Pediatric intestinal malrotation in Upper Gastrointestinal Series: key points for diagnosis

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Learning objectives

The purpose of this poster is to describe the golden features for the challenging diagnosis of intestinal malrotation in children, that predisposes to life-threatening volvulus [1]. Every radiologist should be familiar to the main imaging findings of intestinal malrotation - which are often unclear and may lead to catastrophic delay in surgical treatment, not only in emergency situation, but also when the need for prophylactic surgery might be in question [1].
Background

It has been estimated that malrotation occurs in approximately 1 in 500 live births, and predisposes to acute abdomen events, which configure surgical emergencies [1,2]. Malpositioned and malfixed bowel loops are prone to volvulus that may shortly lead to catastrophic results due to acute vascular supply obstruction and lymphatic and venous drainage blocking, or it can lead to a variable degree of intestinal obstruction and necrosis [2].

A delay in diagnosis and treatment may be dramatic, resulting in small-bowel necrosis, shortgut syndrome, dependence on total parenteral nutrition or even death [2].

**Being familiar with the embryologic origin of the gut is essential to understand the pathogenesis of malrotation.**

The gastrointestinal tract is, at the beginning, a straight tube supplied by Superior Mesenteric Artery (SMA), and it is divided by the vitelline duct and SMA in two parts: the proximal portion that will form the distal duodenum, jejunum and proximal ileum, and the distal portion, that contributes to the distal ileum, cecum, appendix, and colon [3].

At 6\(^{th}\) week of gestation it herniates into the base of the umbilical cord entering in the extraembryonic coelom (fig 1) [3]. During the herniation, bowel makes a counterclockwise 90\(^{\circ}\) rotation around the SMA, and, as well explained by The Rope Model by Snyder and Chaffin, this movement results in a reciprocal position changing: the duodenojejunal loop courses downward right of the SMA and the ceco-colonic loop courses upward to the left of the SMA (fig 2)[3].

Further elongation of the midgut involves predominantly the proximal portion [3].

This latter, at 10\(^{th}\) week of gestation re-enters in the abdomen and undergoes an additional but final 90\(^{\circ}\) counterclockwise rotation, resulting in the final duodenal C-loop configuration (distal duodenum courses inferiorly and posteriorly to the SMA and then to the left and upward) [3]. Later, also distal midgut re-enters in the abdominal cavity undergoing 180\(^{\circ}\) counterclockwise rotation until the cecum is located to the right, such that the colon frame passes anteriorly to the SMA (fig 3)[3].

Cecum can further develop after birth, descending into the right lower quadrant of the abdomen (fig 4) [3].

It is important to mention that a mobile cecum in the upper quadrant is a common physiological variation in the newborn and, on the contrary, a normal position of the cecum does not exclude proximal intestine malrotation, because they develop and rotate separately [1,3].
During the first months after birth, a further cecum elongation may occur together with the fixation of the intestine [1-3]. The duodenojejunoal junction (DJJ) is fixed by the Ligament of Treitz, a fibrous tissue extending from the right diaphragmatic crus to the celiac artery, while the second, third and fourth portions of duodenum are fixed in the retroperitoneum as well as the colon frame, that is anchored at the retroperitoneum and at the greater omentum [1-3]. The small bowel loops are fixed by a broad mesenteric band, that elongates from the DJJ to the ileocecal valve in the lower right quadrant, thus allowing movement but preventing volvulus [1-3].

There is a wide range of anomalies of intestinal rotation in children and varying degrees of intestinal fixation [3]:

**Non-rotation** is the most common type of abnormal rotation, and represents the arrest of retroperitoneal fixation after up to 90° of rotation. In these cases the risk of midgut volvulus is very low, since the entire small bowel is located in the right abdomen and the colon in the left abdomen resulting in a wide mesenteric root. It is most commonly found incidentally (fig 5)[1,3].

**Incomplete rotation** instead is characterized by an arrest in rotation and fixation that occurs between 90° and 270°[1-3]. DJJ is on the right or at the level of the spine and the cecum is located in the upper abdomen, resulting in a short mesenteric root, that predisposes to volvulus due to Ladd’s Band (fig 6) [1-3].

Ladd’s Band is a peritoneal band that forms due to the embryonic malposition anomaly, and extends from the cecum to the retroperitoneum in the right upper quadrant [1,3].
Fig. 1: At 6th week of gestation intestinal tract herniates into the umbilical cord entering in the extraembryonic coelom

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Fig. 2: During the herniation bowel makes a counterclockwise 90° rotation around the SMA

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Fig. 3: Midgut re-enters in the abdominal cavity undergoing 180° counterclockwise rotation until the cecum is located to the right, such that the colon frame passes anteriorly to the SMA

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Fig. 4: Cecum can further develop after birth, descending into the right lower quadrant of the abdomen

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Fig. 5: The entire small bowel is located in the right abdomen and the colon in the left abdomen resulting in wide mesenteric root

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Fig. 6: DJJ is on the right or at the level of the spine and the cecum is located in the upper abdomen, resulting in a short mesenteric root, that predisposes to volvulus due to Ladd's Band

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Findings and procedure details

Imaging of malrotation includes a wide variety of radiological exams:

Ultrasound examination, radiation-free but highly operator-dependent, displays only the relationship between superior mesenteric vein and artery, that is inverted in patients with malrotation; and barium enema study is compromised by false negatives, since up to 32% of infants with malrotation have normal cecum position [1]. Plain film may show abnormal features in case of obstruction or in case of extreme malposition of the entire bowel loops, that result entirely located on one only side of the abdomen. But usually plain film shows normal gasification [3].

