

AMSER Case of the Month

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5-year-old male with abdominal pain and vomiting

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Patient Presentation

HPI: Previously healthy 5-year-old male presents to the emergency department with right lower quadrant abdominal pain, nausea, and vomiting. Abdominal pain began 3 days ago with development of nausea and vomiting with poor oral tolerance the following day. Last bowel movement was 3 days ago but patient can pass gas. No fevers/chills, diarrhea, or hematochezia. No history of similar symptoms.

No significant past medical or surgical history

Patient Presentation

Vitals: Within normal limits

Physical exam:

- Constitutional: No acute distress, non-toxic appearing
- Abdominal: Abdomen is soft and non-distended. Diffuse abdominal tenderness without guarding or rebound.

No pertinent labs

What Imaging Should We Order?

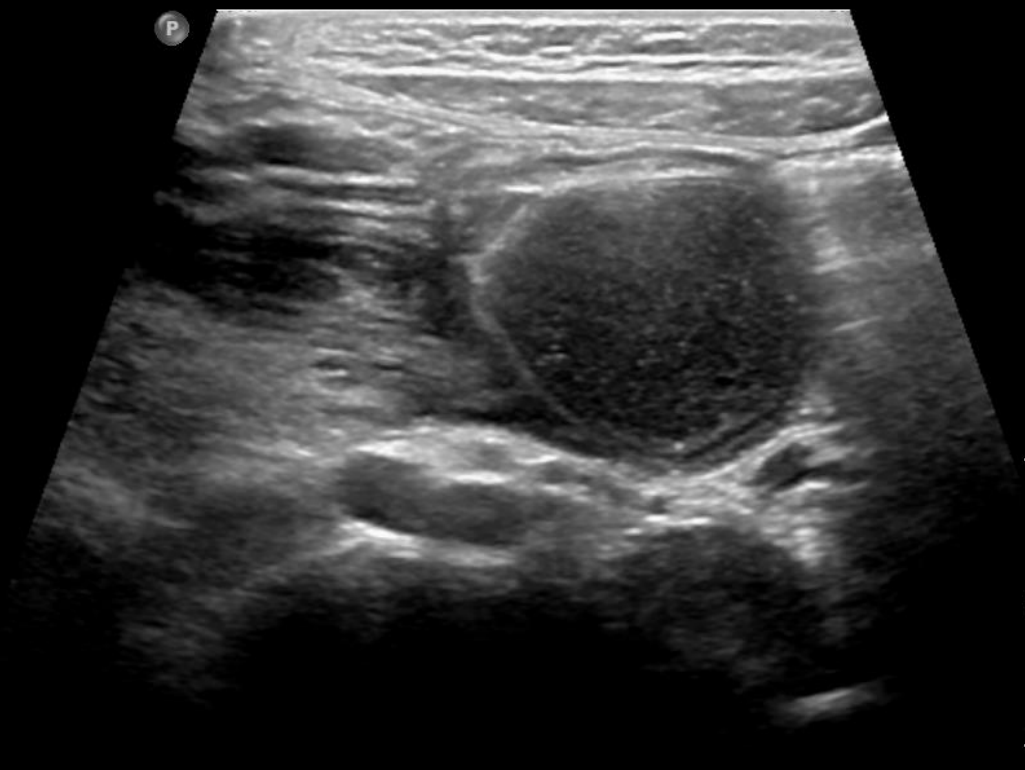
Select the applicable ACR Appropriateness Criteria

Variant 2: Child. Suspected acute appendicitis, intermediate clinical risk. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
US abdomen RLQ	Usually Appropriate	○
US abdomen	Usually Appropriate	○
CT abdomen and pelvis with IV contrast	May Be Appropriate (Disagreement)	⊗⊗⊗⊗
CT abdomen and pelvis without IV contrast	May Be Appropriate (Disagreement)	⊗⊗⊗⊗
MRI abdomen and pelvis without and with IV contrast	May Be Appropriate (Disagreement)	○
MRI abdomen and pelvis without IV contrast	May Be Appropriate (Disagreement)	○
Radiography abdomen	May Be Appropriate (Disagreement)	⊗⊗
CT abdomen and pelvis without and with IV contrast	Usually Not Appropriate	⊗⊗⊗⊗⊗⊗
US pelvis	Usually Not Appropriate	○

