Chirurgia (2013) 108: 407-410
No. 3, May - June
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Associated Type IIIB and Type IV Multiple Intestinal Atresia in a Pediatric Patient

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Rezumat

Atrezie intestinală multiplă asociind leziuni tip IIIB și IV. Studiu de caz

Atrezia intestinală multiplă este o malformație complexă care reprezintă o încercare pentru chirurgul pediatru, mai ales dacă este diagnosticat tipul IIIb de atrezie, la care mortalitatea este foarte mare. Scopul intervenției chirurgicale este acela de a obține o lungime viabilă cât mai mare a intestinului, de a evita sepsisul postoperator și de a preveni complicațiile pe termen lung, cum ar fi sindromul de intestin scurt. Existența unei secții de terapie intensivă neonatală și suportul nutrițional parenteral sunt esențiale pentru supraviețuirea acestor pacienți. Prezentăm cazul unui nou-născut diagnosticat cu atrezie intestinală asociind atrezie "în coajă de măr" la care s-a intervenit chirurgical, realizându-se rezecții și anastomoze intestinale multiple.

Cuvinte cheie: atrezie jejunală, atrezie "în coajă de măr", anastomoză termino-terminală, nutriție parenterală

Abstract

Multiple intestinal atresia (MIA) is a complex congenital defect which represents a challenge for the pediatric surgeon, especially in the rare event of encountering type IIIb or apple peel atresia, which has a high mortality rate. The surgeon's aim is to preserve as much bowel length as possible, to avoid postoperative sepsis and to prevent long-term complications such as short bowel syndrome. Access to a good neonatal intensive care unit and to parenteral nutritional support is crucial in the survival of these children. We report a rare case of multiple intestinal atresia associated with an apple peel atresia, which was managed by multiple intestinal resections and anastomosis without the placement of transanastomotic tubes or stomas.

Key words: jejunal atresia, apple peel atresia, end-to-end anastomosis, parenteral nutrition

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Introduction

Intestinal atresia is one of the most common causes of intestinal obstruction in newborns, with an incidence of 1-3 in 10,000 (1,2). Jejunoileal atresia occurs more frequently than duodenal or colonic atresias and, while single atresias (type I, II, and IIIa) are most commonly encountered, 6–12% of infants are diagnosed with multiple atretic segments (3,4,5). The association of type IIIb or apple peel atresia with multiple intestinal atresia (MIA) is exceptional and has a high mortality rate (6).

Regarding the surgical management of such patients, great care must be placed into preserving bowel length by any means necessary (through multiple end-to-end anastomosis) and assisting the enteral digestive functions by using parenteral nutrition and early enteral feeding (6,7).

This report describes the case of a 1.850 g premature

newborn presenting 3 small bowel atresias associated with an apple peel deformity and discusses the management of this very rare pathology.

Case report

A 33-week preterm baby, weighing 1850 g, with a prenatal diagnosis of upper digestive tract malformation was referred to the Surgical Ward of our clinic 24 hours after birth. A plain abdominal radiography taken on admission showed "double-bubble" images (Fig. 1).

After having been stabilized, on the 2nd day after admission, the newborn underwent a laparatomy procedure through an umbilical incision. The intraoperative findings included an extremely dilated proximal jejunum which ended as a blind pouch about 10 cm from the duodenojejunal flexure and a distal jejunum which was shortened because of 3 separate type IV atresias (with V-shaped mesenteric gaps). These 2 segments between these atretic areas each measured about 15 cm. A typical apple peel (type IIIb) was also encountered (Fig. 2A, B). The distal small intestine measured about 40 cm. The ileocecal valve and the caecum were situated higher in the right upper quadrant due to the malrotation and malposition of the intestine. About 3 cm of the proximal jejunum were resected and the remaining segment was then remodelled in order to obtain an appropriate calibre. A partial resection of the atretic areas was performed: 1-2 cm of each end of the 2 affected segments were removed and 3-4 cm from the distal end were resected. The remaining segments were preserved and connected by 3 end-to-end anastomoses. Great effort was taken in order not to touch or twist the apple peel tract. At the end of the surgery, the total length of the small intestine was of about 70 cm. The reconstructed bowel together with the apple peel tract were carefully placed in the abdominal cavity.

The newborn received parenteral nutrition for the first 12 days after surgery, but feeding also started on day 4, first by using glucose serum then, after testing the digestive tolerance, an amino acid milk formula (Neocade). The baby was discharged on day 45 after the surgery, and by that time he was receiving a full meal (formula) and gaining weight.



Figure 1. *Double-bubble image*

At the 2-year follow-up, the child showed a normal growth, with a good weight gain and overall development for his age, without any symptoms of short bowel syndrome such as malabsorbtion, diarrhea or malnutrition.

