

Imaging-Negative Double Jejunal Intussusception Diagnosed on Laparoscopy With Intraoperative Enteroscopy in a Patient With Peutz-Jeghers Syndrome

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BACKGROUND

Peutz-Jeghers syndrome (PJS) is a rare, autosomal dominant syndrome characterized by **gastrointestinal polyposis, mucocutaneous pigmentation, and a high lifetime risk of malignancy**.^[1] Given their high polyp burden, many patients with PJS develop intussusception.^[2]

CASE PRESENTATION

- A 25 year-old female with PJS and a history of five prior episodes of intussusception secondary to PJ-type polyps presented with **bloating, vomiting, and acutely worsening chronic abdominal pain**
- Patient had initially presented with intermittent abdominal pain six months prior, at which time **seventeen small bowel PJ-type polyps** were resected via anterograde small bowel enteroscopy
- Magnetic resonance cholangiopancreatography (MRCP) and magnetic resonance enterography (MRE) two months later demonstrated **two polyps in the terminal ileum** which were resected via colonoscopy
- Patient then presented with acutely worsening abdominal pain. **Computed tomography (CT) with IV contrast was unremarkable**, but given high clinical suspicion for intussusception, a multidisciplinary decision was made to proceed with diagnostic laparoscopy
- **Two jejunal intussusceptions related to large polyps** were identified and reduced, and **one large (25mm) polyp** was surgically resected (Image 1)
- **One smaller (15mm) duodenal polyp** and **three large (20-25mm) jejunal polyps** were resected on intraoperative enteroscopy via enterotomy (Image 2)
- Patient discharged three days postoperatively with resolution of pain

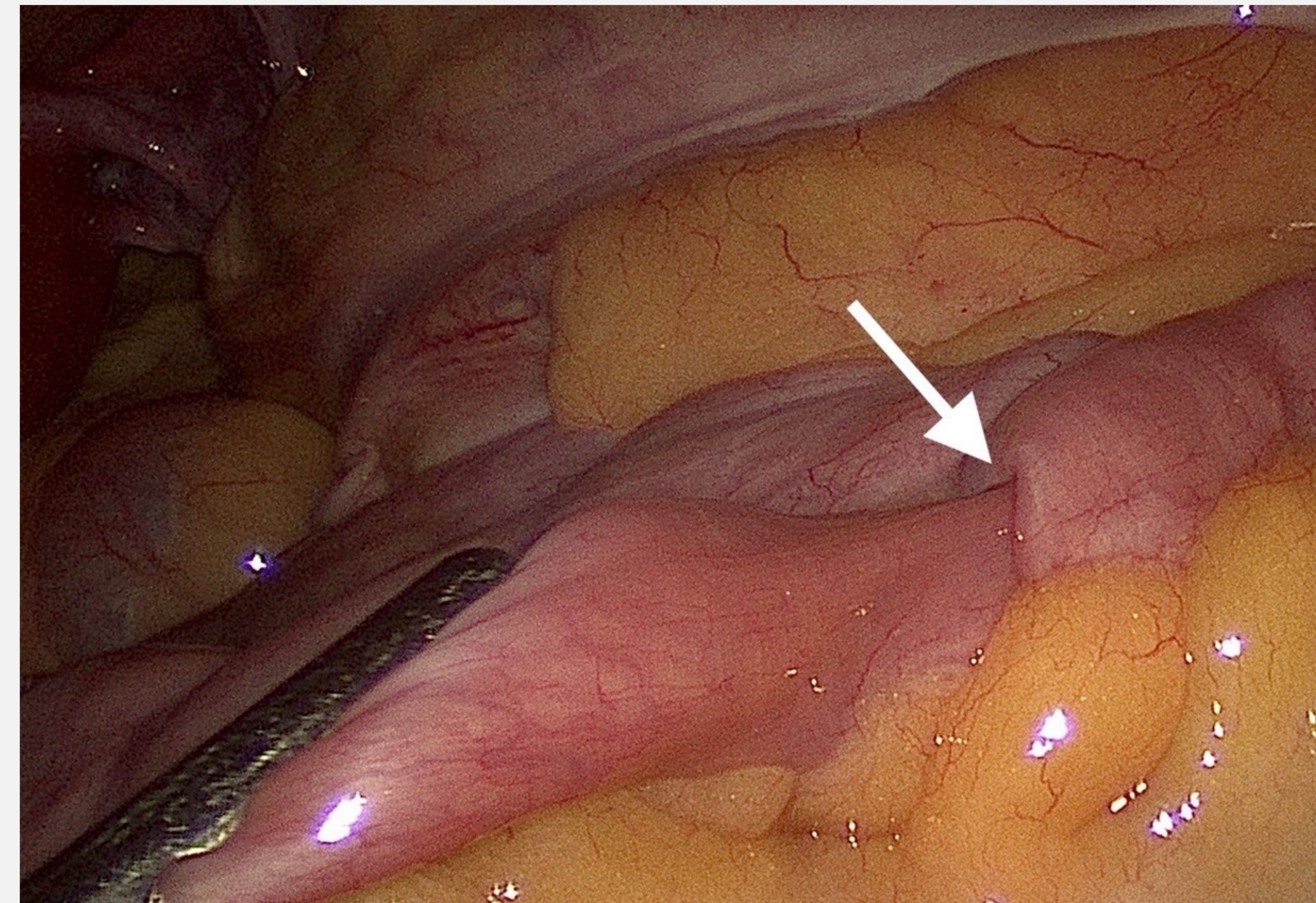


Image 1. Intussusception (white arrow) secondary to a large hamartomatous polyp as seen on laparoscopy

