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ILEAL ATRESIA PRODUCED BY INTUSSUSCEPTION IN UTERO

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Abstract

We experienced 3 cases of ileal atresia, which was considered to be produced by intussusception in the late stage of pregnancy. We attributed ileal atresia to intussusception in utero because of the following findings;
(1) the shortage of the whole length of the small intestine, (2) the normal meconium in the large intestine, (3) defect of the mesentery, (4) no associated malformation, and (5) histologically unique polypoid prominence in the distal blind end.

Histological studies of our 3 cases lead us to the hypothesis about the pathologic process of spontaneous recovery of intussusception in utero. According to this hypothesis, we presumed that the probability of the natural cure is greater than that of the atresia.

INTRODUCTION

Intestinal atresia has been considered to occur due to arrested development of the intestine in the solid stage during the second and third month of gestation. At present, this theory seems to offer a satisfactory explanation of duodenal atresia. However, Louw and Barnard[1] suggested that "vascular accidents" in the fetal intestine might cause some of jejunoileal atresia. Experimentally they succeeded in producing intestinal atresia after ligation of the mesentery of puppies in utero. In 1888, Chiari[2] reported the first case of intestinal atresia produced by intussusception in utero. Recently, we experienced 3 cases of ileal atresia and found that all cases had the polypoid prominence in the distal blind end of the bowel, which corresponded to the intussusceptum according to the histological examination. We investigated the pathogenesis of ileal atresia from such observations.

CASES

Case 1, a female infant was a product of a 23-year old mother with an uncomplicated pregnancy. Polyhydramnios was not observed. She was transferred to our clinic at the 5th day of life because of vomiting bile-stained material and abdominal distension. On admission, the abdomen was markedly distended. The peristaltic waves were visible and the bowel sounds were hyperactive. Dehydration and electrolyte imbalance were noted. The plain films of the abdomen demonstrated gas and fluid levels in the distended stomach and in some bowel loops. No gas was noted in the colon and rectum. A Gastrografin enema was easily passed through the slightly narrow but otherwise apparently normal colon and the cecum, which was normally situated. After the enema, a small amount of normal meconium was passed. A diagnosis of ileal atresia was made and the body fluid and electrolytes abnormalities were corrected. The operation was performed on the next day.

Examination of the abdominal viscera revealed the dilated jejunum and ileum, the latter ending in a bulbous point which was completely sealed off. The distal 30 cm of the ileum was collapsed as was the colon. Both blind ends were not connected by a cordage and the ileal mesentery in this atretic area presented a U-shaped defect. The proximal 20 cm and the distal 5 cm of ileum were resected and an end-to-oblique anastomosis was performed. The postoperative course was uneventful.

The macroscopic observation of the resected specimen showed the proximal segment of the bowel to be considerably dilated and the wall was thick. A polypoid prominence with a length of 0.3 cm and a diameter of 0.1 cm was seen projecting into its lumen at 1 cm distal to the tip of the distal segment (Fig. 1).

The pathological examination of the proximal segment revealed that the mucosa was edematous and the muscularis propria and the mucosal muscle-plate were thick. The Auerbach's plexus was normal. The polypoid prominence in the distal segment consisted of normal intestinal wall and was covered with the mucosa, which disappeared toward the tip. Furthermore, the calcification was observed in the prominence. The muscularis propria of the small intestine proceeded into the polypoid prominence so that the intestinal wall at the attachment of the prominence had no muscularis propria (Fig. 2).

