enteral feeds are slowly resumed in the neonatal ICU. As with other repaired atresias, the length of bowel remaining will also affect overall prognosis.

**Conclusion**

Intestinal atresia can be a devastating disease but it is largely curable if promptly diagnosed and treated. Etiologies among the atresias differ with duodenal atresia being secondary to lack of recanalization or excessive proliferation in early development whereas jejunal/ileal/colonic atresias result from a vascular accident later in development. Bilious emesis and abdominal distention are the most common presentation. The most common diagnostic finding on plain film is an abundance of gas proximally with a scarcity of gas distally. Due to its association and similar presentation to intestinal atresia the critical possibility of malrotation with volvulus must be religiously excluded given the emergent nature of that disease. When diagnosis is unclear or malrotation is low on the differential, a contrast enema can help differentiate between distal atresia and other causes of obstruction (such as meconiumileus, hirschsprung disease, etc). Intestinal atresia can occur anywhere in the gastrointestinal tract but the location and severity determine the treatment. The majority of complications from the intestinal atresias are from comorbid diseases – such as annular pancreas, gastrochisis, malrotation, Trisomy 21, cystic fibrosis – rather than from the atresia itself. Although most patients do well after treatment some will require life-long parenteral nutrition or repeated lengthening procedures depending on the amount of bowel remaining. Overall intestinal atresias are rare but they should always be on the differential for bilious emesis of the neonate.
Table 1. Louw classification with Glosfeld modification of jejuno-ileal atresias.

<table>
<thead>
<tr>
<th>Atresia</th>
<th>Description</th>
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<tbody>
<tr>
<td>Type I</td>
<td>Mesenteric continuity, Serosal continuity, mucosal web</td>
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<tr>
<td>Type II</td>
<td>Mesenteric continuity, serosal discontinuity, bowel connected by fibrous cord</td>
</tr>
<tr>
<td>Type IIIa</td>
<td>Mesenteric defect, complete serosal discontinuity</td>
</tr>
<tr>
<td>Type IIIb</td>
<td>Mesenteric defect, complete serosal discontinuity with “apple peel”</td>
</tr>
<tr>
<td>Type IV</td>
<td>Mesenteric defect, multiple atresias, “string of sausages”</td>
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References: