Intestinal Malrotation

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Learning objectives

- To illustrate the embryologic development of the midgut
- To display a correlation between the stage of embryologic failure and the anatomic result
- To help differentiate normal from abnormal features of intestinal rotation
- To give an overview of the different imaging modalities used to diagnose midgut malrotation

Background

Intestinal malrotation is a congenital abnormality in which the bowel is malrotated within the peritoneal cavity. As a result, an abnormal bowel fixation occurs predisposing to midgut volvulus, which is a potential life-threatening condition and requires an immediate surgical intervention [1].

It occurs in approximately 1 in 500 births and is mostly diagnosed in newborns presenting with bilious vomiting and abdominal distention [1]. Other symptoms include abdominal pain, diarrhea, and constipation [2]. Beyond infancy, intestinal malrotation can be an incidental finding in patients without symptoms (mostly adolescents and adults) or in patients with a malabsorption pattern such as diarrhea, nutritional deficiencies and/or failure to thrive [3]. On the other hand, due to complications such as volvulus and bowel obstruction, intestinal malrotation can have a more acute presentation.

Intestinal malrotation can be associated with many syndromes and anomalies such as small-bowel atresia, gastroschisis, omphalocele, Hirschsprung disease, heterotaxy syndrome, etc… However, this is beyond the scope of this presentation and is not further discussed [2].

The term intestinal malrotation comprises a range of malrotation types, each reflecting the time at which a failure occurs during the embryologic rotation (1, 2, 4). This emphasizes the fact that embryological knowledge of the normal midgut rotation is of utmost importance in understanding the pathophysiology of intestinal malrotation and in differentiating normal from abnormal radiographic features. The radiologist plays an important role in the diagnosis of this entity and its complications.
Findings and procedure details

1. Normal embryologic development of the gut

The gut arises from the yolk sac consisting of three anatomic sections each supplied by a different artery:

- Foregut: celiac trunk
- Midgut: superior mesenteric artery (SMA) (Fig. 1D)
- Hindgut: inferior mesenteric artery (IMA)

Development starts at week 4 of gestation. At this time the midgut starts to grow disproportionately to the abdominal cavity. By 6 weeks, the midgut herniates through the umbilical cord (Fig. 1). This herniation remains in place until week 10. During this stage, both the cephalad (duodenum, jejunum and proximal ileum) and caudad (distal ileum, cecum, colon) portion of the midgut rotate 90° counterclockwise (from a frontal point of view) around the SMA (Fig. 2-3). As a result, the cephalad portion of the midgut lies on the right side of the SMA and the caudad portion of the midgut lies on the left side of the SMA. Within this stage, after the initial rotation, the duodenojejunal portion starts to grow and rotates another 90° counterclockwise [2].

By week 10, the abdominal cavity has enlarged sufficiently at which point the midgut re-enters the abdominal cavity. During this intra-abdominal reduction the duodenojejunal portion undergoes a final 90° counterclockwise rotation while the colon undergoes a 180° counterclockwise rotation. As a final result, the duodenal part of the midgut lies on the left of the SMA and is located posteriorly while the colon lies on the right side of the SMA and is located anteriorly (Fig. 4). By this time, both parts of the midgut have rotated 270° in a counterclockwise manner.

Throughout the further gestation, fixation of the gut to the posterior abdominal wall occurs ensuring the normal anatomy. The small bowel is fixated by a broad mesenteric attachment from the duodenojejunal junction to the ileocecal valve. This broad attachment is a safety mechanism against small bowel volvulus.

2. Malrotation types

As mentioned above, the term malrotation comprises a range of malrotation types, each reflecting the time at which a failure occurs during the embryologic rotation. Three main types are described, i.e. nonrotation, partial malrotation and reversed malrotation [2,5].
In nonrotation, only the initial 90° counterclockwise rotation occurs so that the duodenojejunal junction lies on the right side and the colon lies on the left side of the SMA (Fig. 5-A).

Partial (incomplete) malrotation implies a failure during the final 180° counterclockwise rotation. This term is used in all malrotation types between a nonrotation and a normal rotation (Fig. 5-B).

When rotation is clockwise, the result is termed reversed malrotation in which the duodenum lies anteriorly to the colon.

In the classification of intestinal malrotation, an anomalous fixation of the mesentery must be mentioned as it may be the cause of an intestinal internal herniation [6].

