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INTESTINAL ATRESIA WITH MULTILOBED LIVER: A CASE REPORT

Dr Dnyaneshwar, Ch Sudhakara Babu

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***Correspondence for Author**
Dr Dnyaneshwar

ABSTRACT

Objective To evaluate the causes, clinical presentation, diagnosis, operative management, postoperative care, and outcome in infants with intestinal atresia. **Design** Retrospective case series. **Setting** Pediatric tertiary care teaching hospital. **Interventions** Patients with duodenal obstruction were treated by duodenoduodenostomy , Patients with jejunoileal atresia were treated with resection and anastomosis; tapering enteroplasty, or temporary ostomy, web excision, and the Bianchi procedure . Patients with colon atresia were managed with initial ostomy and delayed anastomosis. **Main Outcome Measures** Morbidity and early and late mortality. **Conclusions** Cardiac anomalies (with duodenal atresia) and ultrashort-bowel syndrome (<40 cm) requiring long-term total parenteral nutrition, which can be complicated by liver disease (with jejunoileal atresia), are the major causes of morbidity and mortality in these patients. Use of growth factors to enhance adaptation and advances in small bowel transplantation may improve long-term outcomes.

KEY WORDS: Cardiac anomalies, diagnosis, jejunoileal atresia.

INTRODUCTION

Intestinal atresia or stenosis can occur anywhere along the GI tract, and the anatomical location of the obstruction determines the clinical presentation. Most newborns present with bilious emesis, a distended upper abdomen, and a scaphoid hypogastrium. Signs of dehydration are not uncommon. Jaundice is present in 32% of these children. A history of polyhydramnios on prenatal ultrasonography (28%), prematurity (35%), or low birth weight (25-50%) can be helpful hints in the diagnosis[1].

The different types of intestinal atresia are named after their location

1. Duodenal atresia - malformation of the duodenum, part of the intestine that empties from the stomach
2. Jejunal atresia - malformation of the jejunum, the second part of the intestine extending from the duodenum to the ileum
3. Ileal atresia - malformation of the ileum, the lower part of the small intestine
4. Colon atresia - malformation of the colon

Duodenal atresia has a strong association with Down syndrome.[1] It is the most common type, followed by ileal atresia The most common cause of non-duodenal intestinal atresia is a vascular accident in utero that leads to decreased intestinal perfusion and ischemia of the respective segment of bowel. This leads to narrowing, or in the most severe cases, complete obliteration of the intestinal lumen.

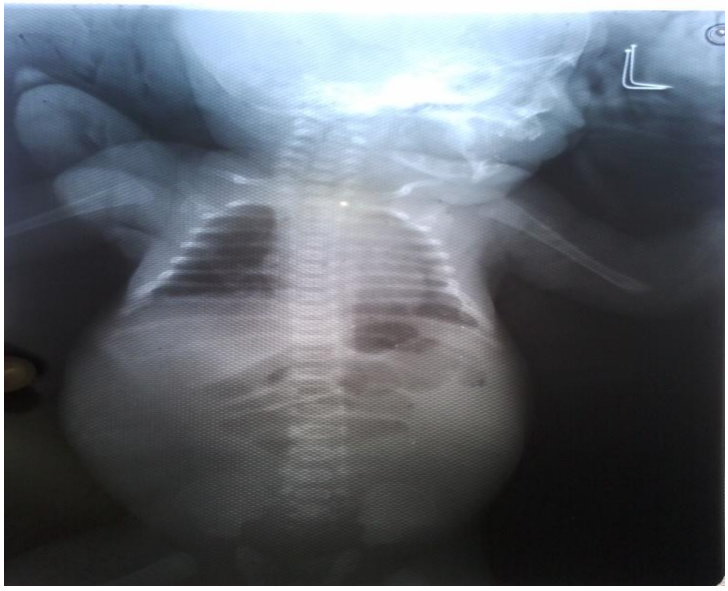
In the case that the superior mesenteric artery, or another major intestinal artery, is occluded, large segments of bowel can be entirely underdeveloped. Classically, the affected area of bowel assumes a spiral configuration and is described to have an "apple peel" like appearance; this is accompanied by lack of a dorsal mesentery. Ileal atresia can also result as a complication of meconium ileus.

Intestinal atresias are often discovered before birth: either during a routine sonogram which shows a dilated intestinal segment due to the blockage, or by the development ofpolyhydramnios (the build up of too much amniotic fluid in the uterus). These abnormalities are indications that the fetus may have a bowel obstruction which a more detailed ultrasound study can confirm.

Some fetuses with bowel obstruction have abnormal chromosomes. An amniocentesis is recommended because it can determine not only the sex of the baby, but whether or not there is a problem with the chromosomes. Fetal and neonatal intestinal atresia are treated using laparotomy after birth. If the area affected is small, the surgeon may be able to remove the damaged portion and join the intestine back together. In instances where the narrowing is longer, or the area is damaged and cannot be used for period of time, a temporary Stoma (medicine) may be placed.

CASE REPORT

A newborn male baby presented to dept of paediatrics Koppal institute of medical sciences koppal with history of bilious vomiting and bloated abdomen. Neonate also presented with constipation, pain abdomen ,cry of unknown etiology.On examination baby looks dehydrated, swollen abdomen. Abdominal radiograph shows multiple air fluid levels in supine and erect posture.