Case 2, a male infant was a product of a 27-year old mother. Polyhydramnios was not observed. Eight hours after birth, cyanosis in his limbs was observed and the abdominal distension was noted. He was

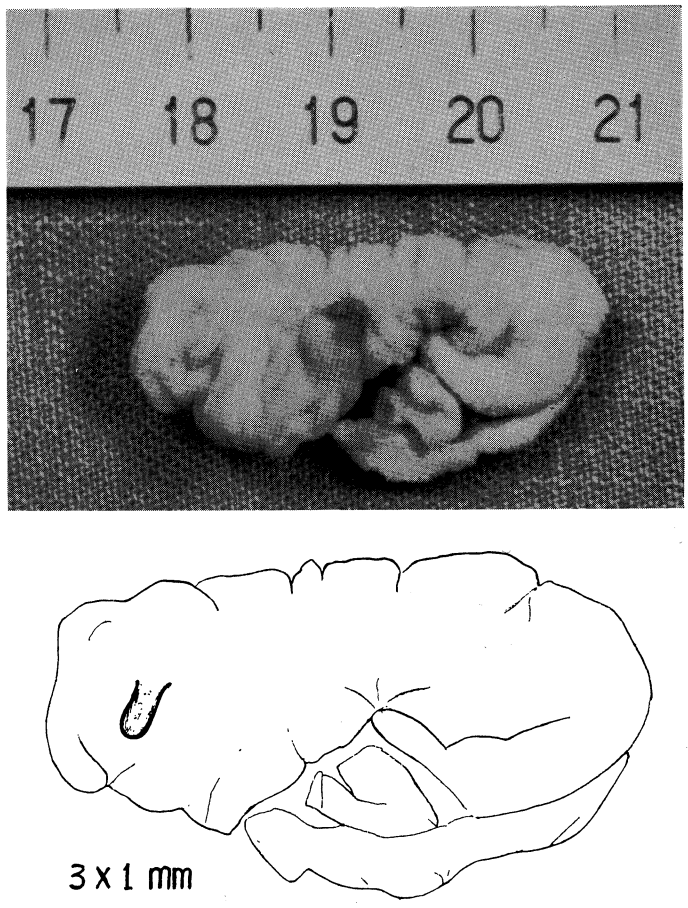

Fig. 1. The mucosal surface of the distal segment after formalin fixation in the case 1. The lower is a sketch of the segment.

transferred to our clinic at the 2nd day of life. The abdomen was considerably distended. The lung-liver border was not clear by percussion and the bowel sounds were inaudible. A small amount of normal meconium was passed by the enema. He had dehydration, electrolyte imbalance and metabolic acidosis. The plain films of the abdomen showed gas and fluid levels in the stomach and some bowel loops. In addition, some free fluid and free gas were noted in the peritoneal cavity. The calcification was not found. With a diagnosis of perforation of the stomach or the small intestine, the operation was performed 27 hours after birth.

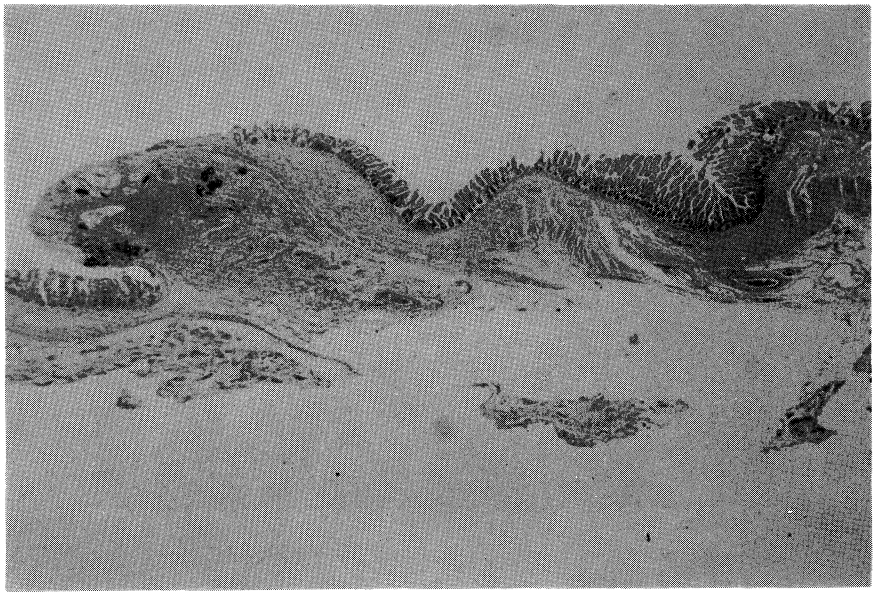

Fig. 2. Photomicrograph of the polypoid prominence in the case 1. (×13, H-E stain)

