

Case Report

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Right retrocolic intrasaccular duodenum: Is it a novel variant of isolated duodenal nonrotation? A case report

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KEYWORDS

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ABSTRACT

Background: Almost all of the studies on anomalies of the midgut rotation and fixation in the literature and related sections in textbooks were designed according to Dott's classification. Focusing only on common rotation anomalies has led to the exclusion and neglect of other rare variants. Isolated pure duodenal nonrotation is such a variant.

Case Presentation: We report a case of an unusual form of isolated pure duodenal nonrotation, in a 3-day-old newborn presenting with bilious vomiting. Ultrasonographic examination revealed the sign of 'whirlpool'. When this finding was evaluated together with bilious vomiting, midgut volvulus was considered and the patient was operated on urgently. Peroperatively, it was observed that the jejunum entered between the leaves of the terminal ileum mesentery. Proximally, the duodenum was located posterior to the right colon in a "sack". This "sack" was surrounded by thick Ladd's bands laterally, mesentery of the ascending colon medially, the posterior surface of the cecum and ascending colon anteriorly, and by the posterior abdominal wall posteriorly.

Conclusion: In isolated duodenal nonrotation, the duodenum may be completely retro-colic. Consequently, the duodenojejunal junction and the ileocecal region may almost overlap. Unlike isolated duodenal nonrotation cases, in the surgical treatment of this variant, separation of Ladd bands alone is not sufficient, additionally, the right colon should be placed in a nonrotation position and care should be taken not to kink the terminal ileum under the cecum.

INTRODUCTION

Malrotation is not a single entity; this term generally applies to any type of abnormalities of the intestinal rotation. [1-4] Frazer and Robbins first described the classic three stages of rotation and fixation process, and Dott translated these preliminary embryologic observations into problems encountered clinically and classified the rotation anomalies depending on which classical stage of rotation these anomalies occur. [5,6] Almost all of the studies on anomalies of the midgut rotation in the literature and related chapters in textbooks are designed according to these commonly known types of rotation anomalies. On the other hand, focusing only on common rotation anomalies has led to the exclusion and neglect of other rare subtypes. Isolated duodenal nonrotation (IDN) is such a subtype. IDN is the type of malrotation with arrested duodenal rotation and normal colonic rotation. Since it is often associated with Ladd bands constricting the

right-sided duodenum, patients generally present with chronic and sometimes acute duodenal obstruction symptoms (chronic abdominal pain, bilious vomiting). Since the symptoms are not specific to IDN, it is almost impossible to diagnose the disease clinically. Radiologically, the upper gastrointestinal contrast study is helpful in the diagnosis of IDN; the position of the duodenojejunal junction is to the right of the spine and the cecum is in the normal position. [7]

Herein, we present a case of an unusual form of isolated pure duodenal nonrotation. The isolated duodenal nonrotation in the current case was defined according to Stringer's classification (Stringer's Type 2A malrotation). [7].

CASE REPORT

A 3-day-old male neonate presented with bilious vomiting for a day. He was born at 38+5 weeks of gestation in March 2020 with a birth weight of 3140g.

Prenatal follow-up was uneventful. The parents were related (cousins) and there was no previous history of intrauterine fetal death. The patient was dehydrated and refusing to feed thus admitted to the neonatal ICU. Bowel habits were reported as normal. Abdominal findings were normal (no abdominal distention or tenderness). The complete blood count and serum biochemical test were normal. Blood gas revealed a lactate value of 4.5 mmol/L (normal values 0.5-1.6 mmol/L). A plain abdominal x-ray was normal. There was a whirlpool sign on abdominal ultrasonography.

Considering the volvulus of the small bowel, urgent laparotomy was done but the volvulus was found self detorted. Other findings were chylous peritoneal fluid, lymphatic stasis in the mesentery, and edema of the small intestine. The anatomy of the colon and small intestine seemed normal at first glance. However, there was a tortuous venous vessel on the left edge of the right colon and thick fibrous bands in the ileocecal region (Fig. 1). When the aforementioned bands in the ileocecal region were dissected and the cecum, appendix, and terminal ileum were released, it was observed that the jejunum entered between the leaves of the terminal ileum mesentery (Fig. 1, 2).