Due to suspected appendicitis, this imaging modality was ordered by the ER physician

Findings (unlabeled)

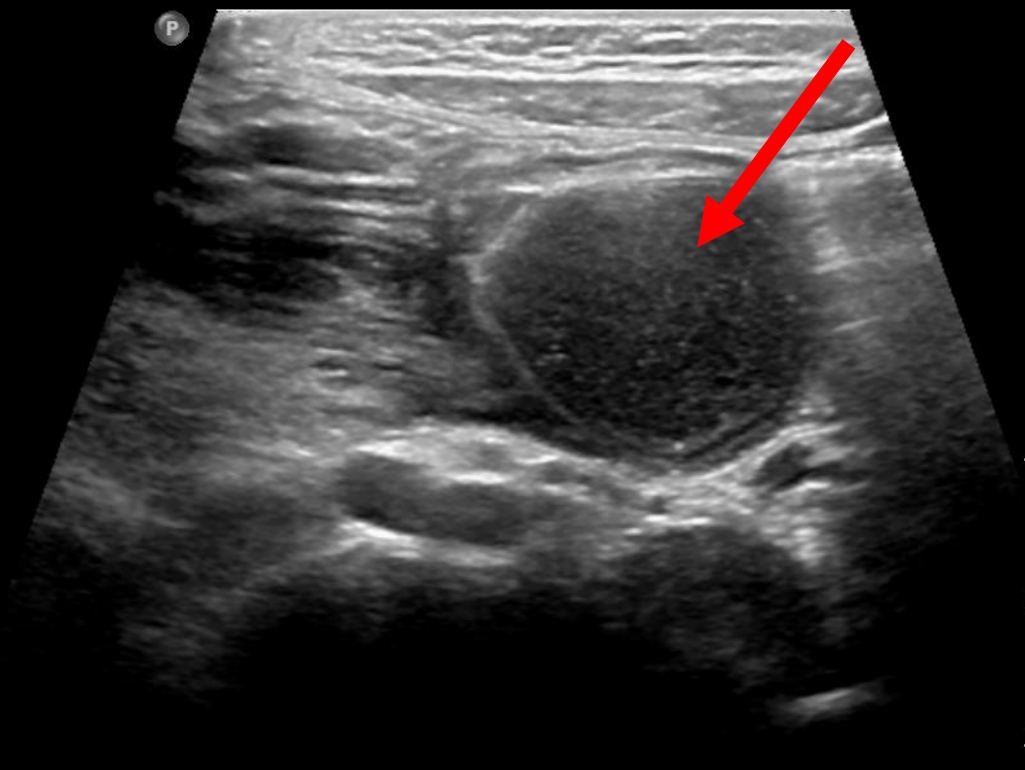


US umbilical area longitudinal

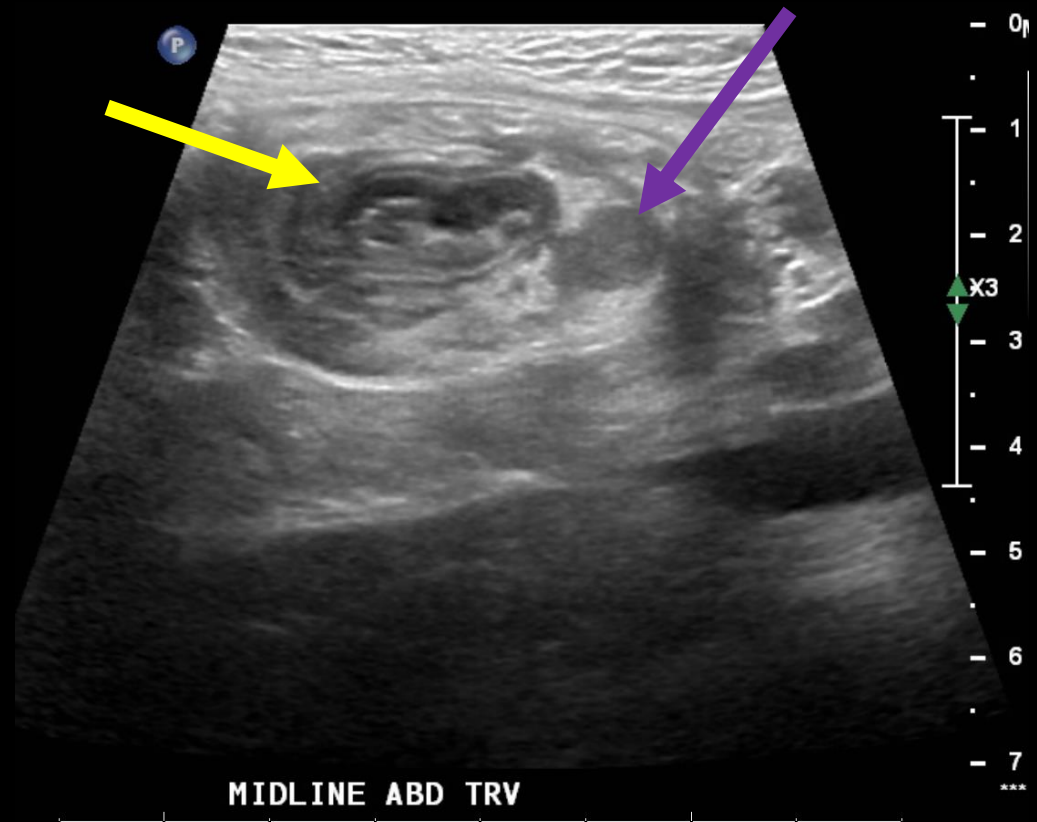


US umbilical area transverse

Findings (labeled)

