Comparative Effectiveness of Imaging Modalities for the Diagnosis of Intestinal Obstruction in Neonates and Infants: A Critically Appraised Topic

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Abbreviations and Acronyms

MR magnetic resonance
PICO patient, intervention, comparison, outcome
MI meconium ileus
US ultrasound

Rationale and Objectives: The purpose of this study was to critically appraise and compare the diagnostic performance of imaging modalities that are used for the diagnosis of upper and lower gastrointestinal (GI) tract obstruction in neonates and infants.

Methods: A focused clinical question was constructed and the literature was searched using the patient, intervention, comparison, outcome method comparing radiography, upper GI contrast study, and ultrasound in the detection of upper GI tract obstruction such as duodenal atresia and stenosis, jejunal and ileal atresia, and malrotation and volvulus. The same methods were used to compare radiography and contrast enema in the detection of lower GI tract obstruction such as meconium plug syndrome, meconium ileus, Hirschsprung disease, and imperforate anus. Retrieved articles were appraised and assigned a level of evidence based on the Oxford University Centre for Evidence-Based Medicine hierarchy of validity for diagnostic studies.

Results: There were no sensitivities/specificities available for the imaging diagnosis of duodenal atresia or stenosis, jejunal or ileal atresias, meconium plug, and meconium ileus or for the use of cross-table lateral radiography for the diagnosis of rectal pouch distance from skin in imperforate anus. The retrieved sensitivity for the detection of malrotation on upper GI contrast study is 96%, and the sensitivity for the diagnosis of midgut volvulus on upper GI contrast study is 79%. The retrieved sensitivity and specificity for the detection of malrotation with volvulus on ultrasound were 89% and 92%, respectively. The retrieved sensitivity and specificity for the detection of Hirschsprung disease on contrast enema were 70% and 83%, respectively. The retrieved sensitivity of invertogram for the diagnosis of rectal pouch distance from skin in imperforate anus is 27%. The retrieved sensitivities of perineal ultrasound and colostography for the diagnosis of rectal pouch distance from skin in imperforate anus were 86% and 100%, respectively.

Conclusions: There is limited evidence for the imaging diagnosis of duodenal atresia and stenosis, jejunal and ileal atresias, meconium plug, meconium ileus, and imperforate anus, with recommended practice based mainly on low-quality evidence or expert opinion. The available evidence supports the use of upper GI contrast study for the diagnosis of malrotation and volvulus, with ultrasound as an adjunct to diagnosis. Contrast enema is useful in the investigation of suspected Hirschsprung disease, but a negative study does not rule out the condition. Colostography is the investigation of choice for the work-up of infants with complex anorectal malformations before definitive surgical repair.

Key Words: Evidence-based medicine; upper gastrointestinal tract; lower gastrointestinal tract; small bowel obstruction; large bowel obstruction; diagnosis; sensitivity and specificity; confidence interval; duodenal atresia; duodenal stenosis; jejunal atresia; ileal atresia; malrotation; volvulus; meconium plug syndrome; meconium ileus; Hirschsprung disease; imperforate anus.

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INTRODUCTION

Duodenal atresia occurs in 1 in 5000–10,000 live births. In one-third of cases, it is an isolated anomaly. In one-third of cases, it is associated with Down syndrome. It can also be associated with other congenital abnormalities, especially cardiac and esophageal anomalies. It results from failure of the duodenum to recanalize during weeks 8–10 of gestation (1). Neonates typically present with bilious or nonbilious vomiting in the first 24 hours after birth.

Infants with less severe duodenal stenosis may have delayed presentation with frequent vomiting and failure to thrive. Duodenal stenosis is often due to the presence of a duodenal web or may be secondary to extrinsic compression from an annular pancreas, malrotation, or a preduodenal portal vein (2).