3. Complications

Due to an abnormal rotation, the normal subsequent mesenterial attachment is disturbed. The result may be a narrow(ed) mesenterial attachment predisposing to volvulus, a well-known cause of intestinal obstruction. Although this complication can occur in any type of malrotation, it is more frequently seen in partial malrotation than in a complete nonrotation. This twist of the midgut can compromise the normal vascular supply making it a surgical emergency due to the potential life-threatening result of midgut ischemia and gangrene.

Another complication is caused by the so-called Ladd's bands, which are peritoneal bands formed to attach an incompletely rotated cecum and is another well-known cause of intestinal obstruction (Fig. 6).

As mentioned before, anomalous fixation of the mesentery can give rise to an internal herniation with potential life-threatening complications such as bowel strangulation and ischemia [3].

4. Imaging features and different imaging modalities

- Plain X-ray:
Conventional radiography is neither sensitive nor specific for intestinal malrotation [3,7]. Several potentially abnormal air-patterns can be found: an abnormal air distribution (Fig. 7), a so-called double-bubble appearance, a gasless abdomen and a dilated duodenum and stomach with paucity of gas in the further abdominal tract (Fig. 8)[2].

- Upper GI series

The imaging modality of choice remains the upper GI study. Knowledge of the normal embryologic development and the normal anatomic result is important in performing and interpreting upper GI-series. The duodenojejunal junction is located to the left of the vertebral body (left-sided pedicle) and this at the level of the duodenal bulb on a standard AP-view [1]. On a lateral view the duodenojejunal junction is located posteriorly [1]. Besides the AP and the lateral view, we use the (prone) right anterior oblique (RAO) view as it depicts the typical inversed C-shape configuration of the normal course of the duodenum (Fig. 9).

In malrotation, the distal duodenum has an abnormal course. On a strict AP-view, this can be seen as an anomalous position of the duodenojejunal junction to the right of the vertebral body and, in some cases, with a lower level of the duodenal bulb (Fig 10A). This duodenojejunal junction can have an anterior location, which can be depicted on a lateral view. In the RAO-view, we often see a C-shape configuration (Fig. 10B) of the duodenal course as opposed to the inversed C-shape configuration of the normal duodenum (Fig. 9B).

In case of an intestinal volvulus, the distal duodenum and the proximal jejunum can have a "corkscrew" configuration (Fig. 11). Once seen, urgent surgery is required in order to avoid catastrophic events such as bowel ischemia and gangrene.

In case of an obstruction due to Ladd's bands, a Z-shaped configuration of the duodenum can be depicted.

Since the upper GI study is of paramount importance in diagnosing malrotation, additional attention should be given to the implementation of this examination:

**As a first key rule,** documentation of the first bolus through the duodenum is mandatory in making an adequate diagnosis! Once the contrast medium passes beyond the duodenum, identification of duodenal bulb and duodenojejunal junction becomes difficult.
As a second key rule, both duodenal bulb and duodenojejunal junction should be documented in AP, lateral and RAO projections. This can be obtained by rotating the patient in a RAO (and over to the lateral) position once the duodenojejunal flexure is reached in a frontal projection.

- Contrast Enema

Contrast enema can be used to illustrate the anatomic position of the colon and the cecum (Fig. 7, Fig. 12). However, this examination is nonspecific [3] because cecal location can be variable in patients without malrotation. On the other hand, the cecum can have a normal position in patients where intestinal malrotation is present. This explains why contrast enema is no longer the primary imaging modality of choice.

Nevertheless, in cases of an unequivocal upper GI study, a contrast enema can be of value in diagnosing doubtful cases of malrotation. Acquisition of delayed abdominal radiographs, performed after an initial and unequivocal upper GI study, serves as a good alternative in documenting the cecal position [1].

- Ultrasound (US)

US is frequently requested by the clinician in order to exclude pyloric stenosis, another cause of neonatal vomiting. Although clinical presentation is very different in these two entities (bilious versus non-bilious vomiting), US is a readily available investigation in the assessment of both the pyloric canal as well as the normal relationship of the SMV and SMA.

An inversion of the normal relationship of SMV and SMA (the SMV swirls to the left of the SMA) is indicative for malrotation (Fig. 11A; Fig. 13). Because these sonographic findings are not very sensitive [2,8], further investigation is warranted in order to exclude intestinal malrotation.