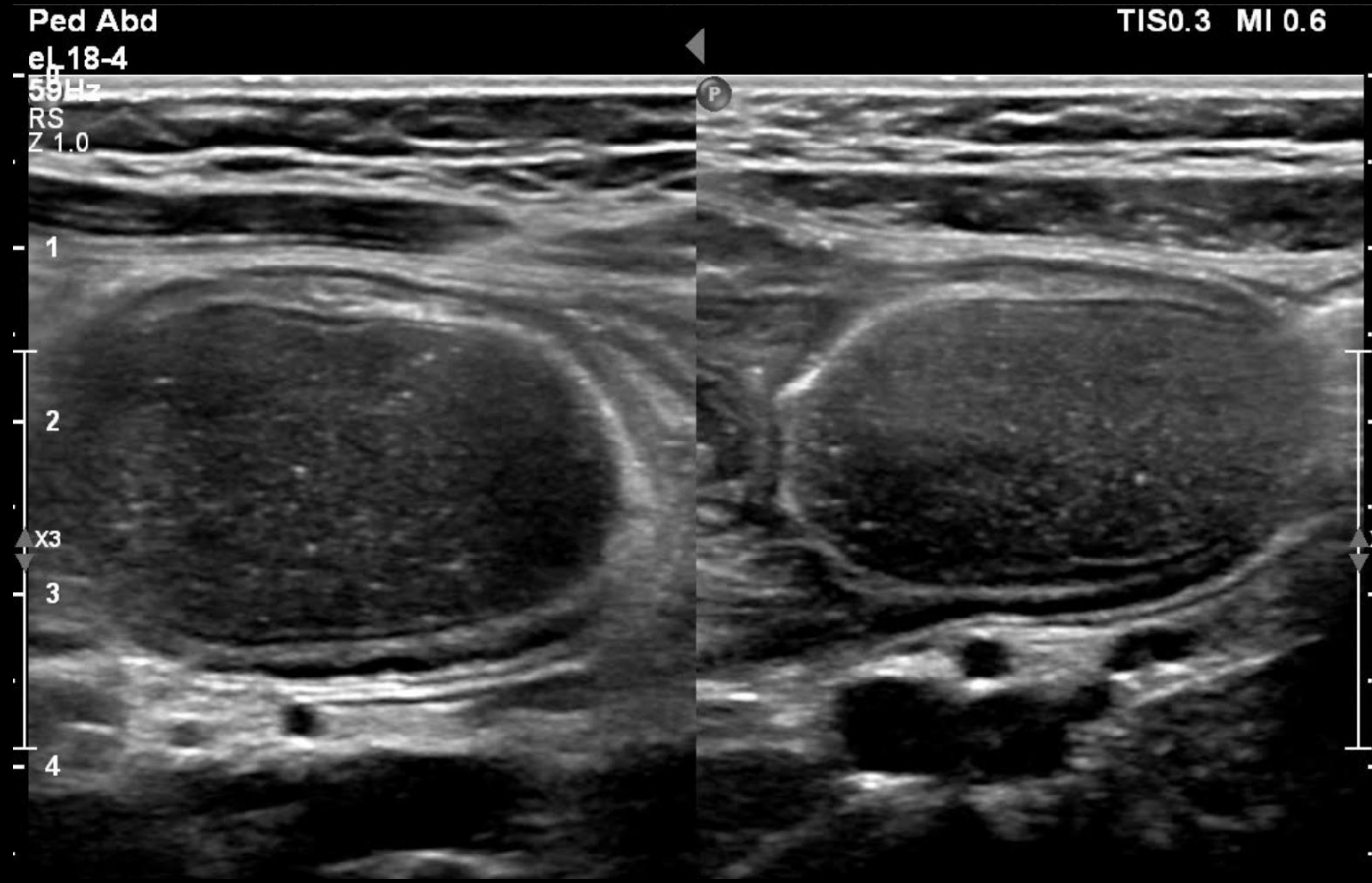


US umbilical area longitudinal



US umbilical area transverse:
Intussusception (yellow arrow) with
suspected cystic lead point (red
arrow) and lymph node (purple
arrow)

Findings (unlabeled)

