

## ILEO-CECAL ATRESIA SECONDARY TO INTRAUTERINE INTUSSUSCEPTION

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Intestinal atresia usually occurs on the jejunum or ileum as a result of suspected mesenteric vascular occlusion. Atresia of the ileocecal segment is very rare and little is known about its origin. We report a very rare case of ileocecal atresia where pathological examination revealed intrauterine intussusception. Resection of the atretic dilated segment and end to oblique anastomosis was undertaken without valve reconstruction.

**Key words:** Intrauterine intussusception ■ Atresia of cecum

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**Received:** May 19, 2012  
**Accepted:** July 16, 2012

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### Introduction

Intestinal atresia is a relatively common malformation, with an incidence of 1:6-8000 live births (1). Although about one third of intestinal atresias are localized in the distal ileum, atresia of the ileocecal junction and atresia of the ileocecal valve are rare. Only four cases have been reported in the English-language literature (2, 3, 4).

There are various theories about aetiology of intestinal atresia. A developmental anomaly in early fetal life, failure of recanalisation, is blamed for atresia affecting the duodenum. Similarly, the number of associated anomalies in proximal jejunal atresia is higher than in distal types. The occlusion of the mesenteric vessels has been hypothesized in the aetiology of distal jejunal and ileal atresia, especially in types II-IV, where a “V” shaped mesenteric defect is present. Intrauterine intestinal perforation, volvulus, malrotation, intussusception, meconium ileus, meconium peritonitis, segmental intestinal thrombosis, smoking, drugs causing vascular contraction, or cocaine may be in the

background (5). Little is known about the aetiology of ileocecal atresia.

Intrauterine intussusception has been described as a rare cause of intestinal atresia. Evans, in a review of almost 1500 cases of intestinal atresia, found only 9 (0.6%) cases due to intrauterine intussusception, but the exact location was not specified (6).

We report a case of ileocecal atresia where pathological examination revealed intrauterine intussusception.

### Case report

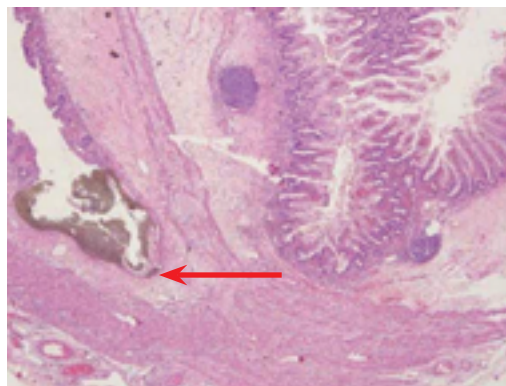
A preterm male neonate (36 weeks gestation), was born by normal vaginal delivery, weighing 1950 g, and the first minute Apgar score was 7/9. The patient was referred with failure to pass meconium, bile stained vomiting and abdominal distension. The abdominal x-ray showed significant dilatation of the small bowel loops, a gasless abdomen and gastrografin enema showed micro colon (Fig. 1).



**Fig. 1** X-ray after contrast enema shows distended small bowel loops, gasless abdomen and microcolon, the contrast dye did not pass to the distended loops.

Laparotomy revealed a distended terminal ileum, and no appendix. No defect on the mesentery was identified. The length of small bowel was found normal, no adhesions and no signs of malrotation were seen.

The dilated terminal ileum, together with the atretic segment was resected and an ileocolic anastomosis, without valve reconstruction, was performed. Pathological examination revealed two sets of bowel wall within the lumen covered by ileal and colonic mucosa, with telescopic protrusion and signs of ischemic necrosis of the intussuscepted segment. All these findings suggested ileocecal atresia secondary on the intrauterine intussusceptions (Fig. 2).



**Fig. 2** Microscopically, two sets of bowel wall, covered with ileal and caecal mucosa and signs of ischemic necrosis of the intussuscepted segment (arrow).

### Discussion

The aetiology of atresia of the intestine differs slightly in different segments. Early fetal disturbance is accountable for duodenum and proximal jejunal atresia and mesenteric vascular occlusion for distal jejunal and ileal atresia (7). Intrauterine intussusception may be responsible for atresia of the ileocecal junction and atresia of the ileocecal valve. We present a unique case, supported by histological evidence, that ileocecal intussusception in utero resulted in ileo-caecal atresia.

It is well known that postnatal intussusception occurs most frequently at the ileocaecal junction and is usually associated with gastrointestinal and respiratory infections. This is explained by the lipopolysachari-

de (LPS) model of intussusception (excessive nitric oxide release results in pathological bowel relaxation and intussusception), and the relative nitrergic hyperinnervation of the ileocecal valve found by Cserni et al. (8, 9). Although little is known about the aetiology of in utero intussusception it is easy to hypothesise a similar mechanism ie. infection, meconium peritonitis.