Similarly, the even rarer jejunal and ileal atresias (3) also present with bilious vomiting and abdominal distension, usually in the first 24 hours after birth. They are thought to result from a vascular incident causing intestinal necrosis in utero and are less commonly diagnosed antenatally than duodenal atresia.

Malrotation is a rotational anomaly of the embryonic gut that leads to abnormal positioning of the duodenal-jejunal (D-J) junction. The D-J junction is not in its normal position to the left of the left-sided vertebral pedicles at the level of the duodenal bulb on the frontal view and posterior (retroperitoneal) on the lateral view of an upper gastrointestinal (GI) contrast study (4). Often, the small bowel is located in the right side of the abdomen and the colon including the cecum is on the left side of the abdomen. The position of the cecum can be variable and may be subhepatic or in the midline (5).

Small bowel volvulus is a complication of malrotation, where the abnormally narrow base or pedicle of the small bowel mesentery becomes twisted around itself, leading to obstruction at the level of the duodenum, followed by intestinal ischemia if prompt surgical treatment is not provided. It should be suspected in any infant with bilious vomiting. Physical examination may demonstrate diffuse abdominal tenderness with or without signs of peritonitis, abdominal distension, and blood in the stool on rectal examination (a late sign suggestive of bowel necrosis) (6). Delayed treatment is associated with high mortality rates or severe morbidity secondary to short gut syndrome if the child survives (5).

Malrotation occurs in approximately 1 in 500 live births (7). Although a previously undiagnosed patient with malrotation may present at any age with an acute volvulus, approximately 80% of patients with malrotation will present in the first month of life (7,8). Of those presenting in the first month of life, most will do so in the first week (9). Some series report that 10–20% present after 1 year of age (4,10–12).

Meconium plug syndrome is a relatively benign cause of large bowel obstruction in neonates who have delayed passage of meconium in the first 24–72 hours of life. Although it can resolve spontaneously, a contrast enema is often required. It is associated with small left colon syndrome, which also usually resolves after contrast enema. Most of the time, there are no further problems. An association with Hirschsprung disease has been reported in 13–38% of cases (13,14).

Meconium ileus (MI) is a common presentation of cystic fibrosis (CF) in the neonatal period. MI is the presenting feature in approximately 10% of patients with CF, and 80–90% of patients who have MI are subsequently diagnosed with CF (15). Infants present with bowel obstruction and failure to pass meconium in the first 3 days of life.

Hirschsprung disease is a functional obstruction of the bowel caused by the absence of intrinsic enteric ganglion cells. It usually presents with failure to pass meconium or delayed passage of meconium in the newborn period. Infants may also present with abdominal distension and bilious vomiting. It may involve variable lengths of colon extending back from the rectum, and rarely can involve the entire colon.

Anorectal malformations/imperforate anus comprise a wide spectrum of anomalies of the anus and rectum and often also of the urinary tract and genitalia. They can vary from simple malformations, which are easily corrected with good functional outcomes, to complex anomalies requiring multiple complex surgeries with poorer outcomes. Imperforate anus is most commonly associated with the VACTERL association (vertebral anomalies, anal atresia, cardiac defects, tracheoesophageal fistula and/or esophageal atresia, renal and radial anomalies, limb defects), and occurs with increased frequency in trisomy 21 (16). The mainstay of diagnosis of anorectal malformations is from physical examination of the neonate. At 16–36 hours post birth, neonates are assessed for the presence of a fistula to the perineum or to the urinary or genital tracts and for associated anomalies. In the majority of patients, a colostomy will be performed and further workup will be arranged after the colostomy has healed. However, in certain cases such as rectoperineal fistula or rectovestibular fistula in girls, colostomy can be avoided and anoplasty is all that is needed. Ultrasound and urine analysis are used to rule out obstructive uropathy, hydrocolpos, or rectovesical fistula.