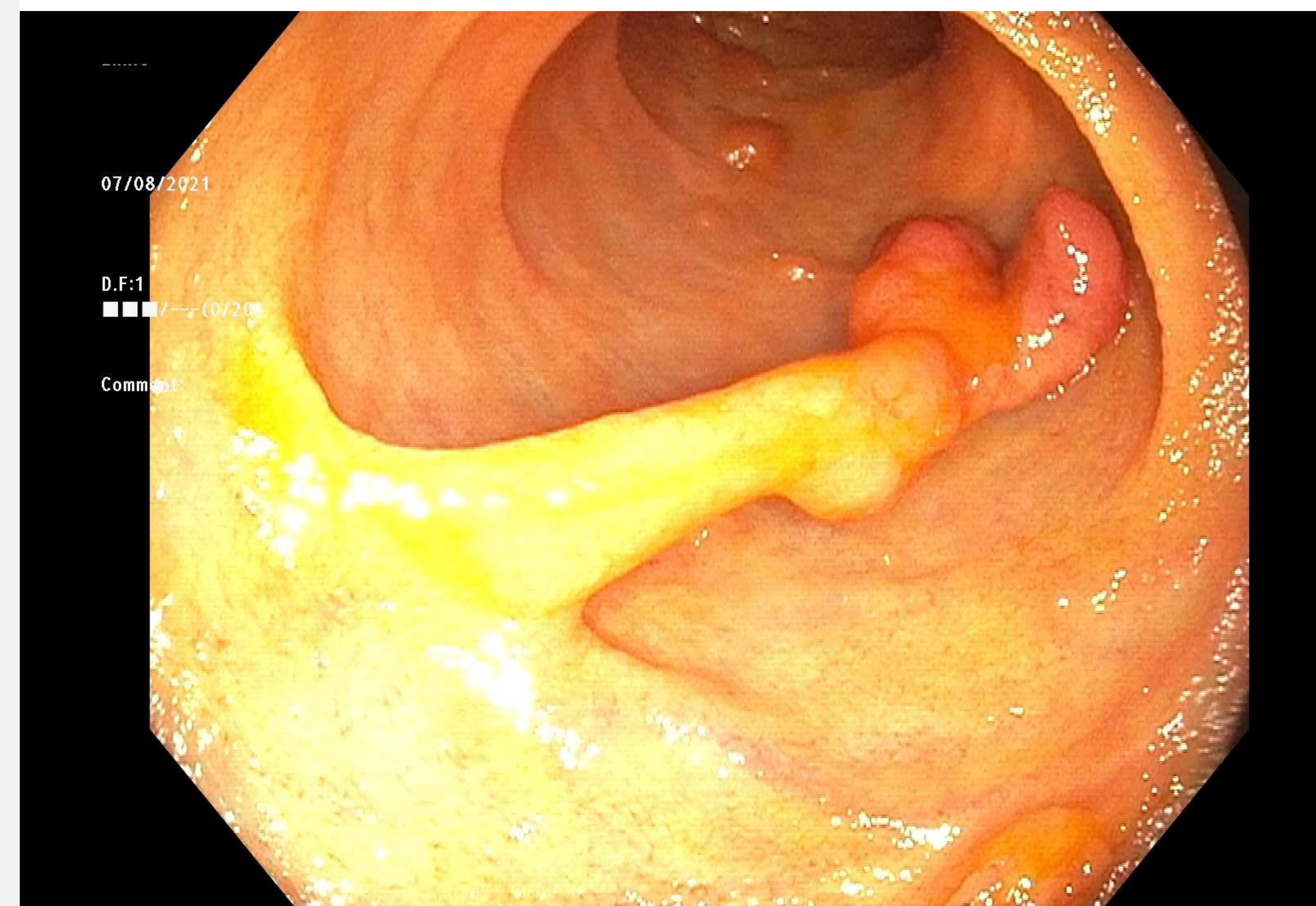


Image 2. Endoscopic view of a pedunculated small bowel polyp with evidence of numerous smaller, sessile polyps

DISCUSSION

- Patients with PJS require multidisciplinary care given their significantly **increased risk of both gastrointestinal and extraintestinal malignancy** and complications such as **intussusception secondary to PJ-type polyps**, which are typically benign (Image 3)^[3,4]
- While CT is generally the preferred imaging study when evaluating for intussusception, estimates of its sensitivity range from 58-100%^[5]