At the time of surgery, newborn had colon atresias, 3 with type I (mucosal web), with type II (fibrous cord), with type IIIa (mesenteric gap defect). The colonic atresia was located in the right colon, transverse colon , and left colon . Newborn was treated with the formation of a temporary stoma , right colostomy , transverse colostomy and left colostomy followed by

delayed bowel anastomosis at age 3 to 4 months. The postoperative and long-term survival rate was 100%. Complications included wound infection , colostomy prolapse , prolonged ileus , and late adhesive bowel obstruction .

DISCUSSION

Intestinal atresia is a common cause of neonatal intestinal obstruction. The causes, clinical presentation, diagnosis, operative management, postoperative care, and outcome may vary considerably according to the location of the obstruction. The 2 major theories regarding the etiology of intestinal atresia are Tandler's[2] concept of a lack of revacuolization of the solid cord stage of intestinal development and the classic study by Louw and Barnard[3] suggesting that a late intrauterine mesenteric vascular accident is the cause of most jejunoileal and colonic atresias. While lack of revacuolization is the probable cause for most cases of duodenal atresia, compelling observations from other studies[4- 10] demonstrate that jejunoileal atresias occur as a result of intestinal volvulus, intussusception, internal hernia, or strangulation in a tight gastroschisis or omphalocele defect. Familial instances of jejunoileal and colonic atresias have also been observed, suggesting that genetics may play a part in these cases.[11- 13]

The classification of intestinal atresias varies somewhat based on the location of the obstruction. Duodenal lesions historically have been classified by the method described by Gray and Skandalakis,[14] with identification of 3 types of lesions. A type I defect represents a mucosal web with normal muscular wall (most common); type II, a short fibrous cord connecting the 2 atretic ends of the duodenum; and type III (least common), one in which there is complete separation of the atretic ends. The classification of jejunoileal atresias initially proposed by Louw[15] also recognized 3 types of lesions. The classification was later refined by Martin and Zerella[16] and by Grosfeld et al[17] to include the apple peel deformity and multiple atresias. According to this method of classification, type I defect represents a mucosal defect with an intact mesentery. Type II defects consist of a fibrous cord connecting the atretic bowel ends. A type IIIa lesion denotes an atretic segment with a V-shaped mesenteric gap defect, while type IIIb defines the apple peel deformity, in which there is a proximal jejunal atresia and the distal bowel is supplied by a single retrograde blood vessel. Type IV describes instances of multiple atresias ("string of sausage" effect). This classification system has also been applied to colonic atresia, with most of those cases demonstrating a type IIIa defect.

The presenting symptoms for patients with any form of intestinal atresia or stenosis are consistent with bowel obstruction and include bilious vomiting, abdominal distension, and failure to pass meconium in instances of lower obstruction. Most infants with duodenal obstruction do not have significant abdominal distension. The number of cases of intestinal atresia that are identified prenatally has increased as a result of the routine use of ultrasonographic monitoring of fetal development. Various studies[18,19] have evaluated the need to perform prenatal examination early (≥18 weeks' gestation) and have shown the benefit of prenatal diagnosis with earlier recognition, prompt surgical intervention, and fewer metabolic complications. Prenatal ultrasonography may identify the presence of maternal polyhydramnios and distension of the stomach and duodenum with swallowed amniotic fluid. These observations are often associated with a high risk of duodenal atresia. Although gastroschisis can frequently be observed on a prenatal ultrasonogram, a notable number of jejunoileal and colonic atresias remain undetected by this method. Because ultrasonography generally has not been an accurate determinant for lower intestinal obstruction, its ability to exclude a malformation is limited.

Postnatally, radiographic studies are useful in the diagnosis of intestinal atresia. Plain abdominal radiographs can identify the classic double-bubble sign (air-filled stomach and proximal duodenum) of duodenal atresia or stenosis. Lack of bowel gas beyond the second bubble is usually diagnostic of an atresia. Jejunoileal or colonic atresias are characterized radiographically by the presence of proximal dilated intestinal loops often associated with air-fluid levels. The presence of intraperitoneal calcifications are indicative of meconium peritonitis and suggest an intrauterine bowel perforation. An upper gastrointestinal contrast-enhanced radiograph should be obtained in cases of partial obstruction and may demonstrate the presence of a stenosis or a mucosal web with a small opening. As the colonic haustral markings are not prominent in the neonate, it may be difficult to discern small bowel from large bowel on plain abdominal radiographs. A barium enema will define the location of a colonic atresia; it is also recommended for instances of presumed jejunoileal atresia to identify whether the colon has been unused (microcolon) during intrauterine life, define the level of obstruction in the small intestine, occasionally identify the presence of obstructing intraluminal meconium pellets in instances of meconium ileus, and identify the location of the cecum to rule out anomalies of rotation and fixation.

CONCLUSION

The survival rate for infants with intestinal atresia, in our experience, has remained relatively stable for the last 2 decades. The major obstacles to a successful outcome continue to be complex congenital heart disease in infants with duodenal atresia and the challenges in the treatment of children with jejunoileal atresia and ultrashort-bowel syndrome, especially those cases complicated by TPN-related liver disease. Early use of growth factors to maximize intestinal adaptation, administration of growth hormone to "grow the bowel," and nutritional modifications may improve the status of patients with short-bowel syndrome.[20] Refinements in small-bowel transplantation and advances in the science of immunology should reduce the current high complication rate and improve graft and patient survival, eventually improving the overall survival rate for these young patients.

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