AMSER Rad Path Case of the Month:

11-year-old female presenting with abdominal pain, vomiting, and constipation

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Patient Presentation

- HPI: 11-year-old female with a family history of Peutz-Jeghers Syndrome (mother) presented to her PCP office for a 3-day history of abdominal pain, constipation, bilious vomiting, and chills. She was referred to the ED. Her last bowel movement was two days prior.

PMH: None
Surgical History: None

Vitals:
BP: 131/79 Pulse: 117 BPM RR: 24 Temp: 98.5°

Physical Exam:
Constitutional: Well-developed, pale, tired appearing
Abdomen: Distended, Diffuse tenderness throughout with slight guarding.
Mouth/Throat: Moist MM, Diffuse melanocytic macules on lower lip.
Cardiac: Tachycardic regular rhythm, warm, well-perfused extremities.
Neurologic: Alert, CNs grossly intact

Pertinent Labs

Complete Blood Count	Result	Reference Range
Hemoglobin	14.2 g/dL	11.5 – 15.5 g/dL
MCV	82.3 F fL	77.0 – 95.0 fL
WBC	62.7 k/uL	5.0 – 11.0 K/uL
Abs Neutrophils	40.8 k/uL	1.50 – 8.00 K/uL
Platelet Count	688 K/uL	200 – 450 K/uL

Comprehensive Metabolic Panel	Result	Reference Range
Sodium	129 g/dL	136 – 145.0 mmol/L
Potassium	4.9 mmol/L	3.4 – 4.7 mmol/L
Chloride	93	101 – 109 mmol/L
BUN	10 mg/dL	6 – 25.0 mg/dL
Creatinine	0.54 mg/dL	0.57 – 1.25 mg/dL

Lab	Result	Reference Range
CRP	29.34 mg/dL	<0.5 mg/dL
Procalcitonin	9.964 nm/ml	0-.4 ng/ml
Lactate	1.08 mmol/L	0.50 – 2.0 mmol/L

What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

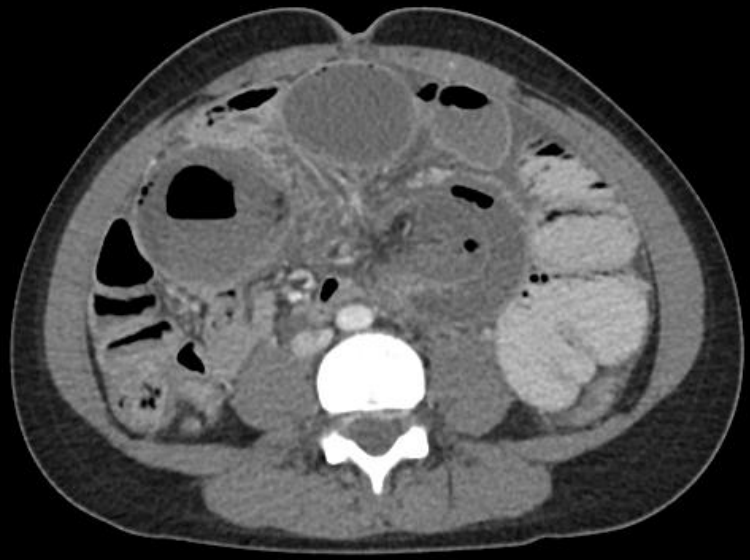
Variant 1: **Acute nonlocalized abdominal pain and fever. No recent surgery. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
CT abdomen and pelvis with IV contrast	Usually Appropriate	☢☢☢
MRI abdomen and pelvis without and with IV contrast	May Be Appropriate	O
US abdomen	May Be Appropriate	O
CT abdomen and pelvis without IV contrast	May Be Appropriate	☢☢☢
MRI abdomen and pelvis without IV contrast	May Be Appropriate	O
CT abdomen and pelvis without and with IV contrast	May Be Appropriate	☢☢☢☢
Radiography abdomen	May Be Appropriate	☢☢
FDG-PET/CT skull base to mid-thigh	Usually Not Appropriate	☢☢☢☢
WBC scan abdomen and pelvis	Usually Not Appropriate	☢☢☢☢
Nuclear medicine scan gallbladder	Usually Not Appropriate	☢☢
Fluoroscopy contrast enema	Usually Not Appropriate	☢☢☢
Fluoroscopy upper GI series with small bowel follow-through	Usually Not Appropriate	☢☢☢

