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# A novel plain abdominal radiograph sign to diagnose malrotation with volvulus

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#### ABSTRACT

Malrotation with associated volvulus is a potentially lethal event for a neonate. The gold standard for diagnosis is an upper gastrointestinal contrast study. However this can delay the diagnosis and the timing of surgical intervention. We present a novel abdominal radiographic sign; duodenal and gastric dilatation occurring in association with limited small bowel gas confined to the right lower quadrant of abdomen and the total absence of colonic air that is indicative of malrotation with associated volvulus. This allows for an earlier diagnosis and expeditious surgery.

# **CASE SERIES**

## INTRODUCTION

Malrotation of the small bowel is a common anomaly occurring in 1 in 500 live births (1). It is a congenital failure of the small bowel to complete the full 270° counter clockwise rotation around the superior mesenteric artery (SMA). This rotation, in normal fetal development, is achieved by three 90° counter clockwise rotations. The first occurs prior to 6 weeks of gestation when the duodenum rotates 90° counter clockwise and lies to the right of the SMA. After the 6th week the midgut herniates from the peritoneal cavity and the duodenum undergoes a second 90° counter clockwise rotation resulting in the duodenum lying posteriorly in relation to the SMA. Then between the 10th and 12th week of gestation the midgut returns to the peritoneal cavity and the duodenum undergoes the final 90° counter clockwise rotation so that the duodenojejunal flexure lies to the left of the midline.

Malrotation when associated with a volvulus can result in severe consequences such as short bowel syndrome, infarction requiring bowel resection or even death. Paediatric surgical trainees are taught to beware the neonate or infant with green-coloured vomitus, and intestinal malrotation should always be excluded if there are concerns (2). The prompt investigation and surgical intervention of these neonates and infants is vital

to ensure a favourable outcome. The investigation of choice of a possible malrotation with associated volvulus in a stable neonate is the upper gastrointestinal tract contrast study (3). This can however potentially lead to a delay in surgical intervention and therefore a worse outcome. In this article we present a novel combination of abdominal radiographic features, namely; duodenal and gastric dilatation occurring in association with limited small bowel gas confined to the right lower quadrant of abdomen and the total absence of colonic air, which in newborn neonates indicates a malrotation with volvulus and may negate the need for further investigations.

#### CASE DESCRIPTIONS

The first patient presented within a few hours of birth with bile stained vomiting to the neonatal unit. The neonate was delivered by normal vaginal delivery, required no post-natal resuscitation, had normal APGAR scores and was also a term-delivery. The initial clinical assessment revealed a well neonate with a non-distended and non-tender abdomen. Initial serological investigations revealed no abnormalities and no acidosis or raised lactate levels. The initial plain abdominal radiograph (Figure 1) confirmed duodenal dilatation (arrow 1),

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gas in proximal small bowel confined to the right of the midline (arrow 2) and absent colonic air.

The second patient was also a term delivery, delivered in a good condition with a normal postnatal examination and serological investigations. There was an antenatal diagnosis of possible small bowel obstruction and the immediate placement of a nasogastric tube confirmed bilious aspirates. The antenatal ultrasound report but not the images were available for review. The postnatal plain abdominal radiograph (Figure 2) confirmed the duodenal dilatation with gas in small bowel confined to right hand side. No colonic gas was evident. The transverse soft tissue shadow lying to the left of the midline (arrowed) is the umbilical cord stump.

The third patient presented in a similar fashion to the first neonate. The initial plain abdominal radiograph revealed the same combination of radiological features (Figure 3). In this case, however, the proximal small bowel dilatation was more impressive than the other two cases.

Surgical intervention, in all of the three neonates, confirmed the diagnosis of malrotation with volvulus and a successful Ladd's procedure was performed. In all the patients the laparotomy revealed viable bowel and there was no indication for either bowel resection or stoma formation. All of the patients underwent an uneventful post-operative course and full enteral feeds were established with a discharge from the neonatal unit within the first week of life.

#### DISCUSSION

Malrotation with associated volvulus can be a catastrophic event for a newborn neonate. The investigation of choice, in a stable infant is an upper gastrointestinal tract contrast study (3). A normal duodenojejunal flexure (DJF) is seen to the left of the spine and at the level of the pylorus (Figure 4). Abnormalities seen with malrotation include; an abnormally placed DJF, spiral or "corkscrew" distal duodenum, and proximal jejunum located on the right in the abdominal cavity (Figure 5) (4).