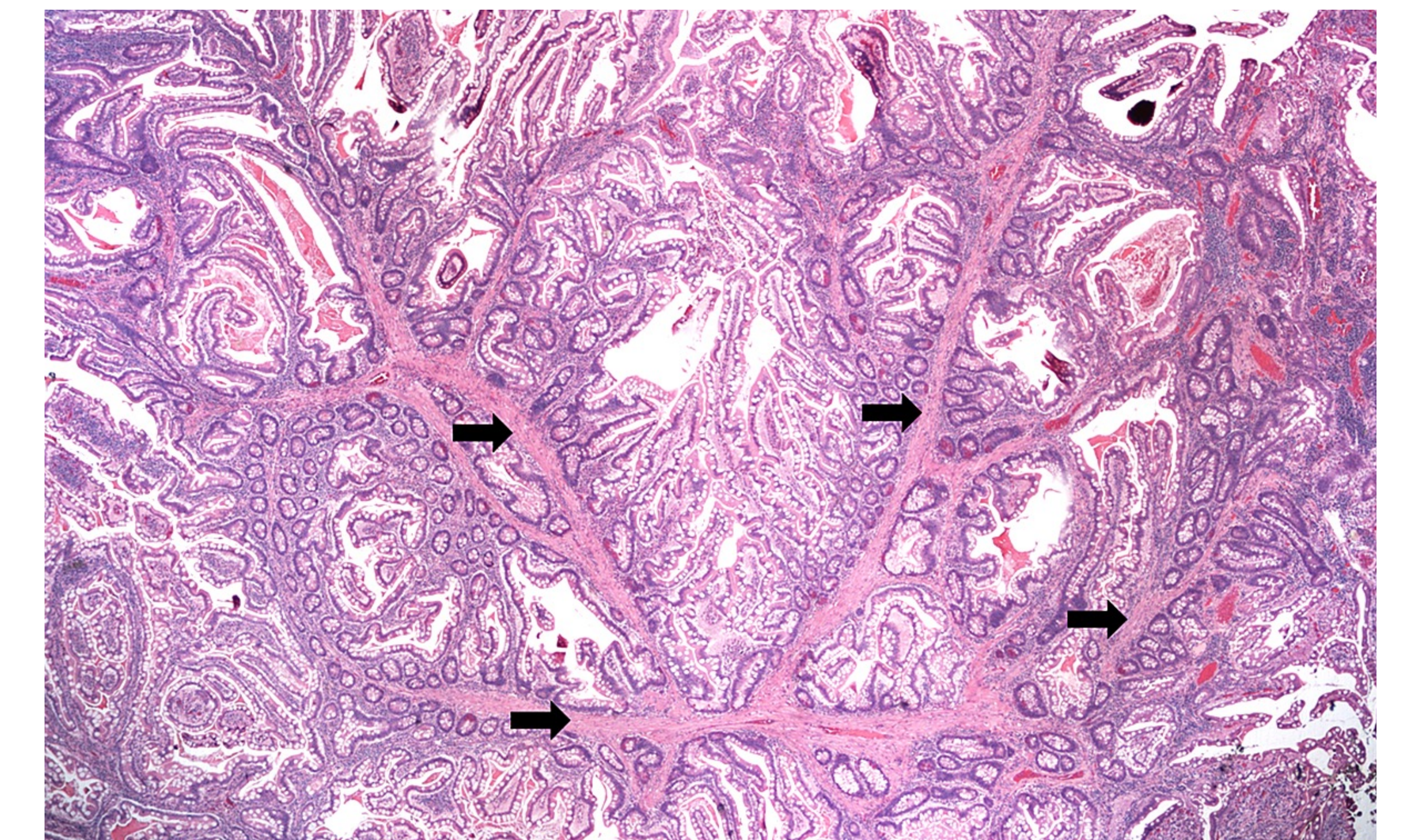


Image 3. A hamartomatous small bowel PJ-type polyp with characteristic "arborizing" smooth muscle bands (black arrows) (Hematoxylin & eosin stain, magnification: 200x)

CLINICAL RELEVANCE

- **PJ-type polyps may grow rapidly**
- In patients with predisposing risk factors and a compatible presentation, clinicians should **consider intussusception regardless of imaging**

REFERENCES

1. Jenne et al. Peutz-Jeghers syndrome is caused by mutations in a novel serine threonine kinase. *Nat Genet.* Jan 1998;18(1):38-43.
2. van Lier et al. High cumulative risk of intussusception in patients with Peutz-Jeghers syndrome: time to update surveillance guidelines? *Am J Gastroenterol.* May 2011; 106(5):940-5.
3. Latchford et al. Management of Peutz-Jeghers Syndrome in Children and Adolescents: A Position Paper From the ESPGHAN Polyposis Working Group. *J Pediatr Gastroenterol Nutr.* Mar 2019; 68(3):442-452.
4. Chen et al. Cancer risk in patients with Peutz-Jeghers syndrome: A retrospective cohort study of 336 cases. *Tumour Biol.* Jun 2017; 39(6)
5. Amr et al. Intussusception in adults and the role of evolving computed tomography technology. *Am J Surg.* Mar 2015; 209(3):580-3.