This imaging modality was ordered by the ER physician

Radiology Images (not labeled)

CT Abdomen Pelvis with Contrast



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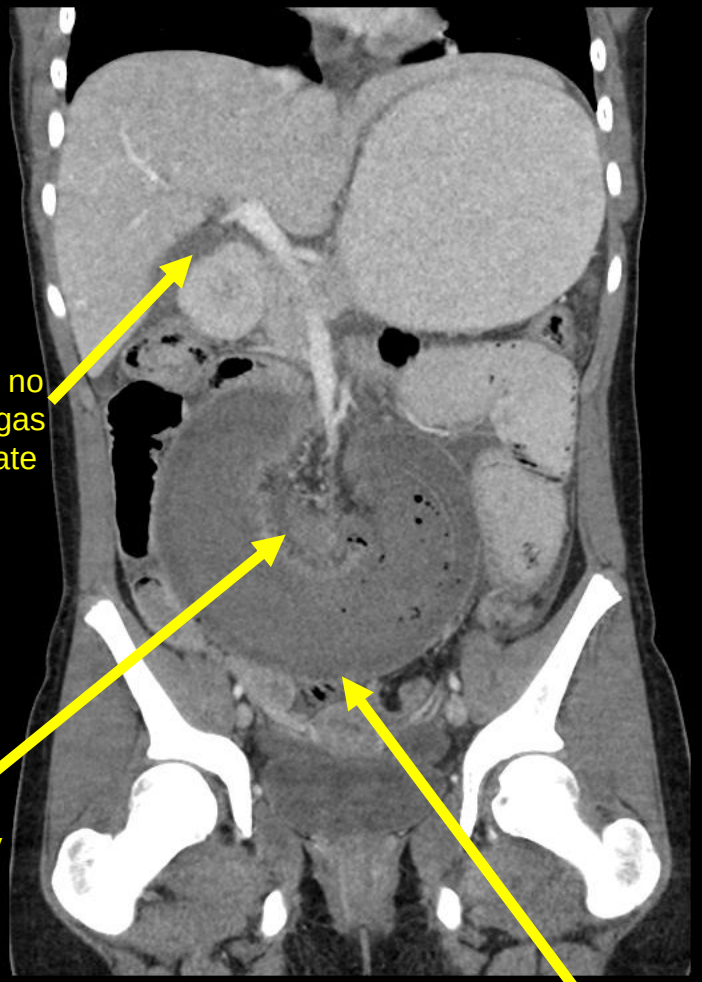