There were about 100 ml of the yellowish brown ascites and the meconium in the peritoneal cavity. The small intestine ended in a bulbous point which was completely sealed off at 100 cm from Treitz's ligament, and the perforation was observed at the tip. Both blind ends were completely separated and the mesentery had a U-shaped defect. An atretic segment was observed 10 cm proximal to the ileocecal valve. The ileum 20 cm proximal and 5 cm distal to the atretic segment were resected and an end-to-oblique anastomosis was performed. After the operation, the patient expired because of the respiratory distress.

The resected specimen was observed. An intraluminal bulb was palpated in the blind end of the distal segment. The mucosa on the mesentery side within the range of 2.5 cm from the blind end was somewhat hypertrophied and proceeded further to the polypoid prominence with a length of 1.5 cm and a diameter of 0.3 cm, at the tip of which the brown, necrotic substance (2×0.5 cm) was observed (Fig. 3).

The pathological examination of the proximal segment revealed that normal ganglion cells were present. The base of the polypoid prominence in the distal segment was covered with the atrophied mucosa of the small intestine, and there were scattered calcified areas. The pro-

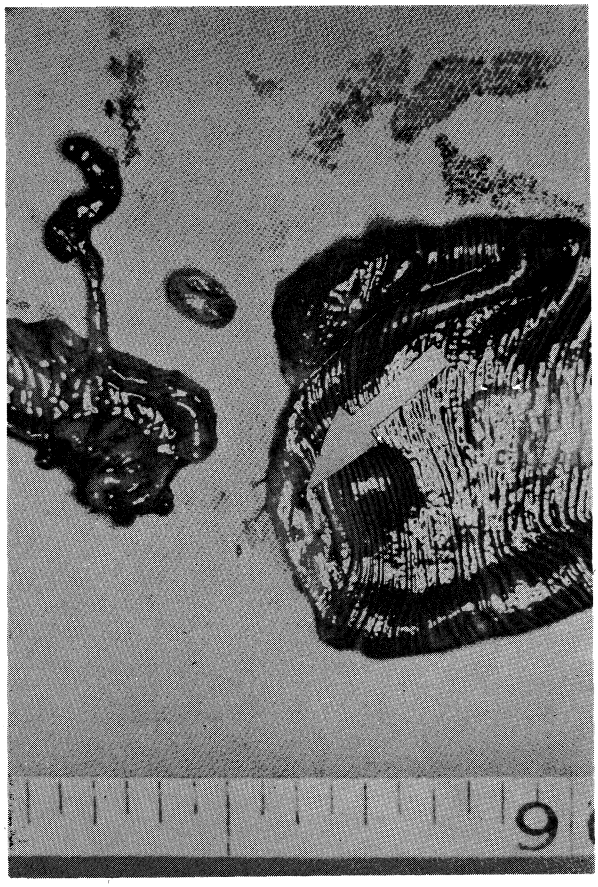

Fig. 3. Resected specimens of the case 3; the left is the distal segment and the right is the proximal segment. The arrow shows the perforation site.

minence consisted of the intestinal wall with the mucosal muscle-plate and muscularis propria (Fig. 4 and 5).

The mucosa of the distal segment was extending to the prominence. The muscularis propria was observed in the prominence, so that the intestinal wall at the attachment of the prominence had no muscularis propria (Fig. 6). The tip of the prominence had the hemorrhagic necrosis, which retained the structure of the intestinal wall as a whole.