Discussion

Intestinal atresia is a frequent cause of neonatal bowel obstruction, with multiple intestinal atresia being reported in 6-12% of cases. The number of interruptions in MIA may vary from 9 to 23, the small intestine being usually affected, although both the duodenum and the large bowel can be

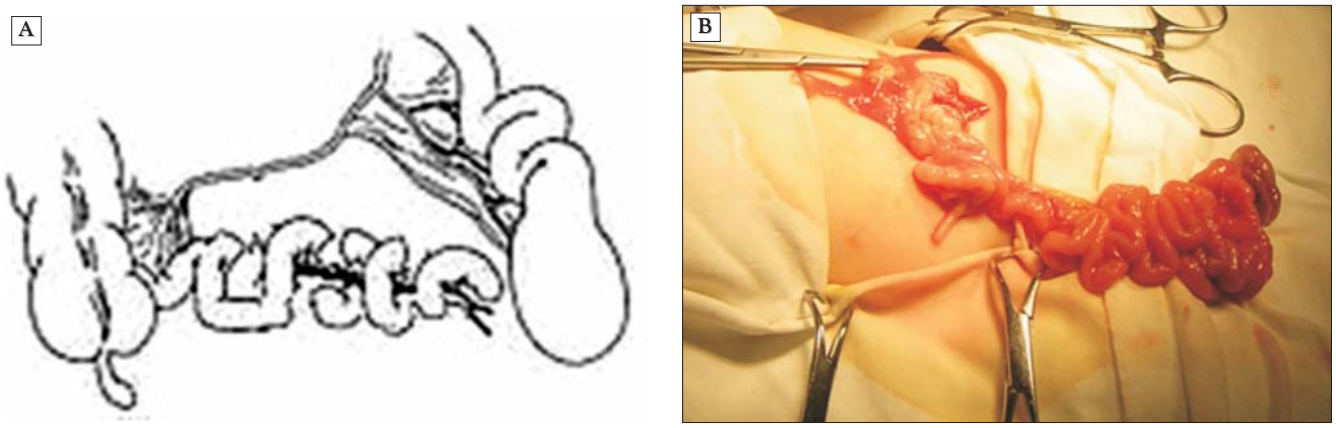


Figure 2. ***(A)*** *Apple-peal disposition;* ***(B)*** *Intraoperative image of the apple-peal*

involved (6,8). The coexistence of all 4 types of jejunoileal atresia in the same patient is exceptional and seems to be the result of multiple ischemic lesions of the intestinal arteries (4,9).

In the past decades, due to the many improvements in neonatal intensive care, operative techniques and parenteral nutrition protocols, the mortality rate has visibly decreased from 90% in the first half of the last century to 16% in the last couple of years. The most important cause of mortality remains short bowel syndrome, encountered in 65% of infants with extensive atresia, types IIIb or type IV (10,11).

The intraoperative aspect of the distal bowel and of the colon is important in order to exclude the association with Hirchsprung's disease (0.8%) (10,12). In our case the biopsies which were taken during surgery have shown no signs of aganglionosis.

When choosing the most appropriate surgical treatment for intestinal atresia, one must take into account the specific type of multiple atresia and all other circumstances pertaining to each case, which can be severely limiting. Newborns presenting this type of lesions are usually preterm infants with low birth-weights, who can associate any to all of the dysfunctions characteristic for preemies; in our case, the child was a 33 weeks preemie, weighing 1850 g at birth, with respiratory distress requiring intensive care with stabilization and supervision, before and after surgery (7,13).

The short residual intestinal length together with the ineffective peristalsis of the proximal segment are the main limitations of the surgical treatment for multiple atresias (6). The usually dilated proximal bowel, which may be ischemic and dysfunctional, could require a limited resection or, more likely, a tapering enteroplasty (10). End-to-end single layer anastomosis has been used in recent years to promote early recovery of bowel function and, although the use of multiple anastomosis may come with a higher risk of postoperative complications such as stricture or leaks, it appears to be the optimal procedure in terms of preserving bowel length (2,6,10,14).

Several MIA cases have been successfully managed with resection of all atretic segments, multiple anastomosis and enterostomas. The present direction when dealing with MIA is to either use multiple anastomosis or silastic stenting, a theory which is supported by newer studies where it has been reported that the flow of nutrients stimulates the motility and function of the distal intestine (2,4,6,14).

Without the use of intestinal salvage techniques, the length of the small intestine is often insufficient, while when using them, the length of viable intestine varies between 49 to 107 cm (14,15). In our case, we performed a partial resection of the atretic areas, a tapering jejunoplasty and three end-to-end anastomoses obtaining 70 cm of viable bowel, which is considered sufficient for a 33-week premature newborn (8,16).

While most studies report weaning from parenteral nutrition after 90 days, in our case, the baby only received parenteral nutrition for the first 12 days after surgery, feeding starting early (on day 4), by gradually increasing the quantity of fluids (2,6,7,14).

Conclusions

We presented the case of a newborn baby-boy, affected by the rare combination of multiple small bowel atresias type IIIb and IV, who was successfully treated by performing multiple resections and primary termino-terminal anastomosis, avoiding the need for stomas and further surgeries. Preserving the bowel length by multiple primary anatomosis in cases with multiple intestinal atresia is of utmost importance, even in the face of challenges such as the lack of good neonatal intensive care units and parenteral nutritional support which are still problems for countries like ours.

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