Adults intestinal malrotations: a simple approach to recognize the most frequent types with MDCT

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Learning objectives

To identify congenital intestinal malrotation, a condition often undiagnosed up to adulthood, and to recognize several types of this anatomical variation. Diagnosis is often missed because of the misconception that this is just a disease of childhood. Congenital intestinal malrotations in elderly are easily recognized only if considered as possible cause of abdominal pain or obstruction. Knowledge of imaging features, clearly depicted by MDCT scans, can easily lead to diagnosis. In this exhibit typical imaging findings suggesting abnormal rotation during embryologic development are presented in order to evaluate and prevent possible complications.
Background

Midgut malrotation is a congenital anomaly referring to either lack of or incomplete rotation of the fetal intestines around the axis of the superior mesenteric artery during fetal development. In rare cases, the major cause of midgut malrotation seems to be an incomplete fixation. In order to understand intestinal rotation and its anomalies, a knowledge of intestinal embryology is essential.

The embryo's gut presents the form of a straight tube at the fourth week of life. During the fifth week, a vascular pedicle develops and the gut can be arbitrarily divided into foregut, midgut and hindgut. The superior mesenteric artery supplies blood to the midgut. The midgut is defined by a rapidly enlarging loop with the superior mesenteric artery running out to its apex. The cephalad portion of the loop gives rise to the first 20 feet of small bowel, and the remainder of the loop forms the distal small bowel and colon up to the splenic flexure. Intestinal rotation primary involves the midgut. The rotation of intestinal development has been divided into 3 stages.

Stage I occurs in weeks 5 to 10. It includes extrusion of the midgut into the extraembryonic cavity, a 90° counterclockwise rotation, and return of the midgut into the fetal abdomen. (Fig. 1)

Stage II occurs in week 11 and involves further counterclockwise rotation within the abdominal cavity completing a 270° rotation. This rotation brings the duodenal "c" loop behind the superiori mesenteric artery with the ascending colon to the right, the transverse colon above, and descending colon to the left. (Fig.2)

Stage III involves fusion and anchoring of the mesentery. The cecum descends, and the ascending and descending colon attach to the posterior abdomen. (Fig. 3)

Stinger classified several types of malrotation according to the embryologic state of development. Type Ia is defined as non rotation of the colon and duodenum, and type Ila is defined as non rotation of the duodenum only. In type IIb, the duodenum and colon show reversed rotation; in type IIc, there is only reversed rotation of the duodenum. In type IIIa, both the duodenum and colon are non rotated. Type IIIb is characterized by incomplete fixation of the hepatic flexure. In type IIIc, incomplete attachment of the cecum is seen. Type IIIId is characterized by an internal hernia near the ligament of Treitz.

Rotational anomalies are one of the most frequent malformations of the digestive tract; the incidence rates vary depending on clinical or autopsy investigation, but they are considered to correspond to 3-5% of surgical obstructions, and they appear in 0.5% of radiological series. Studies about incidence of malrotation in mixed populations show an influence of racial factors, radioactive or toxic insults, and low birth weight; a correlation with gender or number of siblings has not been demonstrated. Malrotation can be associated with various visceral situs and other congenital anomalies including polysplenia, interrupted IVC with hemi-azygos continuation and pancreatic anomalies. Patients with malrotation may present in the first month of life with biliary vomiting or obstruction secondary to midgut volvulus or abnormal peritoneal bands (Ladd’s bands).
Most of the intestinal malrotations, however, are diagnosed incidentally in adulthood. Nevertheless, it is important to recognize this anomaly because it may cause symptoms of intermittent obstruction or abdominal pain, mimicking pain related to pancreatic disease. Not all patients with malrotation present symptoms and the anomaly can be discovered only at autopsy. In some studies the cases of complete malrotation, with the ligament of Treitz to the right and below the 12th vertebra, presented a greater incidence of volvolus, while those with left-sided ligament were cases of incomplete malrotation, usually with a better prognosis.

The classic treatment for intestinal rotation is the Ladd procedure, which entails mobilization of the right colon, division of Ladd's bands and mobilization of the duodenum, division of adhesions around the SMA to broaden the mesenteric base, and an appendectomy. Obviously if the malrotation is complicated with an intestinal ischaemia, a resection is performed.
Fig. 1: Stage I (weeks 5-10)

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Fig. 2: Stage II (weeks 11-12)

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Fig. 3: Stage III (week 12-term.)

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Imaging findings OR Procedure details

Population
Two patients with abdominal pain and previous negative US study and five with extra-abdominal symptoms underwent contrast media MDCT. Scans were performed with a 64 slices MDCT (Siemens) and 128 slices MDCT (Siemens), with injection of intravenous contrast media (Iopamiro 350 mg) 110 +/- 20 ml at 3.0 ml/mL and saline chase 50 ml at 3.0 ml/mL.

