CASE REPORT

Under pressure: a contribution to the pathogenesis of acquired ileal atresia

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SUMMARY

An acquired ileal atresia is a rare occurrence. A 3-week-old neonate is presented, who developed postnatally a type 2 ileal atresia and an ileal stenosis within a pre-existing internal hernia secondary to an adhesion band. The literature reports a total of eight cases (4 females) with acquired ileal atresia in babies ranging in age from 3 weeks to 2 years (median 4 months). Mechanical forces (eg, adhesion band, intussusception and volvulus) onto the intestine are most frequently (75%) implicated as the primary event. The distal ileum is most often affected and a type 3A atresia is identified in six of eight (75%) cases.

BACKGROUND

We prove in this case through the operative findings and the histology that mechanical forces on the intestine lead to the development of this acquired ileal atresia.

Analysing the seven cases (table 1) reported in the literature we arrived at the conclusion that mechanical forces dominate the development of acquired ileal atresia rather than a pure vascular occlusion.

Our report challenges the prevailing assumption that in utero vascular occlusion as demonstrated in animal models is the basis for the development of intestinal atresia.

CASE PRESENTATION

Introduction

Intestinal atresia typically presents shortly after birth with the classical symptoms of abdominal distension and vomiting. Invariably the abdominal radiograph shows dilated bowel loops indicative of obstruction.^{1–3} A postnatally acquired ileal atresia is extremely rare. The case of a 3-week-old baby is

presented and the available literature analysed for pattern of development.

Case description

A baby girl was born by elective caesarean section at 38 weeks gestation with a birth weight of 2.8 kg and a normal Apgar score. After passing meconium within 24 h after birth, she fed well in the first 3 weeks and opened her bowels five to six times per day. Three days prior to her hospitalisation she had a significant decrease in her milk intake and developed absolute constipation associated with abdominal fullness.

On admission, the abdomen was distended with mild general tenderness. The anal tone was normal and there was no explosive release of bowel content as a possible indication of Hirschsprung disease. The abdominal radiograph showed grossly dilated bowel loops and a paucity of rectal gas. Contrast meal radiograph identified a high-grade bowel obstruction with normal gut rotation.

At laparotomy the following findings were present: part of the distal ileum was found to be adherent to the caecum and ascending colon region and an adhesion band originating from the ascending colon stretched to the mesentery of the distal ileum harbouring an internal hernia (figure 1). The loop of the herniated bowel was in close proximity to an ileal atresia 2 cm from the ileocaecal valve (ICV; figure 2). A second stenotic segment was identified in the distal part of the loop, approximately 35 cm from the ICV (figure 3). The adhesions were released, the atretic and stenotic segments were resected and gut continuity restored. Microscopy of the stenotic segment illustrated mucosal erosion (figure 3B, arrow) and serosal fibrosis (figure 3B, double arrow).

The atretic segment showed on microscopy two blind pouches connected by the fibrotic tissue, in

Table 1 Acquired intestinal atresia

Author	Age/sex	Findings	Location	Type of atresia	Follow-up
Michaelis (1971)	19 months/M	Abdominal abscess 2° to strangulation	Mid-ileum	Type 3A	? months NED
Drablos 1978	7 weeks/M	Septic thrombosis ?	Ileocecal region	Type 3A	12 months NED
López Candel 1993	10 weeks/F	Iatrogenic postvolvulus	Dist ileum	Type 3A	postmortem
Ratan 2005	4 months/M	Intussusception?	Dist ileum	Type 2	6 months NED
	3 months/M	Volvulus?	Dist ileum	Type 3A	? months NED
Elemen 2010	2 years/F	Adhesion band	Dist ileum	Type 3A	18 months NED
Osuoji 2011	4 months/F	Intussusception	Dist ileum	Type 3A	? months NED
Houben 2013	3 weeks/F	Adhesion band	Dist ileum	Type 2	3 months NED

Age, age at operation; F, female; M, male; NED, no evidence of disease.

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Figure 1 Graphic illustration of the internal hernia in the distal ileum region with the stenotic segment around 35 cm from the ileocaecal valve (ICV) and the atresia close to ICV.

line with a typical type 2 ileal atresia (figure 2A,B arrows). The mucosa of the proximal pouch shows an inflammatory response. No thromboembolic events were identified.

The postoperative recovery was uneventful: extubation took place 4 days later, oral nutrition was restarted at 6 days and the baby was discharged 11 days after surgery.

OUTCOME AND FOLLOW-UP

At the 3-month follow-up the baby is thriving.