ASK

We questioned whether radiography, upper GI contrast study, or ultrasound perform better for the detection of upper GI tract obstruction and for the diagnosis of duodenal atresia and stenosis, jejunal and ileal atresia, and malrotation and volvulus. Before undertaking this review using evidence-based methods, we note the prevailing popular opinion (before using explicit critical appraisal) that plain radiography is sufficient for the diagnosis of duodenal, jejunal, and ileal atresias, whereas upper GI contrast study is the investigation of choice for malrotation and volvulus.

Similarly, we questioned whether radiography or contrast enema has a better diagnostic performance for the detection of meconium plug, MI, Hirschsprung disease, and imperforate anus (the common causes of lower GI tract obstruction in neonates).

We constructed a focused standardized patient, investigation, comparison, and outcome question to search the available literature as follows: (1) “In neonates and infants with
suspected upper GI tract obstruction, how does radiography vs. upper GI contrast study vs. ultrasound compare to each other for detection of upper GI tract obstruction and diagnosis?” and (2) “In neonates and infants with suspected lower GI tract obstruction, how does radiography vs. contrast enema compare to each other for detection and diagnosis?”

SEARCH

Secondary and primary evidence were searched according to the evidence pyramid described by Haynes (17). For secondary literature (eg, information systems, synopses, and syntheses) (18), we searched Up To Date (19), The American College of Physicians Journal Club (20), Cochrane Collaboration Library (21), Turning Research Into Practice (22), and PubMed (23). For primary literature (original studies), we searched PubMed (23) and EMBASE (24).

A comprehensive Medline search (United States National Library of Medicine database) for original articles published between May 2005 and May 2015 using the Ovid and PubMed search engines was performed using a combination of the following key terms: (“diagnosis”) AND (“small bowel obstruction”) AND (radiography OR upper GI contrast study OR ultrasound)) and (“diagnosis”) AND (“large bowel obstruction”) AND (radiography OR contrast enema)) and (“diagnosis”) AND (“duodenal atresia” OR “jejunal atresia” OR “ileal atresia” OR “malrotation and volvulus”) OR “meconium plug” OR “meconium ileus” OR “Hirschsprung”) AND (“evidence based”) AND “imperforate anus” OR “anorectal malformation” AND (“diagnosis”) AND “radiograph” OR “colostomy” OR “colostogram” OR “magnetic resonance imaging.” (Fig 1).

The search was limited to English-language articles, human, and pediatric studies. The abstracts were reviewed and selected based on well-designed methodology, clinical trials, outcomes, and diagnostic accuracy. Additional relevant articles were selected from the references of reviewed articles and published guidelines.

APPRAISE

Upper GI Tract

The evidence base for the imaging of intestinal obstruction in the pediatric population is limited. Computed tomography is generally avoided because of concerns regarding radiation exposure. Radiography, contrast studies, and ultrasound are commonly used to assist with diagnosis.

Neonates and young children with intestinal obstruction deteriorate rapidly, and prompt clinical and radiological detection of obstruction is essential to reduce morbidity and mortality. Before radiological investigation and treatment, it is essential that children are adequately resuscitated and stabilized (25). Children who are medically unstable and in whom there is a high clinical suspicion of malrotation with volvulus should proceed directly to surgery (6).

Duodenal Atresia and Stenosis, Jejunal, and Ileal Atresia

In many cases of congenital intestinal atresia and obstruction, the diagnosis is made on antenatal ultrasound (26–28). Patients may present with polyhydramnios due to fetal bowel atresias. Findings such as the “double bubble sign” of gastric and duodenal dilatation suggest the diagnosis of duodenal atresia (1). For the purpose of this study, we will focus on the proven diagnostic modalities used after birth, which include plain radiography and upper GI contrast study.

Radiography.—On searching the primary literature, we found a retrospective observational study entitled “Congenital Duodenal Obstruction in Neonates: A Decade’s Experience From One Center” by Chen et al. This was assigned level 4 evidence using the Oxford Center for Evidence-Based Medicine (29). The evidence regarding the diagnosis of duodenal, jejunal, and ileal atresias is extremely limited and the literature is predominantly expert opinion (level 5 evidence). No studies provided a sensitivity or specificity for the use of imaging in the diagnosis of duodenal atresia/stenosis or jejunal or ileal atresia.