There are various other conditions that should be considered on an abdominal radiograph when managing a neonate who presents with bilious vomiting and duodenal obstruction. These include duodenal atresia, the presence of a duodenal web and also an annular pancreas.

In an otherwise well child, duodenal atresia has a typical abdominal radiographic sign, the "double bubble". This appearance is secondary to a distended first part of duodenum and an air filled stomach. The absence of air distal to the second part of duodenum distinguishes this condition from a malrotation with volvulus or other conditions causing a partial duodenal obstruction (Figure 6).

The duodenal lumen may also be partially obstructed by the presence of a duodenal web, diaphragm or intraluminal diverticulum. This results in the appearance of a dilated first part of the duodenum, distal air in the jejunum and a normally placed duodenojejunal flexure (Figure 7). The distal jejunal air would have a normal distribution compared to the confinement to the right in our series of neonates. A duodenal web may also be demonstrated with an upper gastrointestinal contrast study. The abnormal duodenum can be demonstrated with a dilated 1st and 2nd part, the stomach may also be dilated but there will be normal orientation of the rest of the small bowel and colon (Figure 8).

Other rare causes of complete or incomplete duodenal obstruction include the presence of an annular pancreas and duodenal stenosis. With an annular pancreas, the second part of the duodenum is obstructed by a ring of pancreatic tissue arising from the head of the pancreas. This ring of tissue encircles the duodenum either completely or partially resulting in variable degrees of obstruction. Duodenal stenosis usually has a delayed presentation with episodes of aspiration, vomiting and failure to thrive.

There have been recent advances in ultrasound diagnosis and features of malrotation include an abnormal relationship of the superior mesenteric artery and vein and the classic whirlpool sign of mesenteric pedicle twisting (5). The use of this specialised technique remains, however, operator dependent. It is therefore not feasible as a routine method of investigation even in some tertiary paediatric surgical centres.

There are various plain radiograph signs that have been described in the literature for a paediatric malrotation with associated volvulus. These include a gasless abdomen, a "whirled" appearance secondary to volvulus, separation of adjacent bowel loops with "tubular" appearance secondary to ischaemia and also a normal gas pattern (4,6-8). It has also been suggested that there is no clinical value of a plain abdominal radiograph (8). We have, on the contrary, presented in this article 3 cases of neonates who presented with signs of intestinal obstruction with operative finding of malrotation with volvulus. All the supine radiographs were taken within the first 24 hours of life. Consistent features in all of these were; duodenal and gastric dilatation occurring in association with limited small bowel gas confined to the right lower quadrant of abdomen and the total absence of colonic air. These three neonates were subjected to a laparotomy and were found to have malrotation with volvulus but with viable bowel. All underwent a successful Ladd's procedure which involved, a derotation of the bowel, division of Ladd's bands, widening of the vascular pedicle and appendicectomy. In addition, intubation of the duodenum was carried out to exclude a web.

This peculiar radiological appearance is secondary to a malrotation with associated volvulus causing a partial duodenal obstruction leading to proximal dilatation. Duodenal distension (as noted in figures 1-3) is variable and dependant on the longevity and the degree of obstruction. Irrespective of this, duodenal occlusion would appear to be partial, and a small amount of air does pass through into the small bowel which is visible to the right of the midline. On initial post natal abdominal X ray no air appears to pass distally into the colon hence its imperceptibility. As the air makes its way into the colon it is likely that this feature would transform over time.

This constellation of radiographic features has hitherto not been described and in combination indicates a volvulus. This may be an indication for immediate laparotomy and a Ladd's procedure without requirement for an upper gastrointestinal contrast study. There is a need, however, for a larger investigation of neonates who present with bilious vomiting to determine the sensitivity and specificity of this constellation of radiographic features in malrotation with volvulus. Although the absence of this sign does not exclude malrotation with volvulus and its associated sequalae, its presence should ensure there is no delay in surgical intervention by avoiding unnecessary investigations with the associated hazard of radiation exposure.