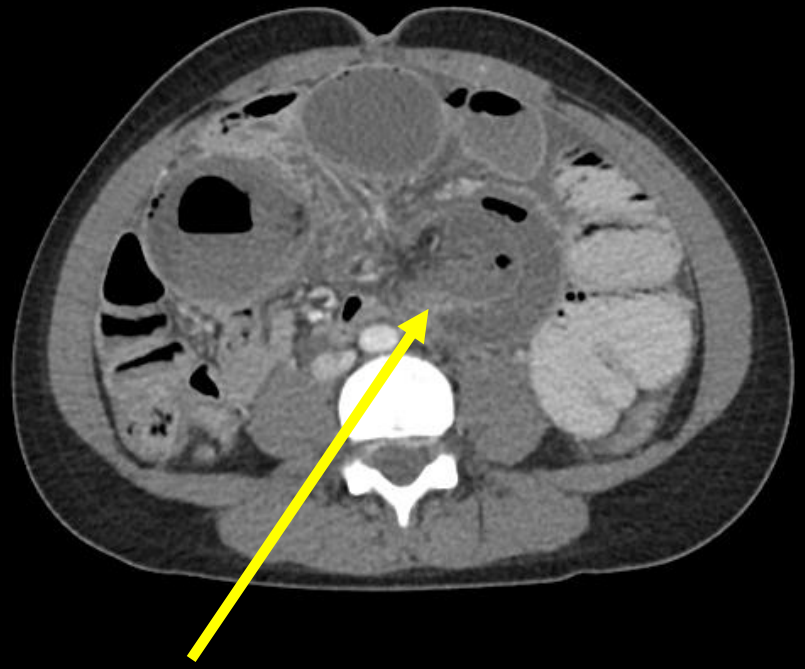

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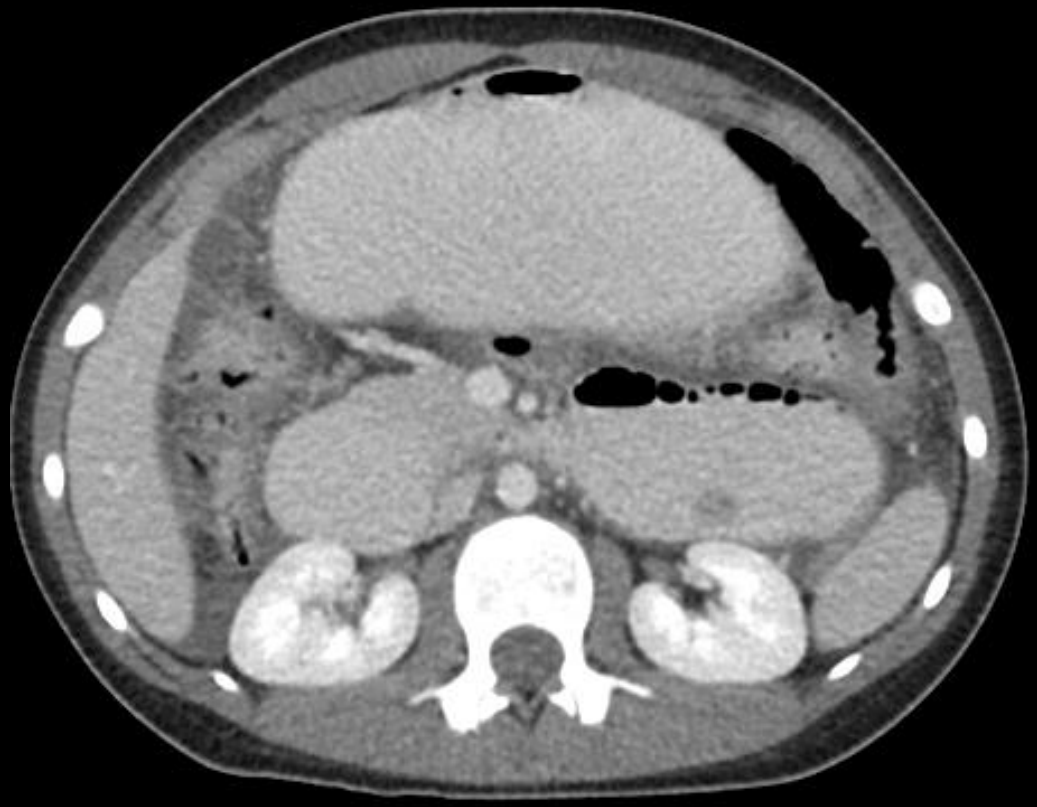