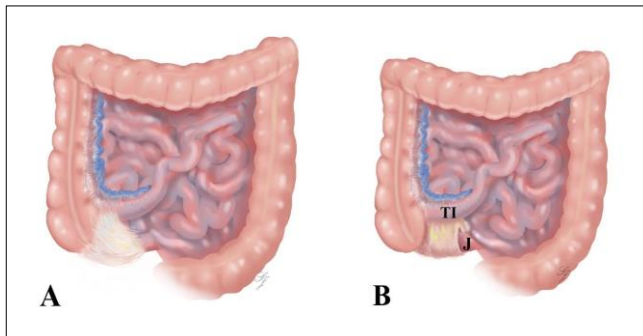


Figure 1: Diagrammatic illustration: A) Intestinal anatomy at first glance. B) Intestinal anatomy after releasing the ileocecal bands. Note that the terminal ileum and the proximal jejunum are almost overlapping. TI: Terminal Ileum, J: Jejunum

After the jejunum was released through the leaves of the terminal ileal mesentery, it was understood that this exit point in the mesentery was actually the duodenojejunal junction (color difference). Proximally, the duodenum was located posterior to the right colon in a "sack" (Fig. 3). This "sack" was surrounded by thick Ladd's bands on the lateral side, mesentery of the ascending colon on the medial side, the posterior surface of the cecum, and ascending colon on the anterior side, and the posterior abdominal wall on the posterior side (Fig. 3). The duodenum was completely located on the right side of the vertebral column. It was understood that the tortuous and thick venous structure extending within the mesenteries of the terminal ileum and ascending colon was superior mesenteric vein (SMV) since it was observed that proximally this venous structure and splenic vein joined to form the main portal vein. In contrast, the

anatomy of the superior mesenteric artery was normal. Ladd's bands did not compress the duodenum directly. Instead, the fibrotic bands compressing the duodenum were the bands extending between the medial and lateral walls of the "sack", and these bands were causing the duodenum to become kinked.

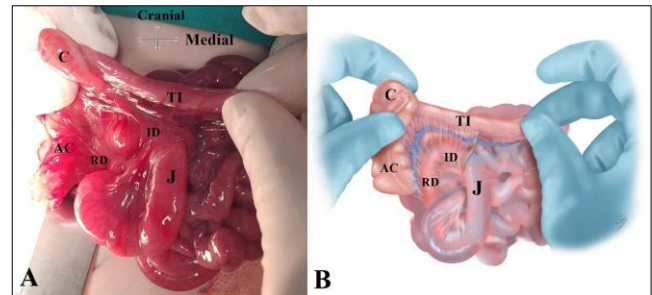


Figure 2: Intestinal anatomy after further releasing the bonds between the cecum, appendix, and terminal ileum: A) Original peroperative photo. Note that the ascending colon can easily be retracted upwards since it has a mesentery. B) Diagrammatic illustration. Note that the jejunum enters into the mesentery of the terminal ileum, the duodenojejunal junction is between the mesentery layers, the duodenum is initially in intramesenteric position and proximally becomes retrocolic localized. AC: Ascending Colon, C: Cecum, ID: Intramesenteric Duodenum, J: Jejunum, RD: Retrocolic Duodenum, TI: Terminal Ileum.

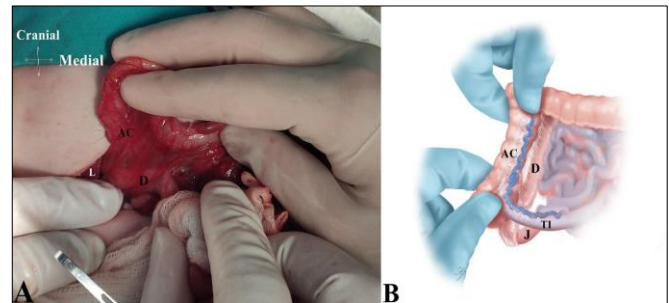


Figure 3: Anatomy of the duodenum within the retrocolic "sack": A) Original peroperative photo. B) Diagrammatic illustration. This illustration is not an exact copy of the peroperative photo. It is designed to make the retrocolic and intrasaccular position of the duodenum more understandable. AC: Ascending Colon, D: Duodenum, J: Jejunum, L: Liver, TI: Terminal Ileum.

The duodenum was released from these bands, and it was laid on the right quadrant as in the classical Ladd procedure. Then, the mesentery between the small intestine loops was released as possible. However, the situation was different for the colon; after the release of the duodenum and duodenojejunal junction from the terminal ileum mesentery and the retro-colic 'sack', they were left in the right quadrant of the abdomen, in accordance with the classical Ladd procedure, and it was observed that in this position the right colon was more likely to press on the duodenum. Therefore, the need to bring the right colon to the classical nonrotation position emerged. When the colon was brought to the non-rotated position, since the ileocecal region is facing to the right unlike nonrotation, the distal part of the terminal ileum was kinked and remained under the cecum. Therefore, the colon was left obliquely, with

the terminal ileum kinked least. The passage to the duodenum and jejunum was confirmed to be normal by giving air to the stomach.

The patient started feeding on the 5th postoperative day while bowel sounds and defecation were normal. The patient was discharged at the end of the second postoperative week. He is doing fine on follow-up of 15 months after surgery.

DISCUSSION

We report a new subtype of isolated duodenal non-rotation in which the duodenum is fully retro-colic in position. Unlike isolated duodenal nonrotation cases, in the surgical treatment of this subtype, separation of Ladd bands alone is not sufficient, additionally, the right colon should be placed in a nonrotation position and care should be taken not to kink the terminal ileum under the cecum.