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# CONCLUSION

Combination of duodenal dilatation, gas in proximal small bowel confined to the right of the midline and absent colonic air on an initial plain abdominal radiograph of a neonate aids expeditious diagnosis of malrotation with volvulus. This allows earlier surgical intervention and an improved outcome. A larger series is required to ascertain the sensitivity and specificity of this sign. For the meantime, for all suspected cases of malrotation the gold standard of radiological investigation remains the upper gastrointestinal contrast study.

# TEACHING POINT

Malrotation with associated volvulus is a potentially lethal event for a newborn neonate presenting with bilious vomiting. An abnormally placed duodeno-jejunal flexure demonstrated on an upper gastrointestinal contrast study, or the presence of duodenal and gastric dilatation with limited small bowel gas confined to the right lower quadrant of abdomen and the total absence of colonic air on an early post natal X ray is indicative of malrotation with volvulus and requires prompt diagnosis and treatment.

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#### **FIGURES**



**Figure 1:** Initial AP abdominal radiograph of 1 day old male neonate presenting within a few hours of birth with bile stained vomiting. An initial radiograph confirmed duodenal dilatation (arrow 1), gas in proximal small bowel confined to the right of the midline (arrow 2) and absent colonic air.

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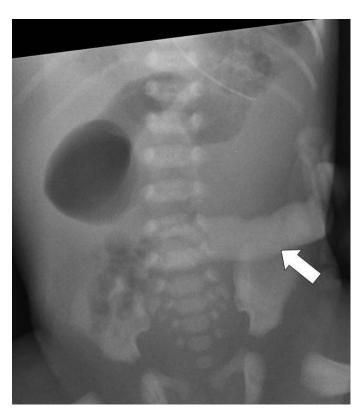


Figure 2: AP abdominal radiograph of 1 day old female neonate with ultrasound antenatal diagnosis of possible small bowel obstruction. Duodenal dilatation is noted with gas in small bowel confined to right hand side. No colonic gas was evident. The transverse soft tissue shadow lying to the left of the midline (arrowed) is the umbilical cord stump.

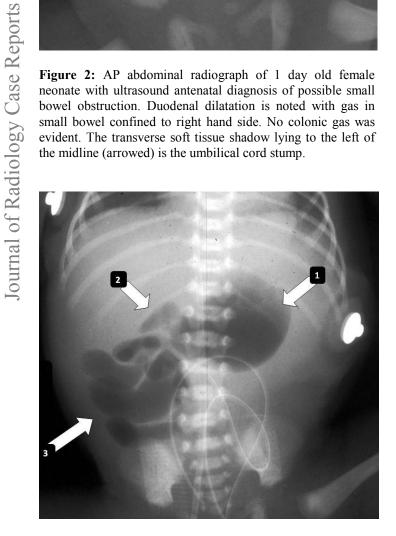


Figure 3: AP abdominal radiograph of a 1 day old male neonate who presented with bile stained vomiting. Gastric (arrow 1) and some duodenal dilatation (arrow 2) was revealed with more impressive small bowel shadows confined to the right side of the abdomen (arrow 3). There is an absence of distal small bowel or colonic air.

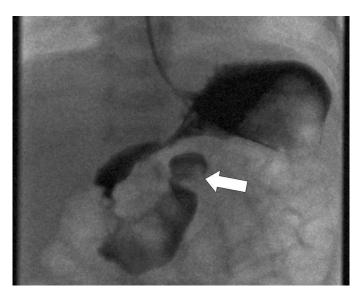
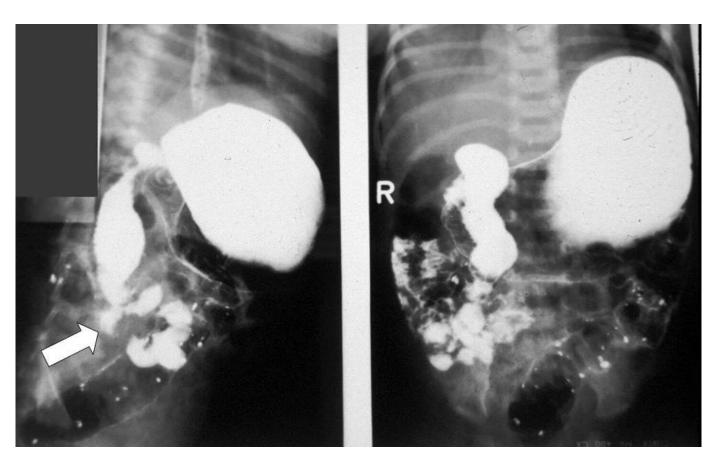
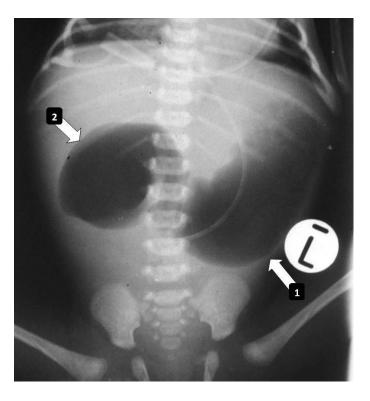