The mainstay of diagnosis of duodenal atresia and stenosis in a neonate is radiography showing the “double bubble sign” of gastric and proximal duodenal dilatation and an absence of gas within the distal bowel (30). This sign was demonstrated in 68.6% of patients presenting with congenital duodenal obstruction in one retrospective observational study (31). In partial duodenal obstruction or duodenal stenosis, an air-fluid level in the stomach may be demonstrated.

Similarly, the even rarer jejunal and ileal atresias are usually first identified on radiograph showing dilated air and fluid-filled bowel loops down to mid or distal small bowel level with no gas beyond the level of obstruction or in the colon.

All infants with intestinal obstruction should have a supine anteroposterior abdominal radiograph and a lateral decubitus radiograph to rule pneumoperitoneum (1).

Upper GI contrast study.—It is recommended that all medically stable suspected cases of duodenal atresia are followed up with an upper GI contrast study to rule malrotation and volvulus masquerading as a double bubble sign (1). In infants who have had the stomach decompressed either through vomiting or through aspiration of a nasogastric tube, air can be injected down the nasogastric tube to act as its own contrast agent and reproduce the double bubble sign. Barium is useful to determine if the patient has duodenal atresia or duodenal stenosis.

Upper GI contrast study with barium may be useful in cases of jejunal or ileal atresia to differentiate atresia from stenosis.

Contrast enema.—It is recommended that all medically stable suspected cases of jejunal and ileal atresia are followed up with a contrast enema to differentiate between MI or Hirschsprung disease and intestinal atresia. All three pathologies may have an associated microcolon. In cases of atresia, the contrast does not reflux back into the dilated proximal bowel loops around the inspissated stool (1).
Malrotation and Volvulus

Radiography.—A plain radiograph is often performed as a first-line investigation to rule out gross pneumoperitoneum or obvious lower GI obstruction as an alternative diagnosis. The most common finding in malrotation and volvulus is a gasless abdomen, but occasionally a double bubble sign (like in duodenal atresia) is demonstrated (6).

Upper GI contrast study.—On searching the secondary literature, we found a study entitled “Intestinal Malrotation” on www.uptodate.com. This recommends upper GI contrast study as the investigation of choice for children with suspected malrotation and volvulus, based on a retrospective cohort study using surgical findings as the gold standard (32). This was assigned level 3b evidence using the Oxford Center for Evidence-Based Medicine (29). In this study, the sensitivity of the upper GI contrast study for the diagnosis of malrotation was 96% and the sensitivity for the diagnosis of midgut volvulus was 79% (32). The study looked only at patients who went to surgery for suspected malrotation and/or volvulus; therefore, negative studies that did not go to surgery were not assessed and an accurate specificity could not be obtained.

Findings at upper GI contrast study in malrotation include malposition of the duodenum (D-J junction is not in its normal position to the left of the left-sided vertebral pedicles at the level of the duodenal bulb on the frontal view and posterior on the lateral view) and duodenal obstruction similar to the appearance in duodenal atresia. In acute volvulus, a beak sign

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of duodenal obstruction is often seen, whereas the cork-screw sign of the twisted bowel may or may not be demonstrated.

In up to 25% of cases, the findings at upper GI contrast study can be subtle or equivocal (6). In these cases, contrast enema or ultrasound may be useful as an adjunct to assist in making the diagnosis.

**Contrast enema.**—If the study is inconclusive, a contrast enema may be performed to confirm the position of the cecum, aiding the diagnosis of malrotation (33). The position of the cecum is variable and therefore contrast enema cannot exclude malrotation. If the imaging findings are inconclusive, but there is a strong suspicion of volvulus, the patient should proceed to exploratory surgery (11).