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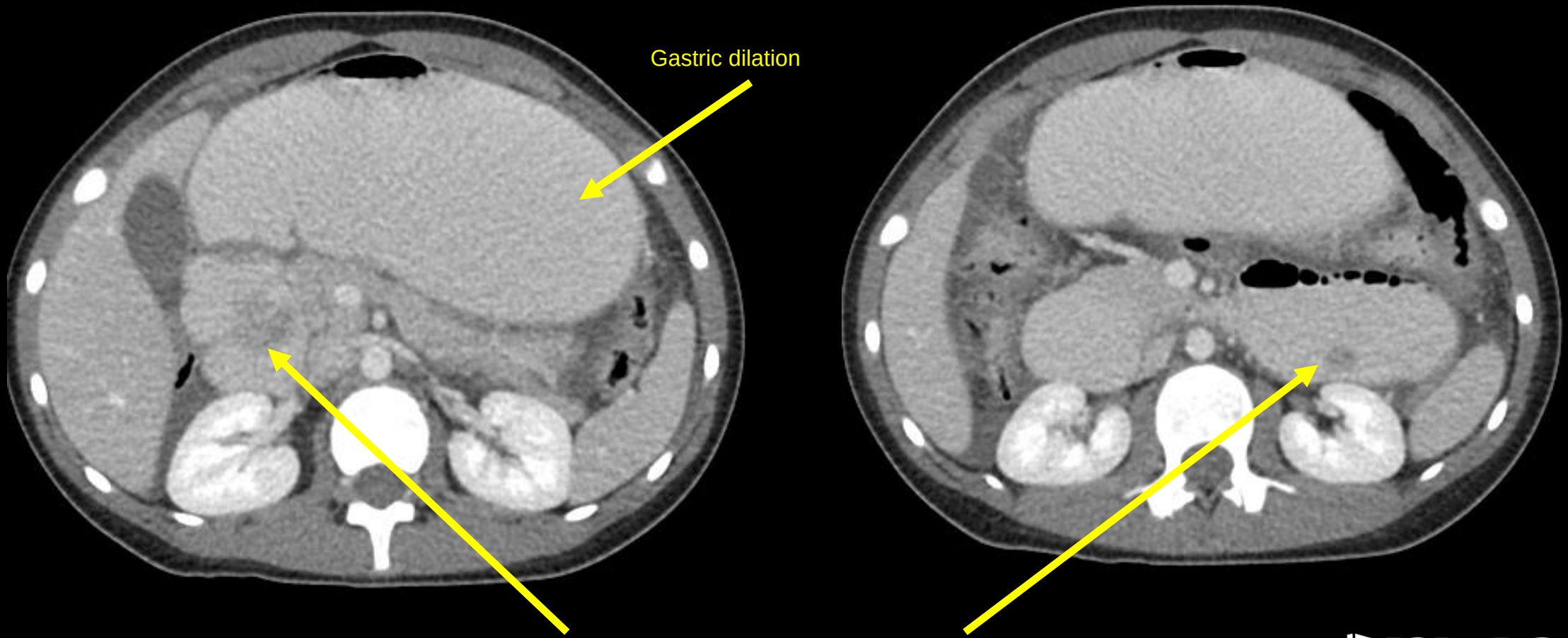


Differential diagnosis

Pediatric Small Bowel Obstruction: Mnemonic AIM x2

- A - Appendicitis , Adhesions
- I - Intussusception, Inguinal hernia/Internal hernia
- M - Meckel's diverticulum, Midgut malrotation with volvulus

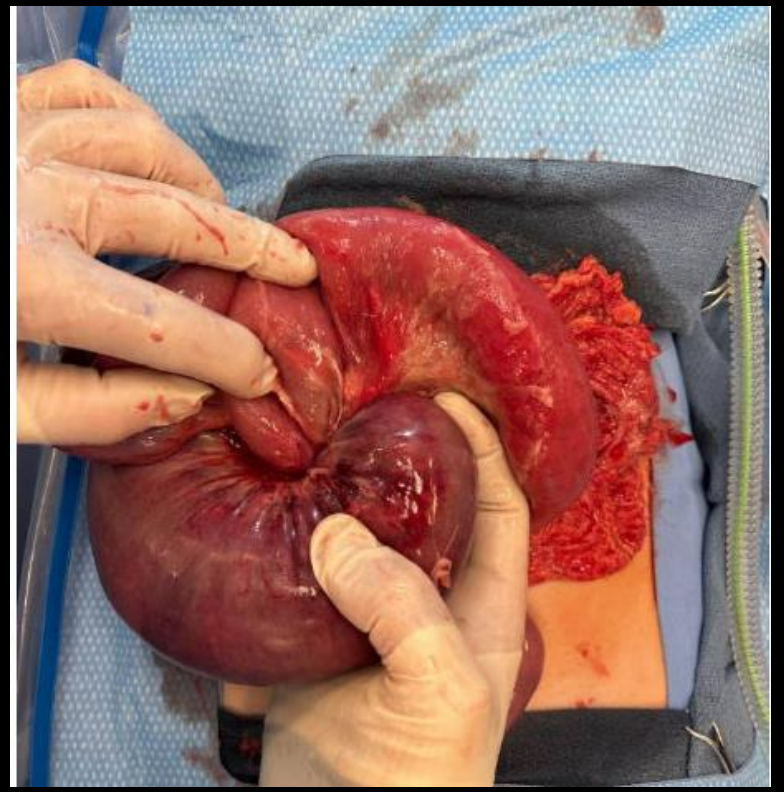
Differential based on Imaging:

- Small Bowel Obstruction secondary to:
  - Intussusception
  - Internal Hernia
  - Adhesions/Congenital band
  - Small bowel volvulus
  - Concern for secondary complication of bowel necrosis