Figure 4: Abdominal upper gastrointestinal Gastrograffin contrast study of a three day old term male neonate who presented with bilious vomiting. A normal duodenojejunal flexure (DJF) is demonstrated on the left of the spine and at the level of the pylorus (arrow). The classic "C" shape of the duodenum is also demonstrated.



**Figure 5:** Upper gastrointestinal barium contrast study in a 4 day old female neonate who presented with bilious vomiting with associated abdominal distension demonstrating features of malrotation with volvulus. The lateral view on the left demonstrates the spiral configuration to the bowel just distal to the second part of duodenum secondary to the volvulus (arrow). The AP view on the right shows the proximal small bowel to be lying to the right of the midline. The classic "C" of duodenum with the DJ flexure lying to the left of the midline is not seen.



**Figure 6 (left):** Abdominal radiograph of a 1 day old term male neonate who presented with bilious vomiting secondary to a duodenal atresia. The "double bubble" is seen with a distended first part of duodenum (arrow 1) and air filled stomach (arrow 2). The absence of air distal to the second part of duodenum distinguishes this condition from a malrotation with volvulus.

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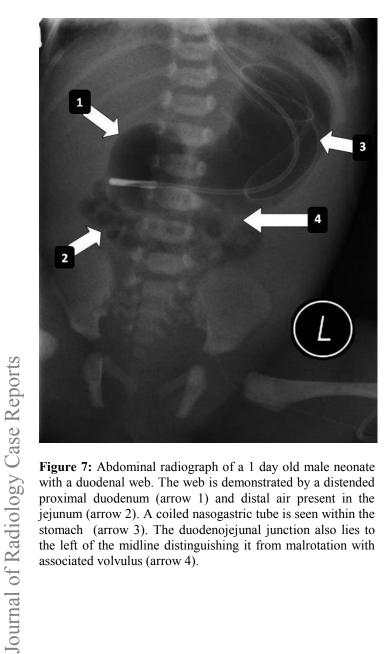
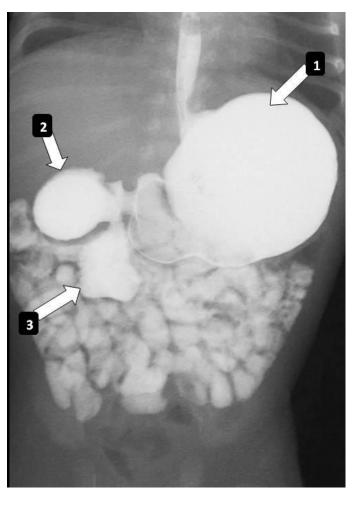


Figure 7: Abdominal radiograph of a 1 day old male neonate with a duodenal web. The web is demonstrated by a distended proximal duodenum (arrow 1) and distal air present in the jejunum (arrow 2). A coiled nasogastric tube is seen within the stomach (arrow 3). The duodenojejunal junction also lies to the left of the midline distinguishing it from malrotation with associated volvulus (arrow 4).



**Figure 8:** Upper gastrointestinal Niopam 360 contrast study in a 1 day male old neonate with a duodenal web. There is an abnormal duodenum demonstrated but with normal orientation of the rest of the small bowel and colon. The duodenal web is demonstrated by a distended stomach (arrow 1), 1st and 2nd (arrow 2 & 3) parts of the duodenum.

#### **ABBREVIATIONS**

DJF = duodenojejunal flexure SMA = superior mesenteric artery

## **KEYWORDS**

Malrotation, volvulus, abdominal radiograph

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