Mall, examining human embryos, mentioned that herniated midgut loops need to return quickly and that in a 40 mm embryo, the loops should be completely inside the abdomen. [5] The rapid rotation of the midgut and simultaneous complete return had long been the dominant idea. However, there was a problem; a growing number of isolated duodenojejunal or cecocolic rotation anomalies had been reported, and Mall's theory was inadequate to explain this situation. Some authors noticed that the process of rotation, actually, was composed of numerous intermediate stages of development.

This confusion in theory and practice was resolved with Snyder and Chaffin's work. [8] The intestinal rotation concept developed by Snyder and Chaffin provides a background for an understanding of "classic" rotational anomalies as well as isolated rotational abnormalities. This concept focuses on 2 areas in detailing intestinal rotation; the duodenojejunal and the cecocolic segments. They demonstrated clearly that the rotation of these 2 segments is not simultaneous. Their description has greatly contributed to explaining and clarifying a variety of confusing clinical and roentgenographic presentations.

In their series of 40 cases, Snyder and Chaffin made a classification including the possible positions of the duodenojejunal and cecocolic segments in case of malrotation. They have mentioned this classification as 'close approximation' and said that "It is felt, however, that with further emphasis on a study of the position of the duodenojejunal loop at surgery, the number showing nonrotation of the duodenum will increase" [8]. Isolated duodenal malrotation appears for the first time in this classification (8 patients). Unfortunately, clinical information and intraoperative findings of these patients were not shared in their study.

Even though Snyder and Chaffin's emphasis on the asynchronous development of the duodenojejunal and cecocolic ring and the classification they created increased the number of case reports on the subject of isolated duodenal nonrotation, it still did not receive the attention it deserved. Lewis reported 4 cases in 1966, Prior reported 3 cases in 1974, Yadav and Pathak reported 2 pediatric cases in 1979. [9-11] In these studies, the duodenal segment was to the right of the midline, and neither of them was retro-colic, as in our case.

Stringer and Jamieson classified several types of rotation anomalies not only according to the classic embryological stages but intermediate stages of development too. [7] According to Stringer's classification, type 2A malrotation was defined as isolated duodenal nonrotation and normal colonic rotation as found in the presented case report.

In the large series of pediatric malrotation studies conducted in the following years, there were no or very few cases of isolated duodenal nonrotation which were limited to 1-2 cases. None of the existing isolated cases of duodenal nonrotation have signs of retrocolic duodenum. Interestingly, in the isolated duodenal malrotation case reports published in the English literature so far, the numbers of children and adult cases were almost the same. In the adult group, the duodenum was not retrocolic as in our case, and the mesentery root was not as narrow as found in our case. [12]

Findings in our case were unique and can be considered a new variant of isolated duodenal non-rotation. This unusual anatomy may be explained by earlier entry of the terminal ileal loop into the abdomen and by fixing the duodenojejunum by adhesions that block the final 90 degrees of normal intestinal rotation.

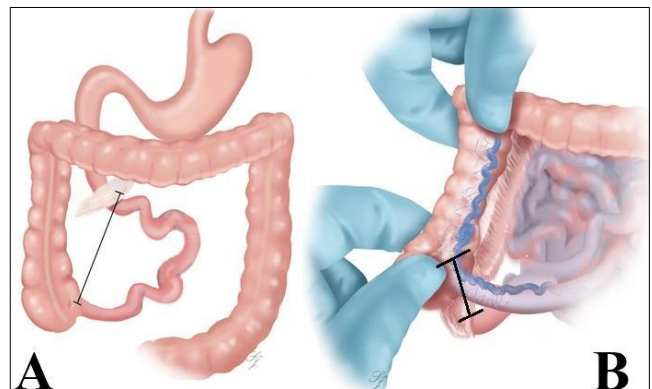


Figure 4: Mesenteric root in 'normal' isolated duodenal nonrotation (re-drawing from Stringer DA, Babyn PA, eds. *Pediatric Gastrointestinal Imaging and Intervention* [4]) (A) and mesenteric root in our patient (B). The long black lines indicate the mesenteric root

Our patient had all the features of self detorsion of the volvulus. In the literature, there is only one patient of isolated duodenal nonrotation who presented with volvulus. The authors conducted that midgut volvulus may occur when fixation of the right colon is deficient. [9] However, the fixation of the right colon was normal in our case. We can explain the volvulus in our patient with the narrowest mesentery root (Fig. 4).

In the surgical treatment of the subtype of IDN that we describe in this case, the right colon should be placed in the classical nonrotation position. However, there is an important issue to be noted here; unlike normal nonrotation cases, the ileocecal junction is faced to the right in isolated duodenal nonrotation.

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Therefore, the terminal ileum may kink under the cecum while setting the intestines to a nonrotating position as suggested by the authors. Therefore, the right colon should be placed obliquely in the abdomen rather than in the full nonrotation position.

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