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## Case Report

# Target sign of intussusception versus whirlpool sign of midgut volvulus ☆,☆☆

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## ABSTRACT

We report the case of a 2-month-old boy who presented with emesis and was initially thought to have an intussusception based on ultrasound findings, but was later found to have malrotation with midgut volvulus. He was surgically detorsed before any bowel necrosis occurred, but later developed recurrent volvulus due to a surgical adhesion acting as an anchor point. The aim of this report is to highlight the imaging similarities and differences between intussusception and the more serious midgut volvulus in order to expedite proper care and preserve bowel. Malrotation with midgut volvulus is a pediatric surgical emergency involving twisting of a congenitally shortened mesentery around the superior mesenteric artery, leading to rapid vascular compromise and ischemic necrosis of small bowel. Prompt diagnosis is critical but difficult, as imaging findings in volvulus can appear similar to those in intussusception. Treatment with a Ladd procedure can safely and effectively reduce the volvulus and prevent recurrence.

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## Background

Intestinal malrotation is a congenital abnormality of the embryologic 270-degree counterclockwise rotation of intestines. The resulting intestines are attached by a shortened “pedicle” of mesentery that is prone to volvulus. It is also associated with fibrous bands of peritoneum known as Ladd bands that can contribute to obstruction. Midgut volvulus is the most

common complication of malrotation in infants, occurring in about one-third of infants with malrotation before 1 month of age, and carries a mortality rate of approximately 4%. [1]

Abdominal ultrasound is a useful initial modality to assess for volvulus, and may demonstrate a circular mass of mesentery and superior mesenteric vein (SMV) twisting clockwise around the superior mesenteric artery (SMA), known as the “whirlpool sign.” Unfortunately, the circular mass may be strikingly similar to the “target sign” or “donut sign” seen in

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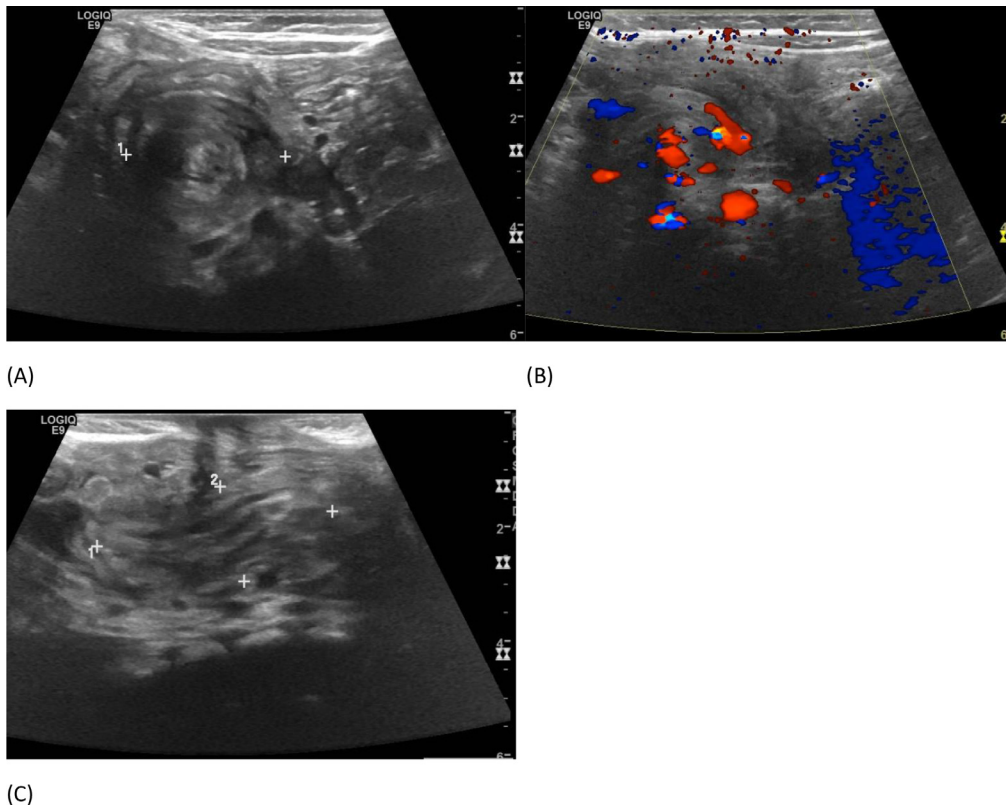