Case 3, a male infant was a product of a 25-year old mother. There were about 130 ml of the amniotic fluid. He became gradually cyanotic and vomited greenish material. Thirteen hours after birth, the normal meconium was passed after the enema. Since then, the abdominal

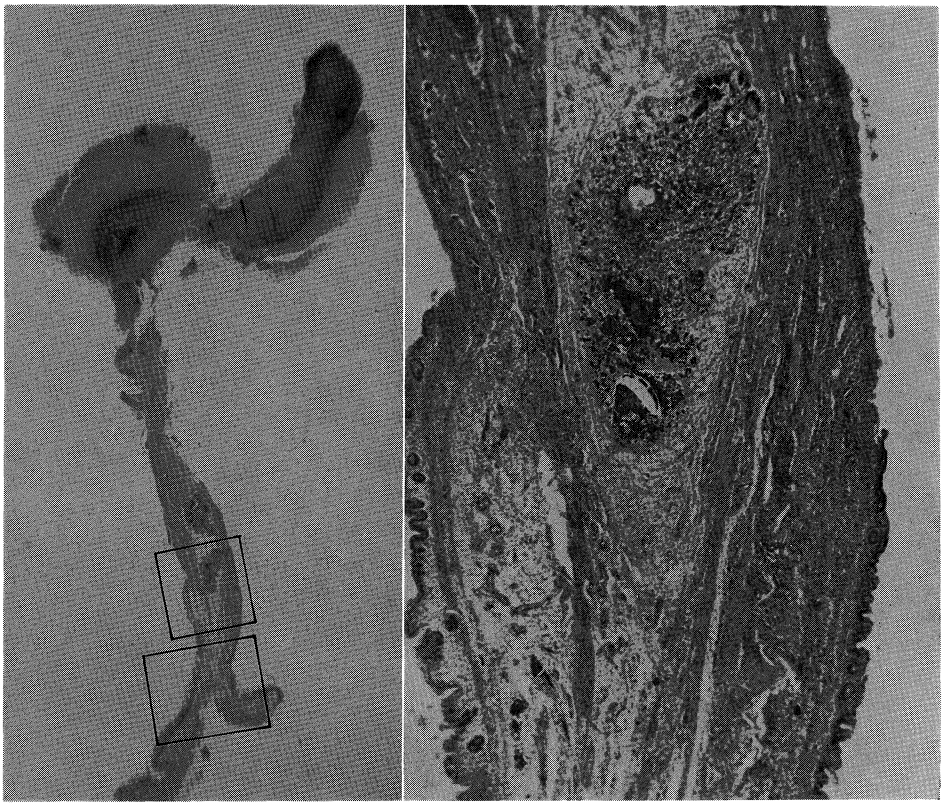

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Fig. 4. Photomicrograph of the polypoid prominence in the case 2. (×3, H-E stain)
Fig. 5. The magnified view of the upper frame in Fig. 4. (×30, H-E stain)

distension and cyanosis were noted in the whole body. Cyanosis was prominent in his face and limbs at the 2nd day of life. The lung-liver border was not clear by percussion and the bowel sounds were inaudible, and the abdomen was considerably distended. The plain films of the abdomen showed free fluid and gas in the peritoneal cavity. The gas and fluid levels were present in the stomach and the small intestine, but not in the colon, nor in the rectum. The calcification was not found. A diagnosis of perforative peritonitis due to ileal atresia was made and the operation was performed at 28 hours after birth.

The abdomen contained about 100 ml of the greenish black fluid. At a distance of 85 cm from the Treitz's ligament, the small intestine ended in a bulbous point, which was completely sealed off. At the tip