DISCUSSION

In our present description, the clinical scenario showed an adherence of the distal ileum to the caecum and the ascending colon region plus an adhesion band stretching from the ascending colon to the mesentery of the distal ileum resulting in an internal hernia (figure 1). It is not clear whether the entrapment of a segment of the ileum within the adhesion is a prenatal or postnatal event. We also do not have any clues to the exact time when the adhesion developed. However, the baby had no illness (including bouts of fever, etc) after birth. She thrived, fed well and had regular bowel openings within the first 3 weeks of life, suggesting the absence of significant bowel obstruction during this period of her life. In any case, the initially undisturbed functioning gastrointestinal tract may have caused a pressure effect onto the more distal gut in

close proximity to the herniated segment of the bowel. The findings of mucosal erosions and serosal fibrosis in this segment of the ileum suggest a chronic pressure effect on the vasculature, rather than an abrupt interruption of blood supply (figure 3B). Acute insult to the vascular supply would cause more dramatic clinical symptoms and signs. Histologically, the pathology would be dominated by mucosal damage, extending to the submucosa as oedematous changes and further changes might develop in the muscularis propria, resulting in damage to the smooth muscle cells. Such acute changes were not present at the stenotic segment of our patient (figure 3B).

The more distal segment close to the ICV may have encountered similar pressure effects and continued to obliterate completely at around 3 weeks postpartum. Microscopic examination identified two blind pouches connected by fibrotic tissue, such as a type 2 atresia of the ileum (figure 2A,B arrows). The mucosa of the proximal pouch was inflamed due to the pressure effect of the obstruction. The aetiology of this atretic segment might be the same as that of the proximal stenotic portion. Apparently, this more distal portion of the ileum at the hernia sac might be subjected to stronger external pressure. Thus, this distal portion underwent atresia earlier. Once the atresia was formed, the proximal bowel segment distended with food contents. These microscopic findings illustrate the sequence of events leading to the postnatal development of an ileal stenosis and an ileal atresia, most likely as a result of mechanical forces within an internal hernia entrapped by an adhesion.

Including the aforementioned case, a total of eight cases (4 females) with acquired ileal atresia were identified (table 1).^{4–9} The patients' ages ranged from 3 weeks to 2 years (median 4 months). The distal ileum is most often affected in 7/8 (87%) cases and the most frequent finding was a type 3A ileal atresia 6/8 (75%). The origin of the atresia was identified in five cases: there were two with an adhesion band, one intussusception, one case of an abdominal abscess (most likely secondary to gut strangulation) and an infant with a postvolvulus bowel anastomosis which resulted in a type 3A ileal atresia 7 weeks following surgery.⁶ In two more cases, circumstantial evidence points to an intussusception and a volvulus as the initiating factors in the development of the acquired ileal atresia.⁸ Only in one case, a septic thrombosis may have resulted in a type 3A atresia.⁵ Mechanical forces could be identified in three quarters of the cases affected by an acquired atresia.

Pressure effects onto the distal ileum appear to be domineering in the development of ileal atresia (table 1). The aetiology may be related to the complex rotational events between gestational week four or five to the completion of week 12: the

Figure 2 Surgical specimen of an atretic segment (A) close to the ileocaecal valve with two blind pouches connected by fibrous tissue—(B, H&E, magnification $\times 5$ and arrow)—consistent with a type 2 intestinal atresia.

Figure 3 Surgical specimen (A) of the ileum showing the stenotic region of the ileum. Erosion is seen at the mucosa of the stenotic region (B, H&E staining, magnification $\times 10$ and arrowhead). There is only fibrosis at the serosa (B, H&E staining and double arrowheads) suggesting chronic pressure effect.

intestinal loop of the midgut elongates and rotates 90° counter-clockwise into the umbilical herniation, which is followed by a further counter-clockwise rotation of approximately 180° during the return of the gut into the abdominal cavity.^{10–12} Possible rotational and mesenteric anomalies may result in various defects instigating the development of an intestinal atresia.^{13–14} More than 100 years ago Spriggs identified adhesion band, volvulus, intussusception and snaring of a loop of bowel at the navel as mechanisms for the development of intra-uterine intestinal atresia.¹⁵ Postnatally such mechanisms may continue to play a role in the development of acquired ileal atresia as illustrated in table 1. Although various animal models provide evidence for the development of an atresia as a result of a primary vascular insult through ligation of a branch of the mesenteric vessel, it appears difficult to translate such results into a clinical scenario based on a thromboembolic process.^{16–18} After all, the intestine is surrounded by a rich network of mesenteric arterial anastomosis (Riolan arcades). In fact only one patient had possible evidence of a thrombotic event as a result of a sepsis.⁵

CONCLUSION

We described a 3-week-old neonate with acquired ileal stenosis and ileal atresia. Available reports imply that mechanical forces on the intestine (eg, adhesion band, volvulus and intussusception) dominate the pathogenesis of acquired ileal atresia, and this type of atresia is most frequently seen in the region of the distal ileum. Typically, type 3A atresia is more commonly identified. There is little evidence to support a pure vascular occlusion (eg, thromboembolism) as demonstrated in animal models, as the basis for the development of an atresia.

Learning points

- Acquired ileal atresia is an exceedingly rare condition caused by mechanical forces pressurising the intestine.
- Acquired ileal atresia is typically identified in the distal ileal region; a type 3A atresia is seen most frequently.
- There is little evidence to support a pure vascular occlusion—propagated by animal models—as the basis for the development of an atresia.

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