

Near Total Jejunio-Ileal Atresia: A Management Challenge

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ABSTRACT

A 2-day-old female neonate with the clinical picture of proximal small bowel atresia, on exploration, turned out to have intestinal atresia of a rare variety, i.e., a near-total jejunio-ileal atresia. The baby had total small bowel length of less than 10 cm. She survived for 3 months on enteral feeding after end-to-back duodeno-ileal anastomosis and thereafter succumbed to septicemia. The case is presented for its extreme rarity and consideration of this extreme form of small bowel atresia as an offshoot of the existing classifications of jejunio-ileal atresia since it has dismal prognosis and presents as a management challenge even today.

Key words:

Classification of small bowel atresia, intestinal obstruction, jejunio-ileal atresia, near total small bowel atresia, neonate, total parenteral nutrition

INTRODUCTION

Intestinal atresia is commonly seen as one of the four types described in any of the currently existing Pediatric surgery specialty literature.^[1,2] None of the classifications in vogue today, include long gap bowel atresias like a near-total/total jejunio-ileal atresia. Though rare, long gap bowel atresias are difficult to treat. They require specially tailored surgical management. Various treatment options are available today. However, existence of a certain minimum length of small bowel is the most crucial factor affecting the outcome of repair, overall salvage of the infant and long-term prognosis of such cases.

CASE REPORT

A female neonate with birth weight of 1.7 Kg, born to a healthy adolescent primigravida by preterm vaginal delivery at 34 weeks, was referred to us on D₂ of life because of non-passage of meconium. There was no history of any drug ingestion/exposure during early pregnancy. The mother was normotensive throughout her pregnancy and had received regular antenatal care. The baby had cried immediately after birth. She had passed urine, but no meconium. On examination, the abdomen was scaphoid. External genitalia and the anal opening were normal. The nasogastric tube aspirate was bilious.

Erect X-ray of the abdomen showed two dilated loops in the upper abdomen; with paucity of gas in the pelvis [Figure 1]. Ultrasonography showed few distended bowel loops mainly in the pelvis with sluggish peristalsis. The liver, spleen, and kidneys were normal. The hematological investigations were normal. The baby was explored with clinical diagnosis of “proximal small bowel atresia.”

On exploratory laparotomy, there was evidence of high jejunal atresia. Entire small bowel, 2-3 cm beyond the duodeno-jejunal (DJ) flexure was atretic. The stomach was dilated and thickened. The duodenum was massively dilated and tortuous and had a length of approximately 7-8 cm [Figure 2]. The jejunum was blind ending and had a length of about 2-3 cm. There was an additional type-I atresia at the DJ flexure. Distally, only 3-4 cm of ileum was present proximal to the IC junction [Figure 3]. The colon was unused but, could be distended by saline infilling.

To preserve the small bowel length as much as possible, single layer extramucosal end-to-back anastomosis was performed between the dilated duodenum and spatulated end of ileum (duodeno-ileal anastomosis) using 5-0 Polyglactin. Enterotomy and web excision was done for type-I atresia at the DJ flexure. Immediate post-operative period was uneventful. After surgery, the baby had a slow recovery of intestinal function. Dye study carried out on the post-operative day 20 showed free flow of contrast across the anastomosis, though it was albeit slow.

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Figure 1: Erect X-ray of the abdomen showing two dilated loops in the upper abdomen; with the paucity of gas in the pelvis

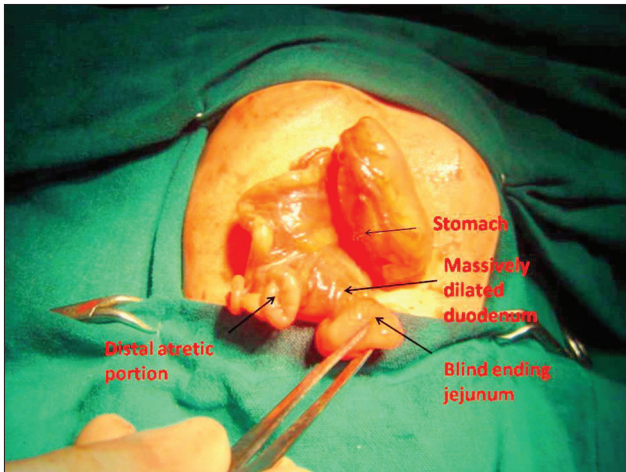


Figure 2: Operative photograph showing massively dilated, tortuous duodenum and atretic jejuno-ileum