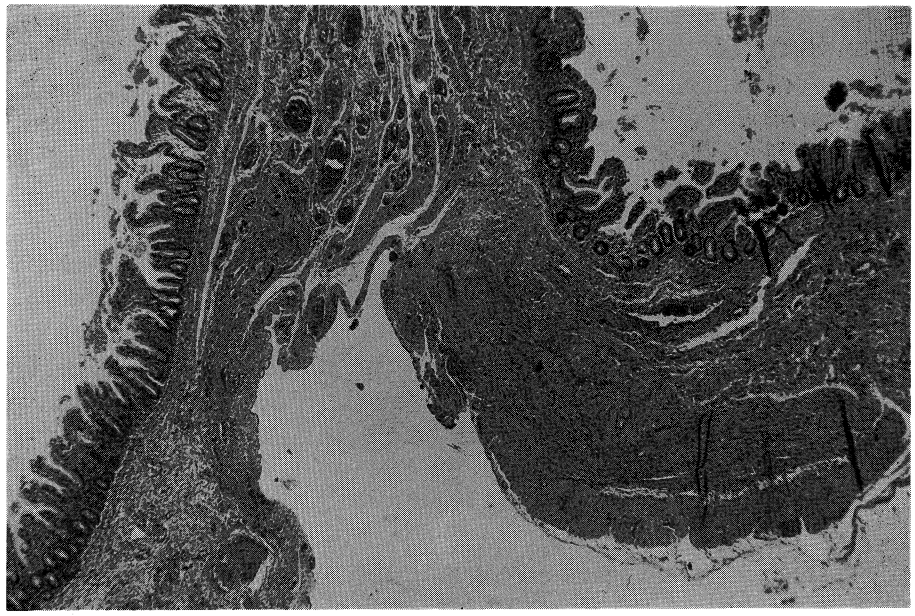

Fig. 6. The magnified view of the lower frame in Fig. 4. (×35, H-E stain)

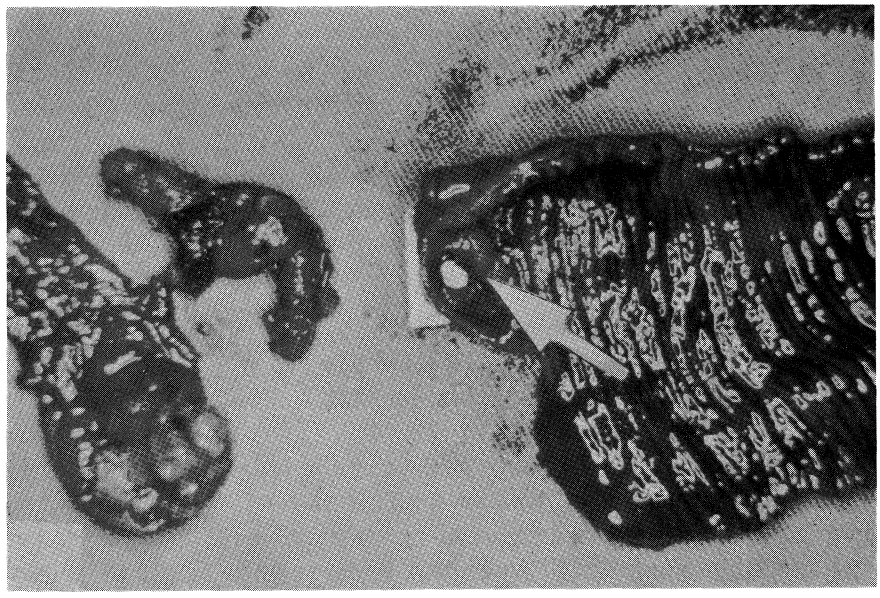

Fig. 7. Resected specimens of the case 3; the left is the distal segment and the right is the proximal segment. The arrow shows the perforation site.

of the bulbous point perforation was noted. Both blind ends were completely separated. The mesentery in this area had a U-shaped defect. An intraluminal bulb was palpated in the blind end of the distal segment, which was 20 cm apart from the ileocecal valve. The ileum 25 cm proximal and 5 cm distal to the atretic segment were resected and an end-to-oblique anastomosis was performed. The postoperative course was uneventful.

The resected specimen was observed. The mucosa on the mesentery side of the distal bowel was somewhat hypertrophied, and proceeded to the polypoid prominence of 1.0 cm length which was 2 cm apart from the blind end. At the tip of the prominence the reddish brown, necrotic mass with a length of 4 cm and a diameter of 0.5-1.0 cm was observed (Fig. 7). The microscopic findings were the same as in the case 2.