**Fig. 1 – Abdominal radiograph of a 2-month-old boy presenting for emesis and poor feeding with episodes of inconsolable crying demonstrated a paucity of distal bowel gas, concerning for obstruction. The differential diagnosis includes intussusception, malrotation with midgut volvulus, appendicitis, incarcerated hernia, and other surgical emergencies.**

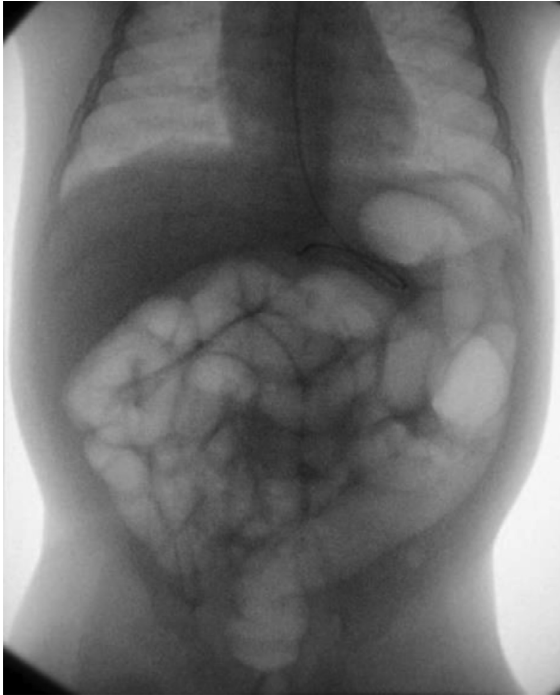
intussusception due to concentric layers of echogenic bowel. Other sonographic findings such as the “pseudokidney sign” of the intussusception in long view can also be deceiving. Normally, the SMV should always be to the patient’s right of the SMA, and the D3 segment of duodenum should be visualized retroperitoneally, posterior to the SMA. These relationships are useful in distinguishing intussusception from the more ominous midgut volvulus. [2]

**Case**

A 2-month-old boy presented to the emergency department for multiple episodes of yellow emesis during the day, poor feeding, decreased stools, and oliguria. The physical exam was benign including a lack of abdominal distention. A portable abdominal radiograph demonstrated a paucity of gas in the distal bowel concerning for small bowel obstruction. (Fig. 1) An abdominal ultrasound was performed by the ultrasound technologist and interpreted by a general radiologist, and was initially concerning for intussusception due to a reported “target sign.” (Fig. 2) A pediatric radiologist (with 15 years of experience) was called to assist with an air enema. An air enema was performed in the presence of a pediatric surgeon, and no intussusception was present, (Fig. 3) and the patient was admitted to the NICU for observation.



**Fig. 2 – Ultrasound of a 2-month-old boy presenting with bilious emesis demonstrated a 2.9 cm circular mass with concentric hyper echogenicity (A) with color Doppler flow (B) that was initially concerning for the “target sign” of intussusception. A longitudinal view of the intestines (C) demonstrated 4.2 x 1.7 cm of intestine concerning for the “pseudokidney sign.” (SMA and SMV not labeled.)**



**Fig. 3 – Normal fluoroscopic images from an air enema in anterior projection demonstrates an absence of intussuscepted bowel.**

The patient continued to have emesis that subsequently became bilious. A follow-up ultrasound was performed the same day by the pediatric radiologist that demonstrated the “whirlpool sign,” most concerning for malrotation with midgut volvulus. (Fig. 4) An upper gastrointestinal series was then performed that confirmed malrotation with midgut volvulus, (Fig. 5) and the patient was taken to the operating room for an urgent Ladd procedure. Intraoperatively, the bowel was found to be torsed 720 degrees, with a segment of mid-

