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Ileal Atresia

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Introduction

Ileal and jejunal atresias are usually described together as jejunoileal atresia (JIA). JIA is a common cause of intestinal obstruction in neonates.¹ It is seen in 1 in 5000 to 1 in 14000 live births.² Intestinal atresia can occur in any location on the small bowel as a solitary or even multiple lesions. Distally located atresia usually presents with delayed symptoms compared to proximal ones. Occasionally, JIA is associated with other malformations such as cardiac anomalies, gastroschisis, and cystic fibrosis.³⁴⁵

Evaluation can be initiated before birth with prenatal diagnosis using ultrasound findings of evidence of intestinal obstruction reported in 29% to 50% of cases.²⁶ Postnatally, the patient presents with signs and symptoms of intestinal obstruction, and diagnosis is made with a plain abdominal radiograph, which shows proximal distended bowel with no distal bowel gas.

Advances in pediatric anesthesia, pediatric surgery, intensive care unit (ICU) care, and nutritional supports over the years have led to better overall survival in patients with intestinal atresia.⁶ Treatment is individualized in most cases and involves resuscitation with intravenous fluid, correction of electrolytes and acidosis, gastric decompression with a nasogastric tube, and operative intervention. The traditional surgical approach is transverse supraumbilical laparotomy, but in recent years minimally invasive approaches such as circumumbilical incisions and laparoscopic-assisted techniques are being widely adopted. The purpose of this article is to review the causes, course, clinical features, evaluation, and recent trends in the management of JIA.

Etiology

The cause of jejunoileal atresia (JIA) has been attributed to an intrauterine vascular accident involving branches of mesenteric vessels in the midgut.⁸⁹ The resultant ischemic necrosis of the fetal bowel is resorbed in utero, leaving behind a blind proximal and distal end of the bowel with a mesentry defect between the ends. Compared to distal vascular disruption, the more proximal vascular disruption results in more extensive bowel defects. There is experimental and clinical evidence in support of the in-utero vascular disruption hypothesis of JIA. Similar atresia was reproduced with induced vascular compromise in experimental animals.¹⁰¹¹¹² Bile, lanugo hairs, and epithelial cells that are seen distal to the atretic segment may suggest that swallowing of the amniotic fluid might have happened before some events. Besides, atresia has been reported in other causes of in-utero vascular disruptions such as intussusception, internal hernia, midgut volvulus, omphalocele, and gastroschisis.¹³¹⁴¹⁸¹⁵¹⁶¹⁷ Thromboembolic occlusion of mesenteric vessels may have contributed in some situations, as showed by the increased risk of small intestinal atresia in mothers exposed to vasoconstrictive drugs and cigarette smoking in the first trimester of pregnancy.¹⁸⁸

Four types of intestinal atresia have been described by Grosfeld classification systems.¹⁹ Type I atresia is described as an internal membrane with serosa continuity and no mesenteric defect; type II involves a proximal and distal blind pouch connected by a fibrous cord with serosal discontinuity; type IIIa has serosa discontinuity with a V-shaped...
mesenteric defect only and type IIIb is the apple peel deformity which described proximal jejunal atresia and a short ileal segment coiled around the ileocolic artery while type IV is characterized by multiple atresias.[19][6] Treatments and prognosis are hugely impacted by the types of atresia. Bowel loss is more common in type IIIb and types IV atresia. Type IIIb (Apple peel) atresia is the least common atresia, with prevalence ranging from 5% to 10% in the literature.[20][2][6] Type IIIb atresias are more likely associated with volvulus with increased risk of distal bowel vascular compromised and have been seen in families demonstrating an autosomal recessive type of inheritance.[2][10] Ileal atresia are rare compared to jejunal atresia, and as the atresia becomes more distal, the less the frequency of occurrence.[21]