Scan protocol
- pitch 1
- Kv 120
- mAs 180

Post-processing protocol
• Axial 1.0 and 3.0 mm
• Coronal 1.0 and 3.0 mm

We observed complete intestinal malrotation in five patients (type Ia); two patients only presented malrotation of small bowel (type IIa). Most frequent findings of malrotation generally are Treitz's angle absence, mesentery "whirl sign" without vascular suffering, vertical or inverted relationship between SMA e SMV, absence of pancreas's uncinate process, completely inverted position of small bowel and colon, cecal position. Furthermore, one patient was admitted to our hospital for abdominal aortic aneurysm. The CT scans showed a complete intestinal malrotation, with a right-sided small bowel and left-sided colon and inversion of superior mesenteric vessels. The patient underwent to an endovascular treatment and the CT performed one month later showed presence of horizontal duodenum with correction of the SMV-SMA relation and normal position of colon and small bowel. Failure of fixation may probably allow abnormal mobility of the duodenum; in this case, the major cause of malrotation seems to be an insufficient fixation.

1. Abnormal relationship between SMA and SMV
Malrotations are generally associated with an abnormal relationship between the superior mesenteric vessels, like SMV inversion and a vertical position of SMV. We observed this imaging finding in four completely-malrotated patients, presenting inversion of SMV in two cases and a vertical position in two cases.
Chou et al. evaluated this relationship on CT in normal patients and in those with abdominal masses. They found the SMV always to the right of the SMA in the first 3 cm of the vessels. They also found that the proximal jejunal vessels have a leftward direction. A rightward course of the proximal jejuna branches coupled with proximal SMV inversion could be an indication of midgut malrotation. Conversely, normal position of mesenteric vessels may be associated with malrotation, but Dufour et al. reported only a
3% of incidence. Thus, when anomalous relationship of the mesenteric vessels is present on CT, the location of the small bowel and the cecum should be carefully evaluated for the possible presence of malrotation. (Fig. 4, 5, 6)

2. Anomaly of pancreas
Mesenteric vascular inversion and abnormal rotation of the intestine may interrupt the normal rotation of the pancreatic primordia with malpositioning of the pancreatic buds, leading to hypoplasia or aplasia of the uncinate process. Between our patients, no one presented anomalies of uncinate process. (Fig. 7)

3. Whirl sign
The whirl sign is a swirl of mesenteric soft-tissue and fat attenuation with adjacent loops of bowel surrounding rotated intestinal vessels. This sign is more frequently associated with midgut and distal intestine obstruction but it's been described for the first time in a case of intestinal malrotation. The tightness of the whirl pattern reflects the degree to which the mesentery and vessels are rotated. We didn't observe this finding in any patient of our study. (Fig. 8)

4. Inverted position of small bowel and colon
Right-sided small bowel and left-sided colon can be easily observed on a CT scan. We found this sign in five patients with complete intestinal malrotation. Two patients with partial malrotation only presented small bowel malposition. (Fig. 9, 10)

5. Cecal and appendix position
Malrotated patients can present abnormal position of cecum, ileocecal valve and appendix. In our study four patients showed the ileocecal valve in the left flank and one patient in pelvis. Other patients presented cecum in normal position. (Fig. 11, 12)

6. Treitz’s angle absence
In this study we demonstrated the lack of duodenal sweep crossing the midline and the abnormal position of duodeno-jejunal junction in all patients with complete malrotation and in one patient with partial malrotation of duodenum. (Fig. 13, 14)
**Fig. 4:** Axial plane. Computed tomography scan through mid-abdomen with intravenous administration reveals superior mesenteric vein (blue arrow) lying to the left of the superior mesenteric artery (red arrow).

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Fig. 5: Axial plane. Scans through mid-abdomen show vertically oriented superior mesenteric vessels.

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Fig. 6: Coronal plane (MIP). CT scans show a rightward course of the proximal jejuna branches. In this image we can also note right and left colon on the left side of abdomen and small bowel on the right.

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Fig. 7: Axial plane. At the level of the pancreatic head, vertically oriented superior mesenteric vessels and aplasia of the uncinate process are visible.

**Fig. 8**: Coronal plane. Whirl sign in a 46-year-old woman with sigmoid volvulus.

© Volvulus of the gastrointestinal tract: appearances at multimodality imaging. RadioGraphics September 2009 vol. 29 no. 5 1281-1293
**Fig. 9:** Coronal plane. CT scans show the presence of a right-sided small bowel and left-sided colon in a patient with complete intestinal malrotation.

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**Fig. 10:** Axial plane. We can note right and left colon on the left side of abdomen (green arrow) and small bowel on the right (yellow arrow).

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Fig. 11: Axial plane. Ileocecal valve on the left flank in a patient with complete intestinal malrotation.

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Fig. 12: Axial plane. CT scans show abnormal position of appendix.

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**Fig. 13:** Axial plane. Note how duodenum does not cross the midline and the absence of Treitz's angle in patient with complete malrotation.

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**Fig. 14:** Axial plane. CT scans show the absence of Treitz's angle in patient with partial malrotation of duodenum (type IIa).

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Conclusion

Congenital intestinal malrotation is a condition often undiagnosed up to adulthood but it can cause chronic abdominal pain and severe complication; actually, diagnosis and treatment are usually delayed, after suffering prolonged symptoms and/or in emergency. Congenital intestinal malrotations in elderly are easily recognized only if considered as possible cause of abdominal pain or obstruction. Knowledge of typical imaging features, clearly depicted by MDCT scans, can easily lead to diagnosis of malrotation.
References