Upper gastrointestinal series is the preferred diagnostic technic, because it is able to depict directly the upper gastrointestinal loops and to identify the obstructed intestinal tract [2,3]. It is usually performed administering non-ionic contrast media (orally or through naso-jejunal tube - this latter allows a better distention of duodenum and a better visualization of the gastrointestinal tract anatomy) [2,3].

Then contrast is administered and patient is placed on his/her left side in order to fill the stomach, and then on his/her right side, aiming to empty the stomach and to fill the first portion of duodenum [3]. When the contrast media has reached the descending portion of the duodenum, the infant is placed supine again, to document on frontal view the entire course of the duodenal loop and the DJJ position [2,3]. Then on the right side again, to evaluate on lateral view the postero-anterior course of the last segments of the duodenum [2,3].

The normal C-loop anatomy of duodenum consists of 4 parts: the duodenal bulb, that is not fixed in the retroperitoneum, the descending portion, the transverse, and the ascending, which instead are fixed posteriorly to the retroperitoneum [2,3].

The ascending part continues in the jejunum crossing the ligament of Treitz and forms a tight downward concavity-flexure. This portion, called DJJ, normally must be placed to the left of the spine, at the level of pylorus and as posterior as the second portion of the duodenum (fig 7)[2,3].

These anatomical relationships are visible in frontal and lateral view.

On the frontal view the main physiological imaging features are:

the DJJ lays normally at or to the left to the left-sided pedicle of the vertebral body and at or above the level of L1-L2 intervertebral space or at the level of the duodenal bulb. The vertical segment of duodenum is normally shorter than the transverse one (fig 7)[2,3].
On the lateral view, the ascending part of the duodenum should be at the same plane or posteriorly to the descending one, and elongates up to the level of the duodenal bulb [2,3].

Malrotation consists of a varying degrees of abnormally positioned and fixed midgut loops, and any variation from these normal features should be evaluated as suspected for malrotation, especially in symptomatic children (bilious vomiting) (fig 8) [2,3]. Additional findings which may lead to diagnosis of malrotation are a Z-shaped configuration of the duodenum in case of obstructing Ladd’s bands, or a corkscrew-shaped duodenum when midgut volvulus occurs (fig 8, 9, 10) [2].

False positive rate is very high since the newborn intestine is very mobile, and radiologists must always be aware that abnormal findings should not automatically indicate malrotation, but they must be evaluated together with collateral findings and clinical features [2,3]. Accurate contrast media administration and meticulous infant positioning is mandatory, since overfilling the stomach, and external or internal (excessive intestinal gas) abdomen compression may cause DJJ displacement and lead to misdiagnosis, as well as the presence of an enteric feeding tube [2,3]. Also abnormal bowel distention may cause DJJ displacement and should suggests alternative diagnosis [2,3].

It is strongly recommended to evaluate the initial passage of contrast in duodenum directly with fluoroscopy, since when the more distal loops of the bowel fill with barium, the DJJ may be challenging to identify [2].

Moreover, a manual palpation should be performed, as suggested by Lim-Dunham et al [4], during fluoroscopy, thus allowing to evaluate the mobility of the duodenum: it has been estimated that in healthy young children the intestinal ligaments are lax, and this condition leads to a great mobility of the DJJ. In patients with malrotation instead a less mobile DJJ has been reported [2].

When diagnosis is unclear, other imaging exams should be reviewed, being always aware that they could be misleading as well: in particular US, that allows to evaluate the relationship between superior mesenteric artery and vein [2,3]. This latter is normally located to the right and anteriorly to the superior mesenteric artery, but this condition can be reversed in 60% of cases [2].
**Fig. 7:** Normal localization of Duodenojejuneal junction: DJJ normally must be placed to the left of the spine (green asterisk), at the level of pylorus and as posterior as the second portion of the duodenum. Red asterisk depict the different degree of abnormal DJJ localizations

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**Fig. 8:** Frontal View: DJJ is projected on the spine; abnormal duodenal C-loop (Z-shaped)

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**Fig. 9:** Frontal View: abnormal duodenal C loop. DJJ located on the right of the spine

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Fig. 10: Frontal View: corkscrew-shaped duodenum

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Conclusion

It is evident that experienced exam performance and evaluation are crucial in this difficult diagnosis, and it is important that radiologists, especially those who care children, are able to identify the main features that can lead to the correct identification of intestinal malrotation, in order to guide little patients’ surgical management.
Personal information

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