**Ultrasound.**—There is emerging evidence of the usefulness of ultrasound in detecting malrotation and small bowel volvulus, by demonstrating abnormal orientation of the superior mesenteric vessels.

On searching the secondary literature, we found a study entitled “Sonographic Features Related to Volvulus in Neonatal Intestinal Malrotation” by Chao et al. This was assigned level 2b evidence using the Oxford Center for Evidence-Based Medicine (29).

In Chao et al.’a 3-year prospective study of abdominal and color Doppler ultrasonography in neonates with suspected malrotation or malrotation and volvulus, water instillation was used to detect duodenal dilatation, edema, and malrotated small bowel. They found that duodenal dilatation with tapering configuration, fixed midline bowel, and mesenteric whirlpool sign were all suggestive of volvulus. The sensitivity and specificity of duodenal dilatation with tapering configuration for the detection of volvulus were 89% and 92%, respectively (Table 1 and Fig 2a); of fixed midline bowel were 89% and 92% (Fig 2b); of whirlpool sign were 89% and 92% (Fig 2c); and of dilatation of distal superior mesenteric vein were 56% and 73% (Fig 2d) (34). Other studies have used ultrasound to look at the position of the superior mesenteric artery in malrotation. However, a normally placed superior mesenteric vein does not rule out malrotation (35,36).

**Lower GI Tract**

In a vomiting neonate, the initial abdominal radiograph allows differentiation between upper and lower GI causes and guides subsequent investigations, with contrast enema as the investigation of choice for lower GI pathology.

**Meconium Plug Syndrome and MI**

Although prenatal diagnosis of MI is often suggested by ultrasound demonstrating hypercohoic bowel, the definitive diagnosis is made after birth, as echogenic bowel can be a normal transient occurrence in utero (37).

**Radiography.**—A plain radiograph is usually the first investigation in the work-up of both meconium plug and MI.

In meconium plug, plain radiography shows inspissated meconium pellets in the large bowel only (14).

In MI, a plain radiograph demonstrates dilated small bowel loops with the inspissated meconium in the distal ileum and the large bowel. Plain radiography is necessary to assess for perforation, calcifications (evidence of prenatal perforation and meconium peritonitis), or other abnormalities. Evidence of perforation indicates complex MI and precludes an attempt at nonoperative decompression. In this case, the infant should proceed directly to surgery.

**Contrast enema.**—Water-soluble contrast enemas with hyposmolar contrast have long been established as the diagnostic modality of choice for meconium plug (hypoplastic left colon syndrome) and also for MI (38). Gastrografin is most commonly used. It is meglumine diatrizoate, a hyperosmolar, water-soluble, radiopaque solution containing 0.1% polysorbate 80 (Tween 80) and 37% organically bound iodine. The osmolarity of the solution is 1900 mOsm/L. Upon instillation, fluid is drawn into the intestinal lumen, hydrating and softening the meconium mass (37). Aside from demonstrating the level of obstruction and outlining the meconium, they also have the therapeutic effect of often relieving the obstruction and avoiding the need for surgery (39).

In patients with MI, the contrast enema typically reveals a small-caliber colon (microcolon of disuse) and meconium pellets in the terminal ileum. The ileum proximal to the obstruction is dilated. For successful decompression, contrast should be seen refluxing into the terminal ileum. Follow-up

<table>
<thead>
<tr>
<th>TABLE 1. Test Performances for the Diagnosis of Upper and Lower GI Tract Obstruction in Neonates and Infants</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Upper Gastrointestinal (GI) Tract</strong></td>
</tr>
<tr>
<td>Selected Study</td>
</tr>
<tr>
<td>Sensitivity, %</td>
</tr>
<tr>
<td>Specificity, %</td>
</tr>
<tr>
<td>Accuracy</td>
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</tbody>
</table>