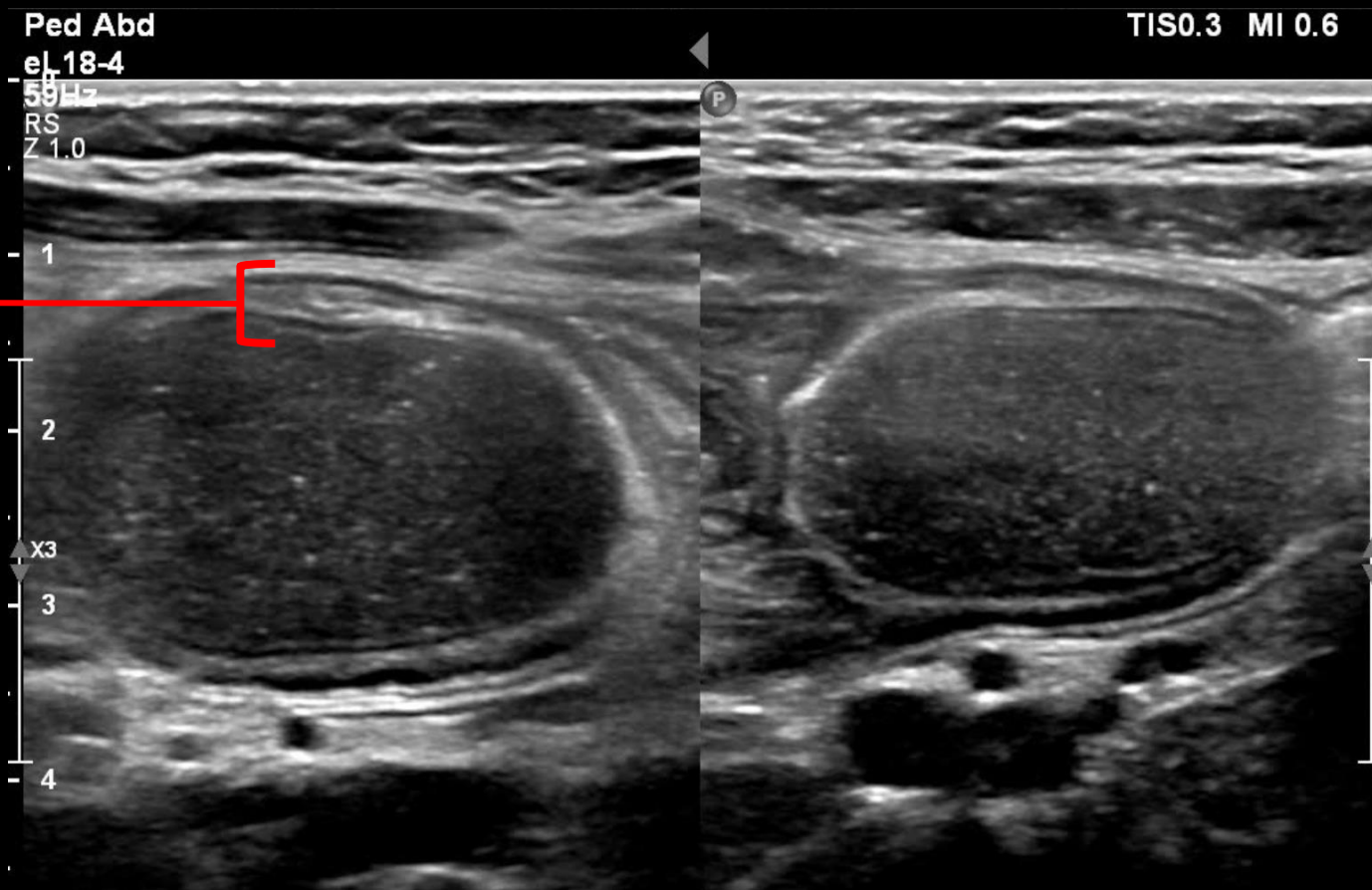


US umbilical area

Longitudinal

Transverse

Findings (labeled)



3.5 x 2.4 x 3.0 cm rounded cystic structure containing complex fluid without solid internal component; no internal vascularity was present (power doppler images not shown)

Final Dx:

Ileocolic intussusception with enteric duplication cyst as
lead point

Case Discussion

- Intussusception is the telescoping of intestine into an adjacent bowel segment
- Classic presentation: Intermittent abdominal pain, vomiting, and a palpable abdominal mass on exam; red “currant jelly” stool later in disease process due to bowel ischemia
- Intussusception is most common from 6 – 36 months old
 - The most common etiology in this age group is idiopathic (1)
 - Intussusception in older children and adults is more likely secondary to a lead point
 - Ntoulia et al. found the incidence of a lead point in cases of intussusception to be 25% (2)
 - Common lead points include lymphoid hyperplasia, Burkitt lymphoma, Meckel diverticulum, enteric duplication cyst, juvenile polyps, and appendicitis(2)
- Diagnosis: Air/contrast enema is the gold standard for diagnosis in children, though ultrasound is useful as a screening tool
 - Ultrasound findings: target sign, pseudokidney sign, crescent in a doughnut sign (3)
- Treatment: Air/contrast enema is effective in reducing the intussusception in most children
 - Indications for surgery: **lead point requiring surgery**, signs of bowel ischemia, unsuccessful enema reduction (3)
- The age of our patient (5 years old) and ultrasound findings raise suspicion for intussusception secondary to a lead point

Case Discussion