**Epidemiology**

The estimated prevalence of jeunoileal atresia (JIA) ranges from 1 in 5000 to 1 in 14,000 live births. About 33% of the affected children are born prematurely, and JIA occurs equally in both sex.[2][7][3][22] Familial cases of JIA have been reported, but the majority of JIA occurs sporadically.[23] Less than 10% of cases of JIA are seen with extra-abdominal organ abnormalities, and this has been attributed to the late occurrence of localized vascular compromise in-utero.[24] There are more associated anomalies such as cystic fibrosis, malrotation, congenital heart disease, Down syndrome (trisomy 21), anorectal, and vertebral reported for jejunal atresia compared with ileal atresia where additional anomalies are rare.[24][3][4][5] No relationship between JIA and paternal or maternal diseases have been reported, and chromosomal abnormalities are seen in less than 1% of patients with JIA.[18][25]

**Pathophysiology**

Structural and functional abnormalities have been reported as part of the sequelae of the ischemic changes from the vascular accident seen in patients with jeunoileal atresia (JIA) in addition to the gross changes in the anatomy of the intestine.[8][26][27] The proximal distended blind loop of the intestine is hypertrophied with normal-appearing villi but has defective peristalsis.[28][29] Both in experimental animals and human newborns, there is a reduction in mucosal enzymes, adenosine triphosphatase production, but hypertrophic and hypercellular ganglia with increased acetylcholinesterase activity in bowel close to the atretic segment.[28][29]

The extent of JIA has been shown to depend on the location and size of vascular compromise, as demonstrated by a complete separation of the atretic ends of the bowel and the accompanying mesenteric defect between the ends seen when mesenteric vessels are disrupted close to the origin.[8] In situations of incomplete vascular compromise, intestinal stenosis has been reported instead. Studies have shown that JIA is mostly due to local vascular events that can occur in situations that favor occlusion or kinking of blood supply to the intestine such as internal hernias, volvuli, intussusceptions, different from duodenal atresia which is usually seen in cases with associated malformation of other systems thereby implicating general factors such as fetal hypoxia instead of local factors as responsible.[8]

Historically, type II and type III JIA with blind ends have high mortality due to functional obstruction from defective peristalsis after direct anastomosis of proximal and distal blind ends.[8] The defective peristalsis and the mucosal necrosis of the blind ends seen in some cases of JIA are as a result of vascular insufficiency.[8] It has been postulated that the damage done to the blind ends is not enough to cause the death of the areas, but being close to the infarcted part of the bowel, a conclusion can be drawn that these areas are left with insufficient blood supply.[8] Therefore, resection of the dilated proximal blind end and the proximal part of the distal atretic end in JIA has been reported with better functional outcomes.

**Histopathology**

The histopathology for jeunoileal atresia (JIA) is a proximal distended hypertrophied blind loop of the intestine with normal-appearing villi but has defective peristalsis.[28][29] The mean thickness of the inner circular muscle and outer longitudinal muscle layers of the proximal segment is higher within 3cm-5cm of atretic end in both ileal and jejunal atresia. The interstitial cells of Cajal (ICC), which is important for gastrointestinal motility, is lesser in the proximal and distal atretic ends of JIA than in healthy bowel.[30] The concentrations of ICC vary at different locations on the proximal segment with the number of ICC at 8 cm of the proximal segment greater than the number at 3 cm and 5 cm
of the proximal segment of JIA [30]. Both in experimental animals and human newborns, there is a reduction in mucosal enzymes, adenosine triphosphatase production, but hypertrophic and hypercellular ganglia with increased acetylcholinesterase activity in bowel close to the atretic segment.[28][29]

History and Physical

A complete history and physical examination are needed for the diagnosis of jejunoileal atresia. History includes gestational age at birth, complications during pregnancy, family history, co-morbidities, illicit drug use, the result of prenatal screening such as ultrasound evidence of obstruction like polyhydramnios, dilated bowel, and congenital anomalies such as down syndrome. Perinatal history, including problems with the delivery, feeding history, onset of abdominal distension, bilious or non-bilious emesis, and passage of meconium, should be documented. Physical examination should include a general assessment of the severity of illness, abdominal distension, evidence of peritonitis, respiratory compromise from aspiration or splitting of the diaphragm, signs of dehydration, jaundice and congenital anomalies down syndrome, congenital heart disease, and anorectal malformation.