SMV, superior mesenteric vein; US, ultrasound.
Figure 2. Graph of conditional probabilities for ultrasound diagnosis of upper gastrointestinal (GI) obstruction in neonates and infants. Graph of conditional probability and area under the curve (AUC). The x-axis represents the pretest probability of the disease in question, and the y-axis represents the posttest probability. The pretest probability is chosen, and a vertical line is drawn from the corresponding point on the x-axis to the “test-negative” or “test-positive” curve. From this point of intersection, a horizontal line is drawn to the y-axis. The posttest probability is the value at the point where the horizontal line intersects the y-axis. The posttest probability is the value at the point where the horizontal line intersects the y-axis. (a) Graph of conditional probabilities for ultrasound diagnosis of volvulus in upper GI obstruction in neonates and infants. Sensitivity = 0.89; specificity = 0.92; and AUC = 0.81. (b) Graph of conditional probabilities for ultrasound detection of fixed midline bowel in upper GI obstruction in neonates and infants. Sensitivity = 0.89; specificity = 0.92; and AUC = 0.81. (c) Graph of conditional probabilities for ultrasound detection of the whirlpool sign in upper GI obstruction in neonates and infants. Sensitivity = 0.89; specificity = 0.92; and AUC = 0.81. As we can see from the above three graphs (a–c), the high sensitivity and specificity means that ultrasound is reliable in ruling in and ruling out volvulus and can reliably detect the presence or absence of a fixed midline bowel and the “whirlpool” sign. (d) Graph of conditional probabilities for ultrasound detection of superior mesenteric vein dilatation in upper GI obstruction in neonates and infants. Sensitivity = 0.56; specificity = 0.73; and AUC = 0.60. As we can see from this graph, the available evidence suggests that ultrasound cannot be reliably used to assess the presence or absence of superior mesenteric vein (SMV) dilatation in the setting of upper GI obstruction in neonates and infants.
Contrast Enema for Diagnosis of Hirschsprung’s Disease in Neonates and Infants Lower GI Obstruction

Figure 3. Graph of conditional probabilities for contrast enema for the diagnosis of Hirschsprung disease in lower gastrointestinal (GI) obstruction in neonates and infants. Sensitivity = 0.70; specificity = 0.83; area under the curve (AUC) = 0.70. Graph of conditional probability and AUC. The x-axis represents the pretest probability of the disease in question, and the y-axis represents the posttest probability. The pretest probability is chosen, and a vertical line is drawn from the corresponding point on the x-axis to the “test-negative” or “test-positive” curve. From this point of intersection, a horizontal line is drawn to the y-axis. The posttest probability is the value at the point where the horizontal line intersects the y-axis. As we can see from this graph, contrast enema is reliable in patients with a high pretest probability and a positive test. It is not so reliable in patients with a high pretest probability and a negative test. These patients will require further investigation.

Radiographs are advised to outrule perforation as a complication and to assess for resolution of the obstruction. The enema can be repeated once or twice at 12–24 hour intervals if necessary to completely relieve the obstruction.

On searching the secondary literature, we found a study entitled “Pulling the Plug”—Management of Meconium Plug Syndrome in Neonates” by Cuenca et al. (40). This was assigned level 4 evidence using the Oxford Center for Evidence-Based Medicine (29). In their study, 30% of patients had spontaneous resolution of the meconium plug without any specific treatment. Of those infants requiring treatment, barium enema was successful in 97% and only one enema was required in 98% of successful cases. On searching the primary and secondary literature, we found no studies of sensitivity or specificity for contrast enema in the diagnosis of meconium plug.

The success rate of patients with uncomplicated MI, treated with Gastrografin enemas, historically range as high as 83% (37). Several more recent studies report much lower success rates closer to 36–39% (39,41), with corresponding lower complication rates, possibly due to a more conservative approach in terms of number of enemas performed per patient and pressures used. However, these studies are of small size and are prone to bias. Operative treatment is associated with significant morbidity and mortality (41). On searching the secondary literature, we found no studies of sensitivity or specificity for contrast enema in the diagnosis of MI.