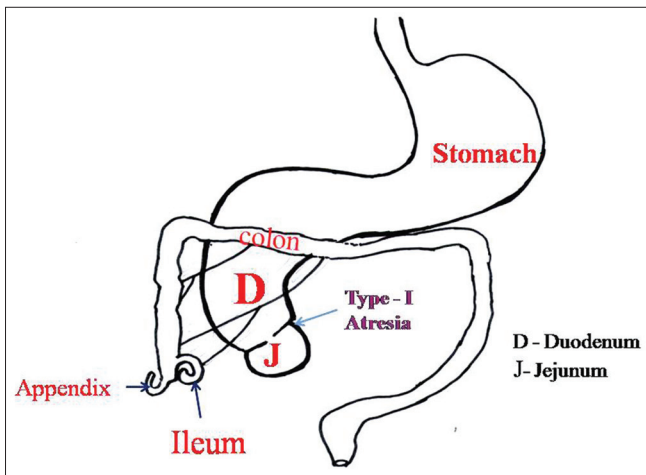


Figure 3: Schematic representation of the atresia

The baby received total parenteral nutrition (TPN) for about 3 weeks. The TPN was gradually tapered, when formula feeds were well-tolerated to meet the child’s daily

requirement. However, the family could not afford special elemental diet for long and the baby had to be put on expressed breastmilk (EBM) feeds. The EBM feeds were progressively increased upto 140 ml/day, when she was discharged. However, the baby had very little weight gain and eventually succumbed to intractable sepsis at 14 weeks of life.

DISCUSSION

The currently existing classifications in standard international literature viz. the Grosfeld Modification of Louw Classification (1979) describe four types of intestinal atresias: Type I or mucosal web, type II or fibrous cord, type IIIa or mesenteric gap defect, type IIIb or “applepeel” atresia and type IV or multiple atresias.^[1,2] Intestinal atresias were initially thought to be produced by a “failure of recanalization” of the solid cord stage of the bowel (Tandler1900). However, this theory has been confirmed only for atresia of the duodenum.^[2] Later work confirmed the theory of Spiggs that the origin of intestinal atresias is mechanical accidents, including vascular occlusions. This theory was confirmed further by Louw and Barnard (1955) who obtained intestinal atresias by inducing mesenteric vascular accidents in puppies.^[1,2] They confirmed that intrauterine clinical instances of intestinal volvulus, intussusception, internal hernia or even constriction of the intestines in a tight gastroschisis could lead to intestinal atresia.^[2]

None of the studies published so far mentioned the possibility of getting a near-total/total jejuno-ileal atresia. However, it can be postulated that an insult of the jejunal and ileal branches of the superior mesenteric artery or a very large intrauterine midgut volvulus and resultant resorption of the entire small bowel during early intrauterine life could be the possible reasons for such extensive atresia.

The standard treatment for commonly occurring jejuno-ileal atresias is exploratory laparotomy upon initial hemodynamic stabilization. The grossly dilated aperistaltic terminal portion of the proximal bowel is excised and anastomosed end-to-back to spatulated distal atretic segment. Recovery of bowel function depends upon the degree of dilatation of the proximal bowel, residual length of the small bowel and presence/absence of the IC valve. The outcome of neonates born with jejuno-ileal atresias has improved considerably in recent years due to many new acquisitions in neonatal intensive care and anesthesia, operative procedures and use of TPN.^[1,3]

However, the treatment of long gap intestinal atresias has not been standardized. Various available treatment options are described in isolated case reports with guarded

success. Federici *et al.* have reported use of gastroduodenal intraluminal silicone tube stents across five successive anastomoses in a case of apple peel atresia with ultimate salvage of the baby.^[4] The Bianchi technique^[5,6] or the serial transverse enteroplasty procedure^[7] have been employed for operative lengthening of residual short bowel syndrome. All these procedures started from some available small gut, but for a near-total jejuno-ileal atresia, as the one described in this case, the only logical and life-saving option would be an intestinal transplantation, a method available in only very few institutions in the world. Though the results of intestinal transplantation in 6 children with intestinal atresia as reported by Nishida *et al.*^[8] have been encouraging, for lack of highly advanced neonatal health-care infrastructure, it is still not possible in many centers including ours.

Prognosis of long gap bowel atresias is universally dismal. Nixon *et al.* have applied the Waterston classification to identify three risk groups in long gap bowel atresias. They have reported 0% survival rate for Group C with high jejunal atresia (within 15 cm of DJ flexure).^[2,9] Wilmore's review of 50 infants with significant small bowel resection suggests that though most infants with >35 cm of residual intestine and intact IC valve survive, survival drops to 50% with bowel length of 15-25 cm and decreases further to 0% in infants with <15 cm residual small bowel even with intact IC valve.^[3] Child in the current report being <1.8 Kg, belongs to Group C of the Waterston classification. Furthermore, the total small bowel length in this neonate from pylorus to IC junction (including the duodenum) was <10 cm; which is incompatible with survival with traditional methods of treatment.

This case is reported for its extreme rarity, dismal prognosis and the management challenge it presents even today. We strongly propose an addition of near-total jejuno-ileal

atresia, as an offshoot to the presently existing classifications of small bowel atresia. By collecting the database of this extremely rare form of jejuno-ileal atresia, in near future we might be able to formulate management guidelines to salvage these neonates for whom we can offer no definitive solution today.

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