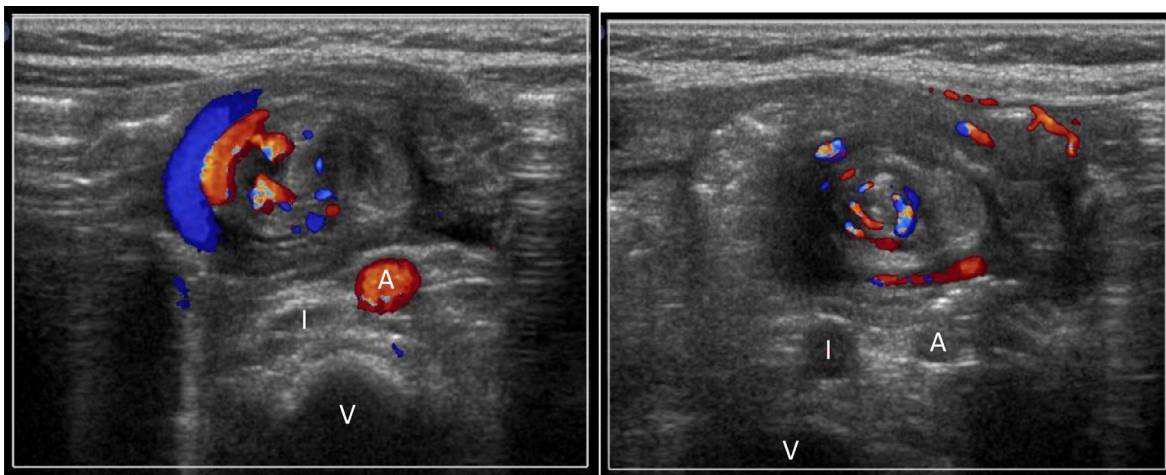
ileum tethered to the antimesenteric border of an adjacent loop of ileum consistent with an adhesion. The bowel was released and mobilized. No bowel necrosis was identified; therefore, resection was avoided. The remainder of the Ladd procedure was successful and the patient was discharged on post-operative day 5.

The patient returned to the emergency room on post-operative day 6 for recurrence of bilious emesis and poor feeding. A conventional abdominal radiograph demonstrated dilated loops of small bowel, (Fig. 6) and a follow-up abdominal ultrasound demonstrated abundant portal venous gas and pneumatosis intestinalis. (Fig. 7) Out of concern for bowel ischemia, the patient was taken to the operating room for an exploratory laparotomy. During the procedure, a 20cm portion of distal small bowel was noted to be torsed around itself (ie creating a closed loop obstruction), and anchored on the portion of mid-ileum which previously had been tethered by adhesions. These adhesions had been taken down during the prior Ladd procedure, leaving the surface of the antimesenteric ileum “raw” and “ragged” resulting in a nidus for this adhesion. The torsed bowel was mobilized with intra-operative evidence of reperfusion and no evidence of perforation. As such, no bowel resection was necessary. The bowel was oversewn to cover the “ragged” bowel serosa. The patient was discharged from the hospital without further complication.