Hirschsprung Disease

Contrast enema.—Contrast enema is the imaging modality of choice for the diagnosis of Hirschsprung disease. It helps ascertain the length of aganglionic colon and aids in surgical planning. Washing out of the distal colon with enemas should be avoided before the contrast enema as it may distort a low transition zone. The catheter should be placed just inside the anus without inflation of the balloon to avoid distortion of a low transition zone and perforation. Radiographs are taken immediately after hand injection of contrast and 24 hours later. Positive findings include a transition zone with proximal dilated bowel, a microcolon, or retention of contrast in the bowel 24 hours post the enema on the delayed radiograph. Contrast enema is less reliable in the neonatal period than in older infants (42).

On searching the secondary literature, we found a study entitled “Diagnostic Tests in Hirschsprung Disease: A Systematic Review” by de Lorijn et al. This was assigned level 3a evidence using the Oxford Center for Evidence-Based Medicine (29). This systematic review showed that contrast enema had a sensitivity of 70% and specificity of 83% (12 studies for a total of 425 patients; Table 1 and Fig 3), which was significantly lower than those of rectal suction biopsy and anorectal manometry (43). Further studies have confirmed this (44–46).

Imperforate Anus

In approximately 10% of patients, there are no clinical signs to reveal the location of the anorectal anomaly after 24 hours and radiography can be useful in this group. A prone cross-table lateral radiograph can be performed to look for air in the rectum and identify the distance of the rectum from the imperforate anus at the skin. If the distance is <1 cm, anoplasty can be performed rather than colostomy in the absence of urinary tract or genital tract abnormalities (47).

Radiography.—On searching the secondary literature, we found a study entitled “Prone Cross-table Lateral View: An Alternative to the Invertogram in Imperforate Anus” by Narasimharao et al. (48). This was assigned level 4 evidence using the Oxford Center for Evidence-Based Medicine (29).
This showed that cross-table lateral radiograph is superior to invertogram radiograph in delineating the most caudal point of the rectum in neonates. Prone cross-table lateral radiographs were performed with the infant in prone position with the hips flexed in a genupectoral position for 3 minutes before a radiograph centered over the greater trochanters. Invertograms were performed holding the infant upside down for 3 minutes before a radiograph centered over the greater trochanters. Infants were much less distressed in the prone lateral position. However, one study showed a low sensitivity of invertogram of only 27% when compared to surgical findings as a gold standard (49).

Infants with imperforate anus should also have radiography of the spine and sacrum performed to assess for associated anomalies.

Ultrasound.—Ultrasound of the perineum has been shown to have a sensitivity of 86% in assessing the bowel to skin measurement distance when compared to surgical findings as the gold standard (49). This is far superior to plain radiography.

Ultrasound of the abdomen should be performed to outline associated urinary obstruction or hydrocolpos or a persistent cloaca. Ultrasound of the spine may be performed in infants less than 3 months to assess for associated tethered cord or other spinal anomalies (47).

Colostography.—Colostography is a contrast study of the distal colon through the mucus fistula in patients who have had a colostomy for imperforate anus. Water soluble contrast is injected by hand at a pressure sufficient to distend the distal rectum and demonstrate any fistulas. The injection is continued until the infant voids, and images are taken during micturition to reveal, in a single radiograph, the sacrum, rectum height, perineum, fistula location, bladder, vesicoureteral reflux (if present), and urethra (47). This is very important in preoperative planning (50). Sensitivity of augmented pressure distal colostography has been shown to be 100% in one study using surgical findings as the gold standard (49).

Magnetic Resonance Imaging (MRI)
MRI is being increasingly used in the evaluation of infants with complex anorectal malformations and fistulas (51). Prenatal MRI often delineates the anatomy and can distinguish high anorectal malformation from cloaca before birth (52). It is useful at detecting the presence of fluid in the rectum suggesting a rectovesical fistula.