Evaluation

Intestinal atresia may be suspected in-utero with suspicious prenatal ultrasound findings, and in a neonate, obstructive symptoms such as abdominal distension, bilious emesis are usually the presenting symptoms.

Prenatal detection of jejunoileal atresia (JIA) on ultrasound, based on the evidence of intestinal obstruction, has been documented in some series with the rate of detection ranging from 29% to 50%.[2][6][7] JIA may be seen on ultrasound as polyhydramnios, ascites, dilated bowel loops, and enhanced bowel echogenicity.[31][32][33] Prenatal diagnosis may enhance care by preparing clinicians and parents so that the neonate can receive prompt care, and complications associated with early feedings such as emesis, electrolyte imbalance, and aspiration can be avoided. Prenatal ultrasound has low sensitivity, especially for distal lesions; it cannot determine the number of atresias or identify the location of the obstruction and not capable of assessing the viability of the gut distal to the obstruction.[34][35] Lesions in the proximal intestine (jejunum) are more likely to be detected compared to lesions in the distal lesion, and this is a result of proximal bowel dilation from net swallowed amniotic fluid.[34][35]

Postnatally, JIA presents with signs and symptoms of intestinal obstruction such as abdominal distension, emesis, and in some cases, delayed passage of meconium. Normal appearing meconium may be seen, but most often, light-colored plugs are passed from the rectum. In cases of distal bowel ischemic, as seen in type IIIb, blood may be seen in the rectum.

Radiographic examination of the abdomen with plain abdominal X-ray using swallowed air as a contrast is a useful diagnostic tool. For proximal JIA, there are presences of few dilated proximal bowel with no distal gas. Intraperitoneal calcification can be seen in prenatal bowel perforation or meconium peritonitis. Malrotation is ruled out using an upper gastrointestinal series contrast study. A contrast enema is used to reveal the atypical appearance of the colon, as may be seen in meconium ileus or Hirschsprung disease. If the contrast passes into the dilated loop of bowel, then atresia is ruled out. Other studies such as echocardiogram, renal ultrasound, rectal biopsy, and cystic fibrosis screening may be used to evaluate patients for associated congenital anomalies such as cardiac malformation, renal anomalies, Hirschsprung disease, and cystic fibrosis.

Treatment / Management

Following confirmation or suspicion of the diagnosis of jejunoileal atresia, preoperative management includes decompression with a nasogastric tube, fluid and electrolyte resuscitation, and intravenous broad-spectrum antibiotics in the event of perforation or evidence of infection.

The surgical approach depends on the location of the lesion, the anatomy, intraoperative condition, and the remaining bowel length.[7] The most common technique is resection of proximal dilated and atretic bowel with primary end to end anastomosis with or without tapering enteroplasty of the proximal bowel.[36][10][37] The decision for a temporary ileostomy is individualized and is performed when there is questionable bowel viability, significant size...
discrepancy between proximal and distal bowel or intestinal perforation.[7][6][38] Surgery can be laparoscopic-assisted or open, and the decision for the surgical approach depends on the surgeon's preference, the patient's presentation, and anatomy. In a laparoscopic-assisted approach, the bowel is exteriorized through the umbilical incision and returned into the abdominal cavity after primary anastomosis. In open surgery, traditional transverse supra- or infraumbilical incisions are used; however, a circumumbilical incision is found to be cosmetically better while producing the same outcomes as the transverse abdominal incisions.[39] One of the retrospective studies reported shorter operative time, time to start, and time to full enteral feedings in the laparoscopic-assisted approach compared to open surgery. Still, there were no differences in postoperative complications, morbidity, or mortality between the two groups.[38][20] However, if the proximal small bowel is significantly distended, laparoscopy has limited utility as the neonatal bowel can be exteriorized through a significantly small incision.

