Right Retrocolic Intrasaccular Duodenum and Intramesenteric Duodenojejunal Junction: Is It a Novel Variant of Isolated Duodenal Non-rotation?

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ABSTRACT

Midgut malrotation with volvulus is one of the true surgical emergencies of childhood. 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 negligence of other rare variants. Isolated pure duodenal non-rotation is such a variant. Herein, we report a case of an unusual variant of isolated pure duodenal non-rotation which presented with signs of midgut volvulus that was successfully treated.

Key Words: Newborn, Malrotation, Non-rotation, Isolated duodenal non-rotation, Midgut volvulus.

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INTRODUCTION

Midgut malrotation with volvulus is one of the true surgical emergencies of childhood. Frazer and Robbins first described the classic three stages of rotation and fixation process,¹ 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.² 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 negligence of other rare variants. Isolated pure duodenal nonrotation is such a variant. In this report, we present a case of an unusual variant of isolated pure duodenal non-rotation.

CASE REPORT

The patient was a newborn aged 3 days. He was born at 39 weeks of gestation with a birth weight of 3140 g. There was no problem in prenatal follow-up. The patient presented to our clinic on the third postnatal day due to bile-stained vomiting.

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There were no abdominal distention and tenderness. Blood analysis was normal except for the lactate being 4.5 mmol/L. Plain abdominal x-ray was not diagnostic. There was a "whirlpool" sign on the ultrasonographic examination.

Laparotomy revealed chylous fluid and lymphatic stasis in the mesentery, and loops of the small intestine were oedematous giving evidence of a previous torsion of the loops. The anatomy of the colon and small intestine seemed normal at the 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 (Figure 1A). When the aforementioned bands in the ileocecal region were separated, it was observed that the jejunum entered between the leaves of the terminal ileum mesentery and this exit point in the mesentery was actually the duodenojejunal junction (Figure 1B). Passing this junction, the duodenum was entering the backside of the right colon and was located in a "sack" (Figure 1C). This "sack" was surrounded by a thick Ladd's band on the lateral side, by fibrous tissues on the medial side, by the posterior surface of the cecum and ascending colon on the anterior side, and by the posterior abdominal wall on the posterior side. It was understood that the tortuous and thick venous structure extending along the medial wall of the right colon was the superior mesenteric vein (Figure 1D). The fibrotic bands compressing the duodenum were the bands extending between the medial and lateral walls of the "sack". The duodenum was released from these bands, and it was laid on the right quadrant. However, the situation was different for the colon. When the colon was brought to the non-rotated position, as the ileocecal region was facing to the right unlike non-rotation, 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 patient remains well 8 months later.



Figure 1: (A) Intestinal anatomy at first glance. (B) Intestinal anatomy after releasing the bands between the cecum, appendix, and terminal ileum. (C) Anatomy of the duodenum within the retrocolic "sac". (D) Abnormal course of the superior mesenteric vein.



Figure 2: (A) Mesenteric root in 'normal' isolated duodenal non-rotation (re-drawn from Stringer DA, Babyn PA, eds. Pediatric Gastrointestinal Imaging and Intervention10) and (B) mesenteric root in our patient. Hshaped lines indicate the mesenteric root.

DISCUSSION

The issue of isolated duodenal non-rotation was discussed by Snyder *et al.*who reported 8 cases in 1954, for the first time in the literature.³ Lewis reported 4 pediatric cases in 1966, Frior *et al.* reported 3 cases in 1974, and Yadav *et al.* reported 2 cases in 1979.⁴⁻⁶ In the large series of pediatric malrotation studies conducted in the following years, there were no or very few cases of isolated duodenal non-rotation which were limited to 1-2 cases.⁷⁻⁹ In this series, midgut volvulus was detected in only one case. In these studies, the duodenal segment was to the right of the midline, and none of them was retrocolic, as in the present case.

One of the most important features of our case is that the patient presented with volvulus. In the literature, there is only

one patient of isolated duodenal non-rotation who presented with volvulus, as reported by Frior *et al.*⁵ They concluded that midgut volvulus may occur when fixation of the right colon is deficient. Unlike Frior *et al.*'s case, the fixation of the right colon was normal in this case. We can explain the volvulus in our patient with the narrowest mesentery root (Figure 2).

Unlike normal non-rotation cases, the ileocecal junction is faced to the right in isolated duodenal non-rotation. Therefore, the terminal ileum may kink under the cecum while setting the intestines to a non-rotating position as suggested by the authors. Therefore, the right colon should be placed obliquely in the abdomen rather than in the full non-rotation position.

Our case differs from previously reported isolated duodenal non-rotations, due to the pure retrocolic position of the duodenum, intramesenteric duodenojejunal junction just below the terminal ileum, narrowest mesentery root and presentation with volvulus without mobile right colon. To the best of our knowledge, this is the first time in the literature to describe this type of isolated duodenal non-rotation. For this reason, we believe that we can label this type of isolated duodenal non-rotation as a 'novel' variant.

PATIENT'S CONSENT:

The patient's parents provided written consent for publication.

COMPETING INTEREST:

The author declared no competing interest.

AUTHOR'S CONTRIBUTION:

MH: Designed and draft the study.

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