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Nonbilious Vomiting in a 4-Week-Old Male: A Case Report and Review of the Literature

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Case Report

A 4-week-old male was born at 38+6 weeks via spontaneous vaginal delivery with a birth weight of 3.41 kg. His birth was uncomplicated, and he was discharged home after a 48-hour stay in the hospital. After discharge, he was breastfed exclusively, had consistent wet diapers and stools, and continued to grow and gain weight. However, at 4 weeks of age, he presented to his primary care physician (PCP) with increasingly frequent spit ups and decreased stools for 1 week. Per the mother, the spit ups had started on the second day of life, but they had initially improved significantly with head elevation after feeds. Over the last few weeks, they had become more frequent and voluminous, which elicited concern. His PCP suspected the infant to have gastroesophageal reflux and failure to thrive, so he switched the infant to Enfamil reflux formula. At the following visit, the mother reported that the frequency of spit ups had worsened, so the PCP ordered an upper gastrointestinal (GI) series with follow through and subsequently admitted the patient to the hospital for severe gastroesophageal reflux, failure to thrive, and intestinal malrotation (see Figure 1).

Final Diagnosis

Gastroesophageal reflux and malrotation.

Hospital Course

On the initial physical examination in the hospital, the patient was awake and appeared well-hydrated. His abdomen was soft, nontender, nondistended, and had active bowel sounds and no organomegaly. The rest of his physical examination was also normal. Overall, he appeared stable, nontoxic, and in no acute distress. The patient was subsequently evaluated by a pediatric surgeon, who placed him on reflux precautions and monitoring for reflux episodes. The reflux precautions included smaller feedings and keeping the infant upright after feedings as well as keeping the head of his bed elevated. With these reflux precautions, he started gaining weight, and the episodes of spitting up decreased. The decision was then made to proceed with correction of the congenital malrotation to prevent future progression to volvulus and bowel ischemia. A nasogastric tube (NGT) was placed, and the patient was placed on NPO (nothing by mouth). After a day, the NGT was removed and oral feeds were started, but this resulted in bilious vomiting. The next day, the NGT was replaced due to increased vomiting and abdominal distension secondary to ileus. After 2 days of bowel movements, flatulence, and decreased abdominal distension, the NGT was once again removed. The infant began tolerating oral feedings without subsequent emesis. On postsurgical day 7, the infant was discharged home, where he continued to tolerate feedings and gain weight.

Discussion

Malrotation of the intestines, also referred to as intestinal rotation abnormality, occurs in 0.5% of the population and can lead to devastating consequences, including volvulus, ischemic bowel, short bowel syndrome, or even death.[1,2] The condition results from abnormal rotation of the intestines during embryological development and is most often diagnosed in the first year of life.[1] While it is commonly agreed that the Ladd procedure is the treatment of choice for symptomatic malrotation, there are still differences in opinion about whether or not to treat incidental findings of malrotation with prophylactic surgical intervention.

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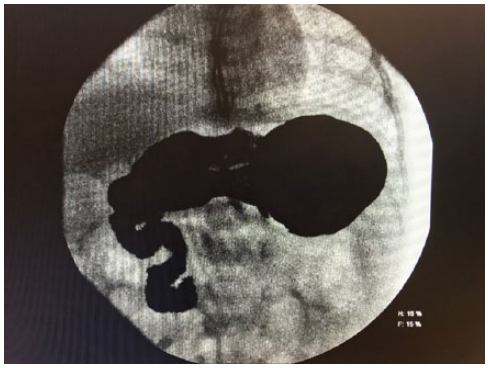

Figure 1. Upper GI series shows the duodenojejunal segment is positioned vertically in our patient providing evidence of malrotation.

During normal embryologic development, the gut tract expands so rapidly that the embryonic cavity cannot accommodate its size. As a result, the intestines extrude out into the amniotic space outside of the fetal abdomen. As the bowel returns to the abdomen by the 8th to 10th week of gestation, the loops of intestine normally rotate 90° counterclockwise around the superior mesenteric artery. They continue to rotate an extra 180° counterclockwise, resulting in a complete rotation of 270°. This motion returns the bowel into the posterior abdomen with the duodenojejunal junction (location of the ligament of Treitz) placed to the left of the midline and the cecum placed in the right iliac fossa.

Intestinal malrotation occurs when the normal embryological process of bowel rotation happens incorrectly or incompletely. Symptomatic malrotation occurs in as many as 1 in 200 live births, but the asymptomatic incidence is unknown because many of these cases are never diagnosed.[1] With incomplete intestinal rotation, the duodenojejunal portion of the small intestines is positioned vertically and the cecocolic portion of the intestine is rotated 90° instead of the full 180°. The cecum is positioned in the mid to upper left portion of the abdomen instead of the right lower quadrant, and peritoneal bands, also known as Ladd bands, fix the cecum to the lateral wall of the abdomen.[2] Occasionally, these bands can cause intermittent intestinal obstruction if they compress the duodenum.