Two specific caveats are the child with multiple segmental atresias and the child with the atretic lesions separated by a short segment. Primary surgical repair is usually performed for multiple atresias to preserve bowel length (generally with a proximal protective diversion to allow the numerous distal anastomosis time to heal). In patients with the atresia separated by a very short segment, resection of the short atresia with primary anastomosis is advocated to decrease the number of intestinal anastomoses, provided the patient has sufficiently normal intestinal length.

**Differential Diagnosis**

Conditions such as intestinal malrotation with midgut volvulus, internal hernia, congenital small left colon syndrome, Hirschsprung disease, meconium ileus, colonic atresia presents with signs and symptoms of bowel obstruction similar to jejunoileal (JIA). Many patients with malrotation may be asymptomatic but are at increased risk of midgut volvulus. Volvulus can present at any age but are seen in 30% of children less than one-month-old.[40] Both volvulus and JIA presents with emesis, and malrotation is excluded using upper gastrointestinal series. Most presentations of Hirschsprung’s disease are seen during the neonatal period with signs and symptoms of intestinal obstruction or persistent abdominal obstruction following surgical anastomosis of small bowel at the JIA site. Contrast enema and rectal biopsy are utilized to exclude Hirschsprung disease from other causes of intestinal obstruction. In cystic fibrosis patients, meconium ileus is an important cause of intestinal obstruction similar in presentation to JIA. Like JIA, prenatal screening for cystic fibrosis and prenatal ultrasound findings with evidence of meconium ileus should trigger early workup and management to minimize complications associated with cystic fibrosis.[41]

**Prognosis**

The mortality for patients with jejunoileal atresia (JIA) has approached zero over the past several years.[31] Advancement in pediatric anesthesia, surgical technique, and total parenteral nutrition has been credited with reduced morbidity and mortality. The prognosis of JIA depends on the presence of short bowel syndrome (SBS) with intestinal length less than 25cm, requiring long-term parenteral nutrition.[6] The risk for SBS is more in patients with type III and type IV JIA. JIA is responsible for about 10% of intestinal failure, and the two most popular bowel lengthening procedures are serial transverse enteroplasty procedure (STEP) and longitudinal intestinal lengthening procedure (LILT).[6] SBS and cardiac anomalies have been largely responsible for morbidity and mortality.[18][2]

In type IIIb JIA, poor outcomes are associated with prematurity, low birth weight, and associated anomalies.[20] Type IV (multiple-segment atresia) is associated with dysfunction of the central nervous symptoms in 25% of nonfamilial cases and severe immunodeficiency linked to a mutation in the tetratricopeptide repeat domain-7A (TTC7A) gene.[42][43][44] The familial form of type IV is fatal and is associated with prematurity and reduced intestine length.[45] [46]

**Complications**

Postoperative complications include sepsis and anatomic leak with the varying rate among studies and the reported rate of 5% to 8% and 5% to 7% respectively in two of the retrospective studies.[6] Other important complications include adhesive bowel obstruction and short bowel syndrome (SBS). SBS is one of the major complications with a
prolonged hospital stay, more feeding problems, increased rate of infection, morbidity, and mortality compared to patients without SBS.[6]

**Postoperative and Rehabilitation Care**

While awaiting the return of bowel function, intravenous hydration and decompression with a nasogastric or orogastric tube are continued. Enteral feeding is started with the return of bowel function, and the feeding rate is advanced as tolerated until goal feed rate is achieved. Oral intake is resumed when the patient is awake and able to suck. In some select patients, parental nutrition is started in the immediate postoperative period, while some patients will require parenteral nutrition later in the postoperative course due to prolonged ileus.

**Deterrence and Patient Education**

Jejunoileal atresia (JIA) is a condition where there is no opening or passage through a segment of the small intestine (jejunum and atresia). It occurs in 1 in 5000 to 1 in 14000 live births and equally in males and females. It is generally accepted that jejunoileal atresia is caused by an in-utero vascular accident during fetal development. It may be associated with other anomalies such as cystic fibrosis, and cardiac abnormalities.