- Enteric duplication cysts (EDCs) are a rare congenital malformation of the GI tract most found during childhood and occur in 0.2 percent of children, more common in males. Enteric duplication cysts are characterized by an epithelial lining consisting of alimentary tract mucosa surrounded by a layer of smooth muscle with close approximation to the GI tract. (4)
- EDCs can be present anywhere along the GI tract but are most common in the ileum (5).
- Clinical presentation: Complications of EDCs include recurrent abdominal pain, intestinal obstruction, intussusception, and volvulus. Other symptoms of mass effect are possible given the location of the lesion. Bleeding, ulceration, and perforation are possible if the cyst wall contains gastric mucosa. (6)

Case Discussion

- EDC Imaging findings: Abdominal ultrasound is the preferred imaging modality for detecting EDCs except for esophageal lesions. Ultrasound will disclose a cyst adjacent to the GI tract with the double-wall or gut signature sign. Identification of a cyst with the gut signature sign is pathognomonic for EDC. The Y sign, caused by splitting of the muscle layer between the cyst and gut, is also highly specific for EDC. (7,8)
- The gut signature sign is preserved in benign conditions and describes the appearance of the gastrointestinal wall
- Treatment: Surgical excision is the definitive treatment and is recommended in symptomatic and incidental EDCs due to the risk of morbidity and mortality (4).
- This case demonstrates a rare example of an EDC acting as a lead point for intussusception. This patient subsequently underwent exploratory laparoscopy converted to laparotomy with ileocecectomy for resection of the cyst. Pathology confirmed the diagnosis of enteric duplication cyst. The patient's post operative recovery has been uneventful.

References:

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