DISCUSSION

I. Classification

The term atresia means complete intrinsic occlusion of the intestinal wall due to anomalous development of its walls, whereas the term stenosis refers to incomplete occlusion. It is considered that both may be produced by the same cause. Louw[3] classifies them into 4-types as follows:

1. Stenosis or incomplete occlusion;
2. Atresia type 1 or a diaphragm (membrane);
3. Atresia type 2 - blind ends joined together with or without a corresponding gap in the mesentery; and
4. Atresia type 3 or disconnected blind ends with a gap in the mesentery and often considerable deficiency in the length of the small bowel.

All our cases belong to atresia type 3.

II. Pathogenesis of jejunoileal atresia

In 1900, Tandler demonstrated that the human duodenum passes through a solid stage during embryonic life. This is due to epithelial proliferation which commences in the fifth week and soon obliterates the whole lumen of the bowel. The lumen reforms by vacuoles which coalesce and is completely reestablished at the end of the eighth week. Tandler suggested that an arrest of the development of the duodenum during the solid stage (fifth to eighth week) would result in intestinal atresia. His hypothesis was widely accepted in the world. However, Evans[4] considered that an atresia of the intestine was caused by a

mechanical disturbance which produced an abnormal bowel wall. At the beginning of the 19th century, intestinal atresia which was perhaps produced by interruption of the blood supply to a segment of the sterile fetal bowel, was reported in succession, but at that time the appreciation was hardly obtained. In 1955, Louw et al. explained on basis of fetal dog experiments that interruption of the blood supply to a segment of the sterile fetal bowel was a cause of intestinal atresia. Thereafter, Santulli et al.[5] supported the Louw's theory of vascular insufficiency because the following facts could not be explained by the recanalization theory; 1) short length of the small intestine, 2) the normal meconium is found distal to the point of the atresia, 3) the gap is found in the mesentery of the atretic area. Furthermore, Grosfeld et al.[6] reported that intestinal atresia having the polypoid prominence in the blind end of the distal segment was caused by intussusception in the late fetal life, and pointed out the following facts as evidence;

1) mature baby,
2) the presence of bile, squamous epithelium and lanugo hair in the distal bowel,
3) no associated anomalies in the other organ systems,
4) most in the ileum,
5) necrotic substances are passed per rectum,
6) normal caliber of the colon, and
7) the polypoid prominence is present in the distal blind end.

The length of the small intestine of our cases were 130, 140 and 110 cm; these were shorter than normal small intestines (248 cm) as Benson[7] had described. All our cases were matured, passed the normal meconium, and had ileal atresia with no other associated malformation. In addition, necrotic substances were not passed per rectum in all cases but they adhered to the tip of the polypoid prominence in the case 2 and 3. Microcolon was not observed in the case 1. We understood that these cases were ileal atresia due to intussusception in the late stage of the pregnancy because of the presence of the polypoid prominence in the distal bowel.

III. Histological investigation of the polypoid prominence in the distal bowel

Intestinal atresia due to intussusception was reported one after another since Chiari. In Japan, Ueda et al.[8] reported a case of intestinal atresia with the necrotic remains of the intussusceptum in the distal blind end. Gherardi et al.[9] reported a case, in which the polypoid pro-

minence was present in the distal bowel, covered by the mucosa, and had the calcification and muscle layers. He pointed out that it should be a remnant of the intussusceptum. Zachary[10] observed a layer of the mucosal epithelial cells calcified in the polypoid prominence as a remnant of the intussusceptum. The calcification in our cases seemed to be only inflammatory products due to the fact that it was present also in the submucosa and in the Auerbach's plexus. Pathological observation of our 3 cases can be summarized as follows;

1) the muscularis propria proceeds into the polypoid prominence, so that the intestinal wall at the attachment of the prominence has no muscularis propria,
2) the muscularis propria proceeds into the prominence to the necrotic part of the tip,
3) the intestinal mucosa covers the base of the prominence and disappears toward the tip,
4) the calcification is found, which is considered as inflammatory products,
5) the tip of the prominence has hemorrhagic necrosis, and
6) the normal Auerbach's plexus is present in both the proximal and distal segment.