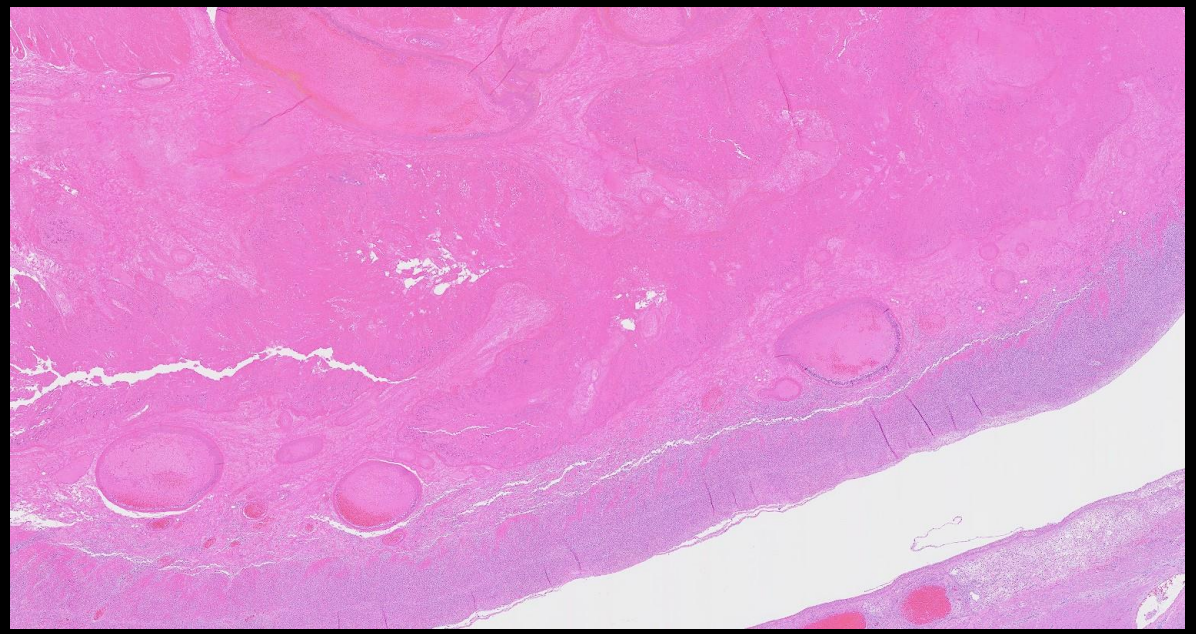
Gross Path

Gross Description:

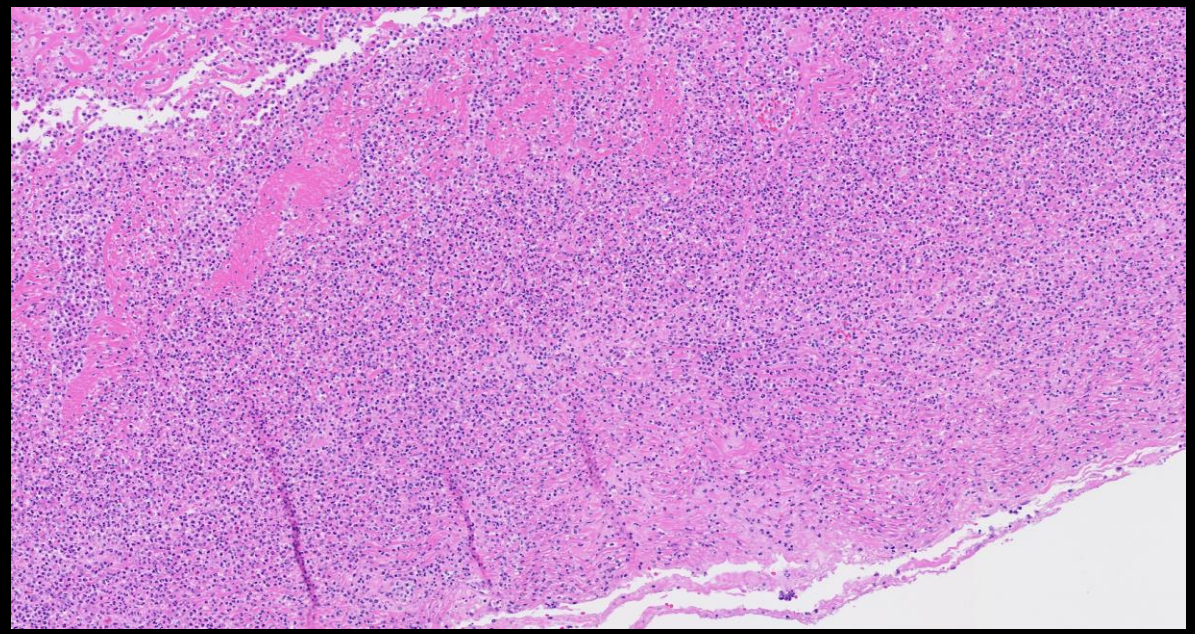
- 38 cm torsed segment of small bowel with ischemic necrosis and acute peritonitis.
- Serosal surfaces are violaceous to dark brown, centrally hemorrhagic.
- Proximal end is remarkable for an area of involution consistent with intussusception with a dilated outer diameter of 4.6 cm.