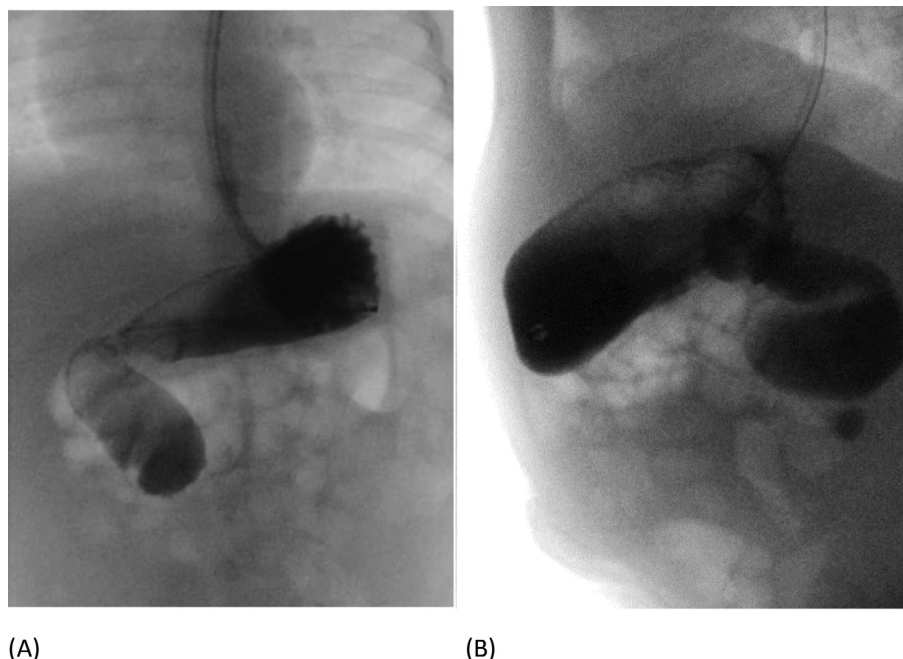
## Discussion

Malrotation with midgut volvulus is one of the few, true pediatric surgical emergencies. If clinical symptoms are consistent with volvulus, exploratory surgery should be considered. If the patient is stable, ultrasound demonstrating the SMA/ SMV relationship and D3 segment of bowel should be performed.

Abdominal radiographs are typically obtained as an initially screening exam, however, both intussusception and midgut volvulus can present with normal radiographs. Left lateral decubitus radiographs, however, should still be ob-



**Fig. 4 – Repeat abdominal ultrasound in the axial plane demonstrates a clockwise encircling of the SMV about the SMA within twisting mesentery, consistent with an ultrasonographic “whirlpool sign” of malrotation with midgut volvulus. The aorta “A,” IVC “I,” and vertebral body “V,” are seen posteriorly.**



**Fig. 5 – An upper gastrointestinal series shows a dilated D2 segment (A) with initial truncation of the duodenum followed by spiraling of the duodenum (B) demonstrating the “corkscrew sign” consistent with malrotation with midgut volvulus**



**Fig. 6 – An abdominal radiograph on post-operative day 7 from a Ladd procedure demonstrates dilated small bowel with numerous air-liquid levels greater than 2.5 cm in width, concerning for small bowel obstruction.**

tained prior to fluoroscopy as the presence of free intraperitoneal air necessitates urgent surgical intervention as opposed to fluoroscopic decompression.

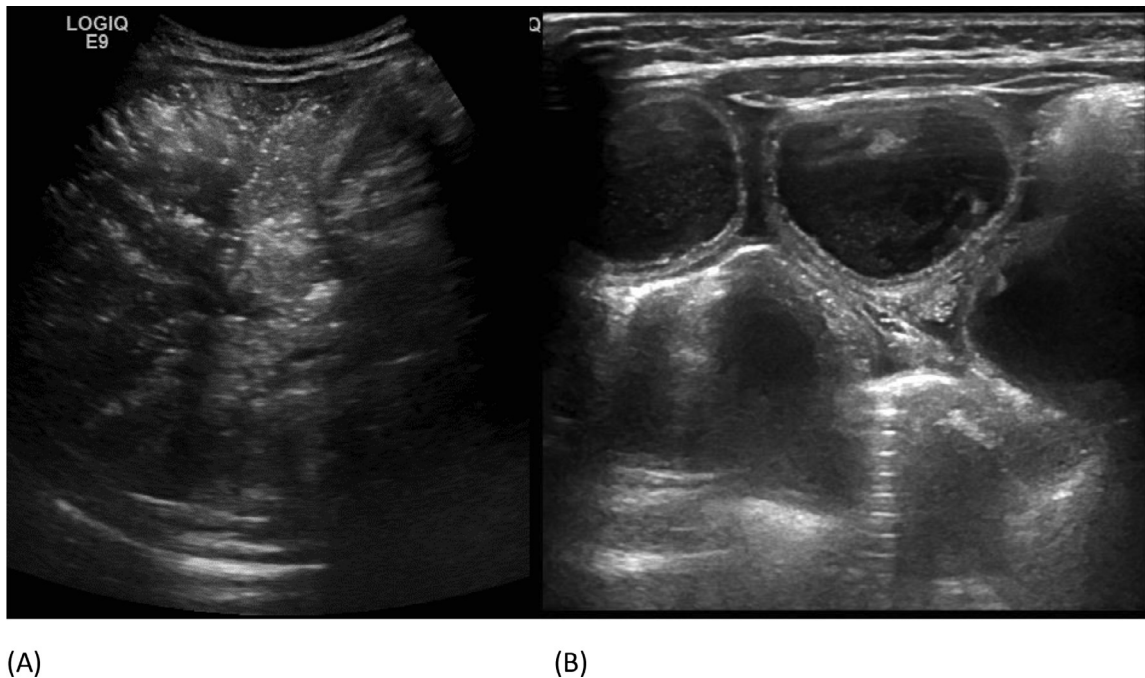
An upper gastrointestinal series (UGI) is the gold standard for detecting malrotation, with or without volvulus, with sensitivities of 93%-100%. [3,4] An UGI series may demonstrate a misplaced duodenum or ligament of Treitz (ex. to the right of midline), “corkscrew” duodenum (as in our case), or duodenal obstruction with bird beak appearance, with distal air. [3,5] A normal UGI with a high suspicion may warrant exploration.