Lower GI Tract

Meconium Plug Syndrome and MI
The evidence for the postnatal diagnosis of meconium plug and MI is severely limited and appears to be based mainly on consensus opinion or low-quality evidence. The use of plain radiography for the diagnosis of large bowel obstruction, followed by hyperosmolar contrast enema under fluoroscopic guidance for diagnosis and treatment, is the accepted algorithm for the investigation and management of meconium plug and simple, uncomplicated MI. Repetition of the enema may be required to fully relieve the obstruction. Infants with evidence of perforation or meconium peritonitis should proceed directly to surgery after plain radiography and should not undergo contrast enema.

Hirschsprung Disease
The main imaging tool for the diagnosis of Hirschsprung disease is contrast enema. It is inferior to both rectal suction biopsy and anorectal manometry in terms of sensitivity and specificity (43). When positive, contrast enema is a useful test and less invasive than rectal suction biopsy. Contrast enema is limited by false-negative results and cannot reliably outrule Hirschsprung disease, especially in cases with a high pretest probability and a negative test (Fig 3).

Imperforate Anus
The investigation and management of imperforate anus and the associated malformations depends on the clinical findings in the neonate, the results of urinalysis, and assessment of associated anomalies such as VACTERL or trisomy 21. In a select group of patients with no clinical signs to reveal the location of the anorectal anomaly after 24 hours, cross-table lateral radiographs are useful when they show air in the rectum <1 cm from the skin. Ultrasound is important to assess for associated urinary or genital tract anomalies, the presence of which will impact on the surgical management. Perineal ultrasound has been shown to be superior to radiography in delineating the location of the distal rectal pouch.

The majority of patients will require diversion colostomy. After the colostomy has healed, this group benefit from preoperative augmented pressure distal colostography to delineate the surgical anatomy and identify fistulas prior to...
definitive surgical repair. MRI has a role in prenatal diagnosis and an emerging role in postnatal preoperative evaluation, especially in complex cases.

**EVALUATE**

**Upper GI Tract**

**Duodenal Atresia and Stenosis, Jejunum, and Ileal Atresia**
More research is needed into the optimal imaging of these rare upper GI atresias and stenosis. At present, we should follow the algorithm of plain abdominal radiography, followed by contrast swallow and contrast enema, based on the limited low-quality evidence.

**Malrotation and Volvulus**
With regard to malrotation and acute volvulus, the investigation of choice is an upper GI contrast study (6,36,53). Plain radiography is usually the initial investigation and is useful to rule out obvious perforation, in which case the child should proceed directly to surgery. Other useful adjuncts to the upper GI contrast study are ultrasound and barium enema, especially in equivocal cases (6).

**Lower GI Tract**

**Meconium Plug Syndrome and MI**
More research is needed into the optimal imaging of meconium plug and MI, as well as the use of contrast enema to relieve the obstruction. At present, it is recommended to follow the established algorithms of plain radiography followed by hyperosmolar contrast enema under fluoroscopic guidance for the diagnosis and treatment of meconium plug syndrome and uncomplicated, simple MI.

**Hirschsprung Disease**
There is still a role for contrast enema in the work-up of infants with suspected Hirschsprung disease. It is less invasive than rectal suction or full thickness biopsies and, when positive, can guide treatment. Radiologists and referring physicians should be aware of its pitfalls and seek further investigations in patients with negative enemas whose clinical symptoms give them a high pretest probability.

**Imperforate Anus**
Multiple imaging modalities including radiography, ultrasound, augmented pressure distal colostography, and MRI have a role to play in the work-up of imperforate anus, depending on the clinical findings and presence of associated malformations. More research is needed to compare these modalities and identify the relevant sensitivities and specificities in the different cohorts of these patients.

**REFERENCES**