Although ultrasound is an excellent imaging modality, it’s results are strongly operator-dependent. Additionally, due to the superimposed intestinal air, both the SMV and the SMA are not always clearly detectable.

- Cross-sectional imaging modalities (CT and MRI)
These imaging modalities are not considered to be the first imaging modalities of choice. However, the diagnosis of intestinal malrotation with cross-sectional imaging is frequently an incidental finding (Fig. 14, Fig. 15), mainly in adolescents and adults [2,3].

Both Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) can be used to display the relationship between SMV and SMA as well as signs of volvulus such as the "whirlpool" sign. CT and MRI can also depict the orientation of both small and large bowel. An additional advantage of these imaging techniques is that other abnormalities, in association with syndromes or anomalies, can be illustrated.

5. Treatment options

The most frequently used surgical approach in patients with malrotation is the Ladd's procedure in which there is a reduction of the midgut volvulus, followed by a dissection of the obstructing peritoneal bands and a reorganization of both small and large bowel in their normal anatomical position. Finally, an appendectomy is performed [2,6].

Nowadays, this procedure is performed using a laparoscopic technique in patients with uncomplicated intestinal malrotation. In complicated cases there may be need for an open approach.

Follow-up is warranted due to postoperative complications such as infections and adhesions. Although rare, recurrent volvulus can occur after a Ladd's procedure [2].