Ultrasound (US) is a useful adjunct for detecting volvulus, with high sensitivity (92%-100%). [6,7] US with Doppler may demonstrate the “whirlpool sign” of a clockwise twisting SMV around SMA on Doppler transverse midline, as in our case.

This case highlights the importance of differentiating the “whirlpool sign” of volvulus from the “target sign” and “pseudokidney sign” of intussusception. When viewing the bowel in short axis, intussusception may demonstrate a “target sign,” or the “crescent-in-donut sign,” depending on the contribution from the hyperechoic mesentery. [8] At the proximal end, or base of the intussusciptions, the hyperechoic mesentery encloses the hypoechoic entering limb of intussusception, and appears as a crescent, creating the “crescent-in-donut sign.” [8] The apex of the intussusception does not have a contribution from mesentery, and demonstrates an edematous and hypoechoic intussusceptum that creates the “target sign” in axial plane. [8]

As the “target sign” and “crescent-in-donut sign” of intussusception and “whirlpool sign” of volvulus appear as circular masses with concentric rings of hyper and hypoechoic layers, the addition of Doppler is crucial to determine the diagnosis. There are previous reports of laparotomies being performed due to accidental confusion of an intussusception for





**Fig. 7 – An abdominal ultrasound demonstrates a significant burden of portal venous gas in the liver (A). Additional views show marked small intestinal dilation with pneumatosis intestinalis (B). Both findings were concerning for bowel ischemia**

volvulus based on ultrasound findings. [8] The finding of a clockwise swirling SMV around the SMA is specific to volvulus. An inverted SMA/SMV relationship may also be present on Doppler, also specific to volvulus, with sensitivity of 71%-100% and specificity of 79%-100%. [9,10]

In either condition, a longitudinal view of intestines should also be obtained. The “pseudokidney sign” refers to the reniform appearance of intussuscepted bowel when viewing the bowel in long axis, with the central hyperechoic mesentery of intussusceptum mimicking renal sinus fat. [8] The pseudokidney sign has been reported as 100% specific for intussusception. [11]

## Conclusion

This case is a reminder of the difficulties in definitive imaging diagnosis of two common pediatric emergencies, intussusception and malrotation with midgut volvulus, and how to avoid common pitfalls. In our case, the presence of a circular mass thought to represent the “target sign” as well as lack of documentation of SMA/SMV inversion led to the initial misdiagnosis of intussusception. The diagnosis was not made until a repeat ultrasound was performed by more experienced hands. The delay in diagnosis led to an hours-long delay of surgical care putting the child at further risk of bowel necrosis. This report highlights the importance of recognizing the imaging “signs” of malrotation with midgut volvulus, including reversal of the SMA and SMV, and an abnormal course of the D3 segment of duodenum. Given the high morbidity and mortality of midgut volvulus, there should be a low threshold to perform

an upper GI examination for confirmation of volvulus, particularly in the setting of an equivocal ultrasound. In the setting of suspected intussusception, a normal air enema should raise the question of an alternative diagnosis.

## Declaration

The views expressed in this manuscript are those of the authors, and do not reflect the official policy or position of the Department of the Army, Department of Defense, or the United States Government. The authors have no financial, personal, or other vested interests in the information contained within this document.

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