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Incidental Malrotation in Adolescent Diagnosed with Appendicitis

By: Dr. David Ho¹, Dr. Jinsy Jacob², Dr. Russell Mordecai¹, Dr. Adam Richards³

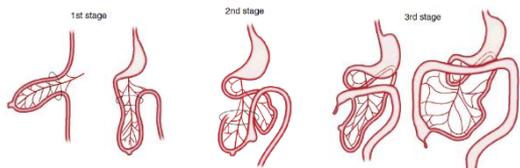
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INTRODUCTION

The human gastrointestinal tract consists of the **foregut**, **midgut**, and **hindgut**. During the **4th week** of development, the duodenum completes its rotation and fixation to the upper left abdominal cavity. In the **5th week** of development, the midgut elongates and is connected to the abdominal wall via vitelline duct. Rapid elongation of the midgut causes it to protrude into the umbilical cord at approximately **week 6** where the cephalic portion of the midgut will give rise to the distal duodenum, jejunum, and proximal ileum. The caudal portion will give rise to the distal ileum, cecum, vermiform appendix, ascending colon and proximal 2/3 of the transverse colon. In **week 7**, the midgut rotates around the SMA which serves as the axis of rotation. The cecal bud develops as a dilatation of the caudal loop of the midgut. The appendix develops during **week 8** at the distal end of the cecum. In **week 9**, the midgut undergoes 90-degree rotation into the umbilical cord. During the **week 11**, the midgut retracts into the abdominal cavity and in doing so, undergoes 180 counterclockwise rotation. **The jejunum returns and assumes position at the LUQ. The ileum assumes position at the RLQ. The appendix will rest in the RUQ but descends to the right iliac fossa, assuming its final position in the RLQ.**



Defects in the first stage of rotation result in extorsion of the cloaca and are rare¹. Defects in the second stage of rotation commonly leads to **nonrotation**. These are the most common of the three defect categories¹. Nonrotation results in placement of the small bowel to the right and large bowel to the left. Finally, defects in the third stage result in defects of elongation of the cecum, causing it to remain in the RUQ.

Reverse rotation occurs when the midgut rotates 180 degrees **CLOCKWISE**, rather than counterclockwise. This will place the colon behind the SMA. **Malrotation** occurs when there is anomaly in the rotation AND fixation of the midgut. This positions the jejunum and ileum to the right of the SMA and the cecum in the sub pyloric region¹.

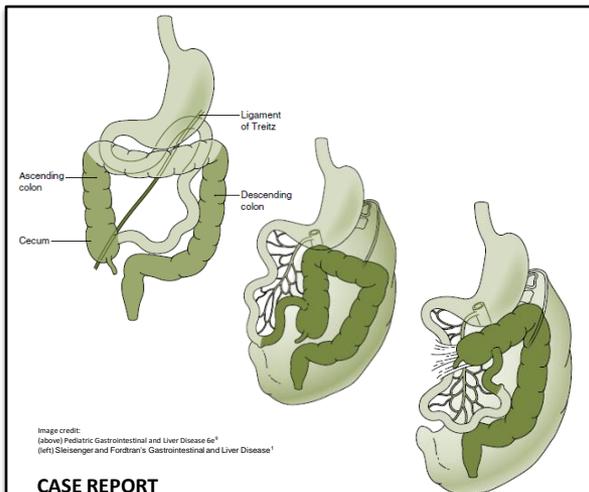


Image credit:
(above) Pediatric Gastrointestinal and Liver Disease 6th
(left) Sleisenger and Fordtran's Gastrointestinal and Liver Disease¹

CASE REPORT

17-year-old male patient with a history of autism presenting with right lower quadrant pain that started in the morning. The appendix was not able to be visualized by ultrasonography. An abdominal and pelvic CT with contrast showed a dilated appendix with surrounding inflammation. **Malrotation was not identified on CT Scan.** Patient was transferred to a nearby Children's Hospital for laparoscopic appendectomy. During the procedure, the pediatric surgeon noted that initially, neither the appendix nor cecum was present in the right lower quadrant. **The appendix was eventually found just caudal to the gallbladder and liver.** No Ladd bands (fibrous tissue that connect the cecum to the retroperitoneum) were located. The cecum was mobile and tended to retreat to the RUQ with the patient in reverse Trendelenburg position. The surgeon was **eventually able to relocate the cecum to the RLQ.** The appendix was found adhered to the back portion of the cecum. The appendix was inflamed but showed no signs of perforation. The appendix was ligated using endoloops and removed without difficulty. Patient tolerated the procedure without complications and recovered in PACU. The diagnosis was acute appendicitis with incidental malrotation / non-rotation.

DISCUSSION

Most patients with intestinal malrotation present in the first month of life. Adult presentation is rare¹. **Approximately 85% of cases occur in the first two weeks of life².** Symptomatic infants will present with vomiting, usually due to an intestinal obstruction or volvulus¹. In a study by Dekoneko et al., it was found that the most common symptom was vomiting, followed by abdominal pain, feed intolerance, and bilious vomiting⁴. Asymptomatic patients may never be diagnosed. As a result, the true incidence is unknown¹. In adults with malrotation, the presentation can vary widely with acute and chronic findings. Acute symptoms result from bowel obstruction or bowel ischemia³. Chronic symptoms may be the result of intermittent bowel obstructions³. Some cases of adult malrotations are diagnosed incidentally during abdominal surgery² as in the case of our patient.

CONCLUSION

This case represents an uncommon finding for a patient in the adolescent age group¹. A pubmed search did not yield many similar cases; however, **a study titled Appendicitis in adults with incidental midgut malrotation: CT findings by Ely et al. found only 8 such cases in their institution between 1998 and 2009⁸.**

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