Prenatal screening with ultrasound demonstrates evidence of intestinal obstruction in 29% to 50% of cases. At delivery, the infant will not be fed if this diagnosis is suspected prenatally. However, if the infant is fed, there will be episodes of emesis and abdominal distension.

Diagnosis is made using an abdominal radiograph, which showed air in the proximal small intestine and no air distally. Upper gastrointestinal series and contrast enema are done to exclude other conditions such as malrotation and Hirschsprung disease. Conditions that mimic this condition are malrotation with midgut volvulus, internal hernia, meconium ileus, Hirschsprung’s disease.

Treatment is with surgical intervention. Following surgical intervention, the infant is first stabilized by rehydration and correction of electrolyte and placement of a nasogastric or orogastric tube to remove fluid from the obstructed stomach and small intestine. The procedure is performed either through a laparoscopic-assisted approach or through an open surgical approach with a single incision on the abdomen. The blockage in the small intestine is identified, and the abnormal part of the intestine is resected and removed, after which the proximal and distal ends are joined together.

Postoperatively, nasogastric tube drainage is continued and nothing to eat by month. It will take the bowel days or even weeks to begin normal function. Oral feeding is started when there is a small fluid coming out through the nasogastric tube. Pain is controlled with acetaminophen or ibuprofen or opioids for more intense pain. The risk associated with surgical anastomosis is a leak at the bowel anastomosis, wound infection, prolonged recovery of the bowel, and shortened intestinal length.

At home, the child continues a normal diet for age, unrestricted activity, and the incisions can be washed with water and soap. Do not submerge under water until at least a week after surgery when the wound must have healed. Give pain medication as needed for pain control. Call your doctor or bring your child to the hospital if you notice redness warmth or drainage from the incision, vomiting, and fever. Follow up with your surgeon in 2 to 4 weeks after surgery for wound checks.

**Pearls and Other Issues**

Historically, jejunoileal atresia is associated with high mortality and morbidity. However, advances in pediatric anesthesia, surgical technique, intensive care unit (ICU) care, and nutritional supports over the years have led to better overall survival in patients with intestinal atresia. Resection of the dilated proximal intestine with end to end anastomosis with or without tapering of the proximal intestine is one of the advances in surgical techniques that have been found to prevent the high mortality associated with the dysmotility of the proximal dilated bowel and the resultant chronic obstruction. The understanding of the proximal bowel dysfunction followed with the improvement in
the surgical technique, along with total parenteral nutrition, has been vital in improving the prognosis of jejunoileal atresia.

Prevention of jejunoileal atresia might not be possible, but close monitoring of high-risk pregnancy and prenatal screening with ultrasound would be helpful for the early diagnosis and prevention of complications.

Enhancing Healthcare Team Outcomes

Jejunoileal atresia (JIA) is the most common cause of intestinal obstruction in neonates. It has been generally reported that JIA occurs as a result of a vascular accident involving mesenteric blood supply in-utero. The atresia can be found anywhere in the small bowel either as solitary or multiple lesions. Advancement in pediatric surgical techniques, pediatric anesthesia, intensive care unit (ICU) care, and nutritional supports over the years has led to better overall survival in patients with intestinal atresia.

While the surgeon may have the primary responsibility for the care of the patient, other members of the interprofessional team include the neonatologists, pediatric anesthesiologists, nutritionists, pediatric nurses, other consultants like cardiologists, or pulmonologists for patients with associated cardiac anomalies and respiratory problems from cystic fibrosis. Each member of the team contributes to the overall care of the patients. Nurses closely monitor the patient's vital signs and provide nursing care. Neonatologists provide neonatal care, especially for patients admitted to neonatal intensive care units. Pharmacists are an important member of the team to ensure that patient is on the appropriate analgesics, and maintain antibiotics stewardship. Nutritionists determine the nutritional requirement of each patient and make an appropriate recommendation. Consultants such as cardiologists and pulmonologists are tasked with managing cardiac and pulmonary complications from associated anomalies.

Questions

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References


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