Micro Path



Necrotic bowel negative for atypia or malignant neoplasm



High Power: Necrotic mucosa with acute inflammation

Final Dx:

Complete small bowel obstruction secondary to jejunojejunal intussusception with necrotic bowel

Interval History

- Patient with presumptive diagnosis of of Peutz-Jeghers given physical exam findings and established diagnosis in mother.
- Intraoperatively patient was found to have perforation of proximal jejunum. She underwent small bowel resection with primary anastomosis with 270 cm of small bowel remaining.
- Intraoperative peritoneal fluid cultures grew enterobacter and patient was started on IV antibiotics

Case Discussion

- Background
  - Pediatric bowel obstructions are one of the most common surgical emergencies in children.
  - If SBO progresses to complications such as bowel ischemia, perforation, or sepsis, especially in the pediatric population, delay in surgical intervention can result in a high risk of morbidity and mortality.[1]
  - Intussusception is defined as the invagination of one segment of the bowel into an immediately adjacent segment of the bowel.
  - Most often the cause is idiopathic and thought to be due to hypertrophy of lymphoid tissue (Peyer patches) from viral infection in setting of classic ileocolic intussusception. Pathologic lead points such as Meckel's diverticulum, polyp, or lymphoma can also cause intussusception.[1]
  - The hamartomatous polyps in PJS are benign growths caused by noncancerous tissue proliferation. Intestinal intussusception has been observed in 47%–69% of adult patients with Peutz–Jeghers syndrome.[6]
  - Ileocolic intussusception is the most common cause of small bowel obstructions in children. Rarely does a part of the ileum or jejunum prolapse into itself.[2]
- Clinical Presentation
  - Affected pediatric patients may present with the classic triad of crampy abdominal pain, red currant jelly stool, and a palpable abdominal mass, although this triad is present in less than 50% of children. [1]
  - Alternatively, pediatric patients with intussusception can present with signs of a bowel obstruction with a distended abdomen and pain. [1]

Case Discussion

- Imaging:
  - Radiographs are usually the first choice to evaluate for signs of bowel obstruction and possible bowel perforation.
  - If bilious vomiting is present and malrotation with midgut volvulus is suspected, an emergent UGI should be performed.
  - Classically US is the modality of choice for intussusception typically shows a 3-cm to 5-cm mass along the expected course of the colon with a characteristic targetoid or donut appearance in transverse images.[1]
  - CT may be utilized in cases where bowel necrosis, pneumoperitoneum and peritonitis are of concern.

- Management
  - Non-surgical management used in stable children is fluoroscopically or US-guided air or contrast/hydrostatic enema for treatment of classic ileocolic intussusception.
  - Surgical reduction performed if image-guided enema is unsuccessful or if there are signs of bowel necrosis or bowel perforation.[3]
  - Periodic surveillance and removal of larger polyps aim to reduce the likelihood of complications in Peutz-Jeghers syndrome and may obviate the need for repeated urgent operations and extensive small bowel resection leading to short bowel syndrome.[4]

References:

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2. Jain S, Haydel MJ. Child Intussusception. [Updated 2023 Apr 10]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2025 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK431078/

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4. Nasri S, Kellil T, Chaouech MA, Zouari K. Intestinal intussusception in Peutz Jeghers syndrome: A case report. *Ann Med Surg (Lond)*. 2020;54:106-108. Published 2020 May 4. doi:10.1016/j.amsu.2020.04.013

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