IV. The pathologic process of spontaneous recovery of intussusception in utero

Intussusception is found most frequently in males at the age of 3 to 18 months. In 1955, Rachelson et al.[11] reported a case of ileal atresia produced by intussusception in newborn. In a review of large series in the literature, they have been able to find that 42 per cent of the 593 non-reposition cases of intussusception is cured by spontaneous auto-anastomosis. It is understood that the frequency of the natural cure may be much higher in the germ-free state as in utero. Histological analysis of our 3 cases lead us to the hypothesis about the pathologic process of spontaneous recovery of intussusception in utero (Fig. 8); intussusception in utero gives rise to the interruption of the blood supply to a segment of the sterile fetal bowel and finally necrosis of the intestine. If a separation takes place at A, peritonitis is produced and both sides of the tube are closed because of germ-free state. The separated intussusceptum at A will be straightened out into the distal lumen and become the polypoid prominence and its tip will develop hemorrhagic necrosis. Therefore, the polypoid prominence is covered by the mucosa and the muscularis propria is characteristically present.

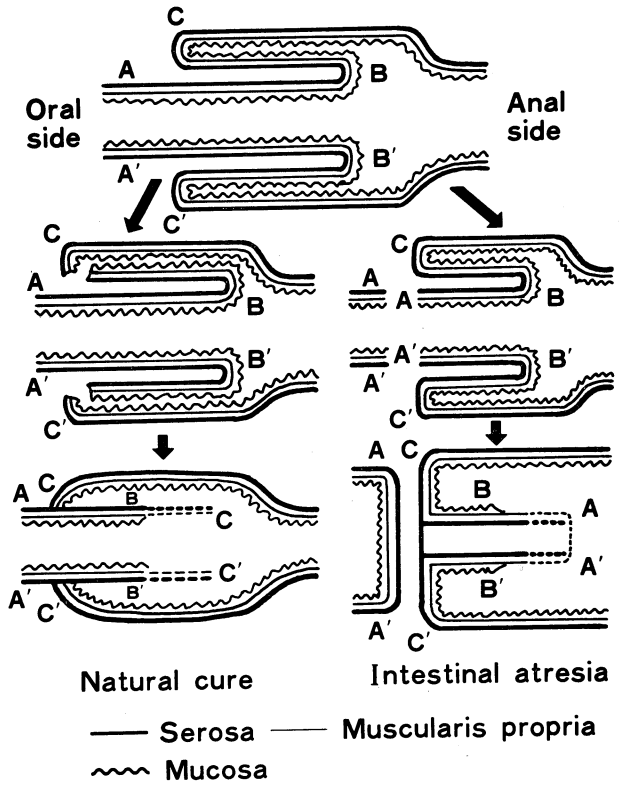

Fig. 8. A schema of the pathologic process of spontaneous recovery of intussusception in utero.

Since the case 1 suffered from intussusception earlier than the case 2 and 3, the necrotic part was absent and the prominence was atrophic. On the other hand, if a separation takes place at C, the intestine which is reversed to the distal side, becomes bicapsular; therefore, the contents of the intestine do not leak into the peritoneal cavity, and intussusception is cured by auto-anastomosis. If the separation takes place between C and B, or close to B between B and A, auto-anastomosis takes place in the same way. As described above, when intussusception is produced in utero, the probability of the natural cure is assumed to be higher than that of the atresia.

V. Frequency of intussusception in utero

According to Louw's publication, the incidence of intestinal atresia is 1 per 3,000 births. Fifty-two per cent of all intestinal atresia is

jejunoileal atresia, about 5 per cent of which is produced by intussusception. It is considerable that intussusception may take place in utero at a fairly high frequency, since the small polypoid prominence is often overlooked and the probability of the natural cure is relatively high.

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