Malrotation can cause a wide range of clinical symptoms. Infants with malrotation that has progressed to volvulus present with bilious vomiting and symptoms of acute bowel obstruction, including obstipation and abdominal distension.[2] They may also present with hypovolemia with or without septic shock.[2] These cases most commonly present in the first month of life and 90% present within the first year. Malrotation may also manifest as recurrent episodes of bilious or nonbilious vomiting, failure to thrive, solid food intolerance, malabsorption, diarrhea, bloating, and abdominal pain.[3] Some infants may present with nonspecific findings, which mimic colic, gastroesophageal reflux, pancreatitis, or biliary obstruction.[3-5] As the children get older and the symptoms become less specific, diagnosis becomes more challenging and less timely. The upper GI series, which has a sensitivity of approximately 96% in infants, is the imaging study of choice when diagnosing malrotation patients.

Treatment of symptomatic malrotation entails surgical intervention. The Ladd procedure, the current standard of care for malrotation, involves lengthening the mesentery and positioning the small and large intestines in the right and left sides of the abdomen, respectively. This surgical intervention aims to reduce the risk of volvulus in the future as well as alleviate other symptoms caused by malrotation. On the other hand, physicians still disagree on the best treatment for asymptomatic malrotation, which is often found incidentally on imaging done for other conditions.

Some physicians argue against unnecessary surgery to prevent complications that may never even occur. Malrotation, and subsequent progression to volvulus, presents most commonly in the first month of life, and 90% of volvulus cases occur in the first year of life.[6] As children get older, their risk of volvulus significantly decreases. Less than a third of malrotation cases in older children lead to the complication of volvulus, and this fraction continues to decrease as individuals continue into adulthood.[7] Therefore, some argue that the risk of volvulus past a certain age does not outweigh the risks associated with surgery. The Ladd procedure, the current treatment for malrotation, is associated with numerous complications in 8% to 14% of patients including intestinal obstruction.[8,9] Despite the procedure, recurrent volvulus can occur in up to 8% of patients.[10] The risks associated with surgery are greater in individuals with other comorbidities, such as heterotaxy syndrome.[2,8,11] This leads many to maintain that the risks of surgical intervention do not outweigh the benefits, especially in those with other significant comorbidities, such as major cardiac disease.[8]

Nevertheless, most surgeons agree that the benefits of a Ladd procedure to correct intestinal malrotation offsets the risks.[12] A Ladd procedure not only prevents the life-threatening complication of volvulus but also relieves other GI symptoms associated with malrotation. The lifetime risk of volvulus secondary to malrotation has been estimated to be up to 20%.[5,13] While the risk of malrotation progressing to volvulus decreases with age,

current literature reveals multiple reports of adolescents and adults presenting with volvulus.[7,10,13-15] Since predicting volvulus is so challenging and it can become life-threatening in a short amount of time, a Ladd procedure is the only way to prevent volvulus in individuals with malrotation.

Furthermore, even if malrotation never progresses to volvulus, it can cause chronic GI symptoms, including chronic abdominal pain, malabsorption, diarrhea, solid food intolerance, common bile duct obstruction, and abdominal distension.[3] Fifty percent to 70% of individuals with malrotation eventually develop symptoms associated with their anomaly.[3,7,16] These chronic and unspecific symptoms are frequently misdiagnosed as irritable bowel syndrome, biliary disease, peptic ulcer disease, or psychiatric disorders.[17] These frequent misdiagnoses result in the patients suffering with these chronic complaints for months, if not years. This anatomical anomaly can also lead to delayed diagnosis of appendicitis due to the unusual location of the appendix, and this delay in diagnosis and treatment leads to increased incidences of complications, such as appendiceal rupture, in these individuals.[17] Therefore, in addition to preventing volvulus, the Ladd procedure can alleviate other chronic GI symptoms.

Last, the Ladd procedure leads to better outcomes, including reduced morbidity and mortality, when performed in nonemergent situations.[10] Elective Ladd procedure can be performed laparoscopically with shorter operative times (63 minutes vs 76 minutes) and potentially less postoperative adhesions and other subsequent complications.[18] Once malrotation has progressed to volvulus, an open procedure is preferred to a laparoscopic one. Furthermore, waiting for malrotation to progress to the emergent situation of volvulus may result in severe morbidity for these patients including short bowel syndrome and death.

Conclusion

Malrotation is a relatively common anomaly that can lead to life-threatening volvulus and bowel ischemia. We present a case of malrotation found incidentally on imaging and use this case as a nidus for discussion over recommended treatment for incidental malrotation. While some physicians advocate against prophylactic surgical intervention in patients presenting with incidental findings of malrotation, many others argue that the benefits of Ladd prophylaxis outweigh the risks. In order to reach a consensus on how best to approach and treat incidental findings of malrotation, further investigation and discussion of this controversial topic is necessary.

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Author Contributions

DN: Contributed to conception and design; contributed to acquisition, analysis, and interpretation; drafted manuscript; critically revised manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring integrity and accuracy.
WS: Contributed to conception and design; contributed to acquisition and analysis; drafted manuscript; critically revised manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring integrity and accuracy.
JD: Contributed to conception; contributed to acquisition, analysis, and interpretation; drafted manuscript; critically revised manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring integrity and accuracy.
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