Images for this section:
**Fig. 1:** Illustration of the initial midgut herniation through the umbilical cord from a lateral (A-B) and an anterior oblique (C-D) point of view. Development of the midgut starts at week 4 of gestation. At this time the midgut starts to grow disproportionately to the abdominal cavity. By 6 weeks, the midgut herniates through the umbilical cord. The midgut is supplied by the superior mesenteric artery (D). Based on Dr. R. Acland's animation [9].
**Fig. 2:** Illustration of the first 90° counter-clockwise rotation from a lateral point of view. (A) displays the initial position in which the cephalad portion of the midgut lies superiorly to the caudad midgut portion. The counter-clockwise rotation is shown as a curved arrow in (A) and (B). The result is illustrated in (C) where the cephalad portion of the midgut lies on the right and the caudad portion lies on the left. Based on Dr. R. Acland's animation [9].
Fig. 3: Illustration of the first 90° counter-clockwise rotation from an anterior oblique point of view. The herniated midgut rotates 90° around its axis (SMA) and this in a counterclockwise manner. The net result is shown in figure. 2C. Based on Dr. R. Acland's animation [9].
Fig. 4: Illustration of the final 180° counter-clockwise rotation during . (A) Shows the final 180° counter-clockwise rotation of the cephalad portion of the midgut which rotates under and to the left of the caudad portion of the midgut. (B) Displays the final 180° counter-clockwise rotation of the caudad portion of the midgut. (C) Illustrates the final result where the colon is located anteriorly and the duodenojejunal junction is located posteriorly. Based on Dr. R. Acland's animation [9].
**Fig. 5:** Illustration of the two main types of malrotation. (A) In nonrotation, a failure occurs after the initial 90° counter-clockwise rotation. The net result is a colon (and cecum) which is positioned to the left and a small bowel which is positioned to the right. (B) In partial malrotation, the failure occurs during the final 180° counter-clockwise rotation. Depending on the degree of rotation, final intestinal position may vary between nonrotation and a normal rotation. (C) When rotation is clockwise, the net result is a colon with a posterior location and a small bowel with an anterior location.
Fig. 6: Illustration of the so-called Ladd's bands. These peritoneal bands are formed to attach an incompletely rotated cecum. They are a well-known cause of intestinal obstruction.
Fig. 7: Conventional radiography and contrast enema in a newborn girl with bilious vomiting. (A) Conventional X-ray of the abdomen demonstrates an abnormal air distribution with a colonic air configuration with haustations (arrowheads) mainly projected on the left side and a small bowel air configuration (arrows) projected on the right side. As a malrotation type was presumed to be present, further radiographic investigation was suggested. (B) Contrast enema confirms an abnormal position of both colon and small bowel. The colon (arrowheads) is located in the left hemi-abdomen. During retrograde filling of the small bowel (arrows), it was demonstrated to be in the right hemi-abdomen. At surgery, a nonrotation was confirmed.
Fig. 8: Conventional radiography and upper GI study in a newborn boy with bilious vomiting and a history of gestational polyhydramnios. (A) Conventional radiography illustrates a dilated stomach and proximal duodenum with paucity of gas in the distal part of the GI-tract. (B) Consequent upper GI study shows a distension of the stomach and proximal duodenum ending in a beaked obstruction. The diagnosis of duodenal atresia was presumed prior to surgery. However, the diagnosis of a midgut volvulus due to a partial malrotation was made peroperatively.
Fig. 9: Upper GI study in a vomiting 5-year-old boy. (A) A-P view displays a normal anatomy with the duodenojejunal junction (circle) situated at the left side of the thoraco-lumbar spine and at the level of the duodenal bulb. (B) Right anterior oblique view shows a typical inverted C-shaped configuration (dashed circle) of the course of the duodenum, illustrating the normal duodenal anatomy (compare to the abnormal findings in Figure 9).
**Fig. 10:** Upper GI-study in a vomiting 2-months-old premature child. (A) A-P view illustrates an abnormal position of the duodenojejunal junction situated entirely to the right of the thoraco-lumbar spine (circle). (Compare to the normal position of the duodenojejunal junction in figure 8A). (B) Right anterior oblique to lateral view shows an abnormal course of the duodenum, running posteriorly (dashed circle) (compare to the normal inverted C-shape configuration of the duodenum as in figure 8B). Barium enema (not shown) displayed an abnormal position of the cecum, located high in the right upper quadrant. Surgery confirmed the presence of a partial malrotation.
Fig. 11: Examination of a newborn patient with persisting vomiting after birth (day 2). Doppler ultrasound examination of the mesenterial vasculature (A) illustrates a malposition of the SMV (arrow) on the left side of the SMA (arrowhead). During the examination a typical "corkscrew"-like swirl of the SMV around the SMA was noticed (not shown). Although these finding are suggestive for malrotation, further radiographic work-up was warranted. An upper GI study was performed displaying a difficult (but present) passage of contrast beyond the proximal part of the duodenum with duodenal narrowing and a starting "corkscrew" configuration (dashed line in D) suggestive for a midgut volvulus. Although there is passage of contrast through this narrowing, the diagnosis of a complete nonrotation with a partial volvulus was made during surgery.
Fig. 12: Examination of a newborn presenting with abdominal distension and vomiting after birth: (A-B) X-ray of the abdomen (A-P and lateral view ) showing a marked distension of the stomach, but with a further gasless abdomen. (C) Contrast enema illustrates an abnormal position of the cecum which crosses the midline but is located superior in the right abdomen suggestive for a partial malrotation. These findings were confirmed at surgery with a 360° midgut volvulus and signs of bowel ischemia. A resection of part of the jejunum was unavoidable.
Fig. 13: Ultrasound of a newborn boy with bilious vomiting illustrating an abnormal relation between the superior mesenteric artery (SMA) and the superior mesenteric vein (SMV) in which the vein (B) is located at the left side of the artery (A). This relation implies an abnormal mesenteric rotation. During surgery, a complete nonrotation was found.
Fig. 14: A 31-years-old woman presenting at the emergency department with lower abdominal pain. Sonographic examination displayed an amount of free fluid in the lower abdomen. However, the appendix could not be visualized. A subsequent CT-scan of the abdomen was performed after administration of a rectal water soluble contrast medium (as well as intravenous iodine). As an incidental finding, a complete nonrotation was detected with the contrast-filled colon (arrowheads) located mainly in the left hemi-abdomen and the small bowel (small arrows) located in the right hemi-abdomen. The cecum was located in the lower abdomen on the midline (circle in C). The thickened appendix is located on the right side of the cecum (large arrow in C) with inflammation of the surrounding soft-tissue and free fluid in the Douglas cave (seen in D): Acute appendicitis in a patient with a complete nonrotation (E).
**Fig. 15:** MRI (coronal T2-weighted sequence) of a 55-years-old man with atypical thoracic complaints. As an incidental finding a complete nonrotation was found with the entire duodenum (small arrowheads) and duodenojejunal flexure (arrows) lies on the right side and the entire colon lies on the left side of the abdomen (large arrowhead).
Conclusion

Intestinal malrotation remains a challenging diagnosis for both surgeon and radiologist. Knowledge of the embryologic development is mandatory in understanding the radiographic features of malrotation and its complications. Upper-GI examination remains the imaging modality of choice.

Personal information

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