A definitive diagnosis of intrauterine intussusception is usually made on histopathological examination. A histological study classifies intrauterine intussusception in types A and B. In type A, the intussuscepted bowel would be located at the distal blind end with good preservation of bowel structure, probably owing to a slight amount of blood supply from the mesentery. In type B the intussuscepted bowel is found outside the distal blind end, and it is necrosed and almost reabsorbed (10). The importance of the ileocecal atresia lies in the absence of the ileocaecal valve.

Most surgeons think that the loss of the valve is well tolerated, however Folaramni et al. recently reported that the ileocaecal valve has significant importance in children (11). The new concept of the ileocaecal valve supported by extensive neuro-histology study suggests that the ileocaecal valve does

not resemble any sphincter, and simple end-to-side intussuscepted ileo-colic anastomosis may replace the valve function (12, 13).

In our case valve replacement was not an option during the first operation, because the microcolon would not allow us to perform an intussuscepted ileo-cecal anastomosis. We may consider ileocecal valve reconstruction if the patient develops chronic diarrhoea in the future.

We report a very rare case of ileocecal atresia secondary to intrauterine intussusceptions, which corresponds histologically with type A, confirmed by pathology examination. When intussusception occurs in the foetus and the time elapsed is enough to allow for gangrene and resorption of the intussuscepted portions of the bowel, intestinal atresia may result. This case provides further evidence for an understanding of the pathogenesis of ileocecal atresia.

**Authors' contributions:** Conception and design: EH; Acquisition, analysis and interpretation of data: NH, AH; Drafting the Manuscript: EH, EK; Critical revision for important intellectual content: ZK.

**Conflict of interest:** The authors declare that they have no conflict of interest. This article was not sponsored by any external organisation.

## References

1. Saxena AK, van Tuil C. Intrauterine Intussusception in etiology of jejunal atresia. *Dig Surg*. 2008;25:187.
2. Ein SH, Venugopal S, Mancor K. Ileocaecal atresia. *J Pediatr Surg*. 1985;20:525-8.
3. Grassi R, Pinto A, Valente T, Rossi G, Catalano O, Rotondo A, et al. Massive enterolithiasis associated with ileal dysgenesis. *Br J Radiol*. 1997; 70:207-29.
4. Cserni T, Magyar A, Nemeth T, Paran TS, Csizy I, Jozsa T. Atresia of the ileocecal junction with agenesis of the ileocecal valve and vermiform appendix: Report of a case. *Surg Today*. 2006;36:1126-8.
5. Aguayo P, Ostlie D. Duodenal and intestinal atresia and stenosis. In: Holcomb G, Murphy P, editors. *Ashcraft's Pediatric Surgery*. Philadelphia; Elsevier; 2010. p. 400-15.
6. Evans CH. Atresias of the gastrointestinal tract. *Int Abstr Surg*. 1951;92:1-8.
7. Strauch ED, Hill L. Intestinal atresia. In: Ziegler M, Azizkhan R, Weber T, editors. *Operative Pediatric Surgery*. New York: McGraw-Hill Comp; 2003. p. 589-97.
8. Cserni T, Paran S, Puri P, Srivastava P. New hypothesis on the pathogenesis of ileocecal intussusceptions. *J Pediatr Surg*. 2007;42:1515-9.

9. Nissan A, Zhang JM, Lin Z, Haskel Y, Freund HR, Hanani M. The contribution of inflammatory mediators and nitric oxide to lipopolysaccharide-induced intussusception in mice. *J of Surg Research*. 1997;69:205-7.
10. Huang WC, Wang CH, Yuh YS. A rare type of ileal atresia due to intrauterine intussusception. *Eur J Pediatr*. 2007;166:1177-8.
11. Carlos P, Joaquin M, Yolanda R, Robert S. Intrauterine intussusception: a rare cause of intestinal atresia. *J Pediatr Surg*. 2009;44:2028-30.
12. Folaranmi S, Rakoczy G, Bruce J, Humphrey G, Bowen J, Morabito A, et al. Ileocecal valve: how important is it? *Pediatr Surg Int*. 2011;7:613-5.
13. Cserni T, Paran S, Kanyari Z, O'Donnell AM, Kutasy B, Nemeth N, et al. New insights into the neuromuscular anatomy of the ileocecal valve. *Anat Rec*. 2009;292:254-61.

**Citate:** Husaric E, Hotic H, Halilbasic A, Konjic E, Karasalihovic Z. Ileo-cecal atresia secondary to intrauterine intussusception. *Paediatrics Today*. 2012;8(2):147-150.