

Background of Peutz–Jeghers syndrome (PJS)

- **Genetic Basis and Key Characteristics**
 - Peutz–Jeghers syndrome: a hereditary disorder transmitted in an autosomal dominant pattern
 - presence of multiple hamartomatous polyps throughout the gastrointestinal tract
- **Cancer Predisposition**
 - substantially elevated lifetime risk of malignancy
 - Gastrointestinal cancers: involving the small intestine, stomach, colon, and pancreas
 - Extra-intestinal malignancies: pancreas, breast and gynecologic tumors
 - Lifelong surveillance
- **Gastrointestinal Manifestations**
 - Polyps most commonly in the small intestine, particularly the jejunum
 - Clinical complications:
 - Recurrent abdominal pain
 - Gastrointestinal bleeding
 - Intestinal obstruction
 - Intussusception
 - Perforation in severe cases

Case Presentation

- **Patient History**
 - A 45-year-old male with a known diagnosis of PJS for seven years
 - Previous interventions included:
 - Multiple upper endoscopies and colonoscopies for gastric and colonic polyp removal
 - Double-balloon endoscopy (DBE) revealing numerous pedunculated and sessile polyps within the small bowel
 - Three prior laparotomies for recurrent small bowel intussusception requiring polypectomy and limited resections
- **Current Presentation**
 - Symptoms:
 - Epigastric abdominal pain
 - Nausea and repeated vomiting
 - Abdominal CT:
 - Multiple segments of small bowel intussusception
 - One segment resulting in mechanical obstruction with proximal bowel dilatation
 - Clinical Decision:
 - Emergency surgical intervention indicated due to obstructive ileus
- **Exploratory Laparotomy**
 - Three separate intussuscepted bowel segments identified
 - One segment with marked proximal dilatation consistent with obstruction
 - Successful manual reduction of intussusception
- **Intraoperative Combined Endoscopic–Surgical Technique**
 - An enterotomy created near the appendiceal region
 - A colonoscope (Olympus CF-230L; 160 cm working length, 12 mm diameter) introduced through the enterotomy
 - Retrograde advancement with visualization of the small intestinal lumen

- **Polyp Identification and Removal**
 - Entire small intestine palpated manually to localize lesions
 - Endoscopic visualization: multiple pedunculated and sessile polyps
 - Snare polypectomy under direct collaboration between surgeon and endoscopist
 - Any suspected serosal thinning or microperforation immediately reinforced with sutures to maintain bowel integrity
- **Operative Outcomes**
 - **Polyp Clearance**
 - A total of 36 polyps removed
 - No additional enterotomies required
 - Larger polyps (>5 mm) preferentially excised in accordance with current management recommendations
 - **Closure and Recovery**
 - Enterotomy closed using an Endo-GIA stapling device
 - Patient discharged on postoperative day seven
- **Follow-Up**
 - Over two years of surveillance:
 - No recurrence of obstruction
 - No gastrointestinal bleeding
 - No new symptomatic intussusception
 - Regular imaging and endoscopic monitoring scheduled according to surveillance guidelines

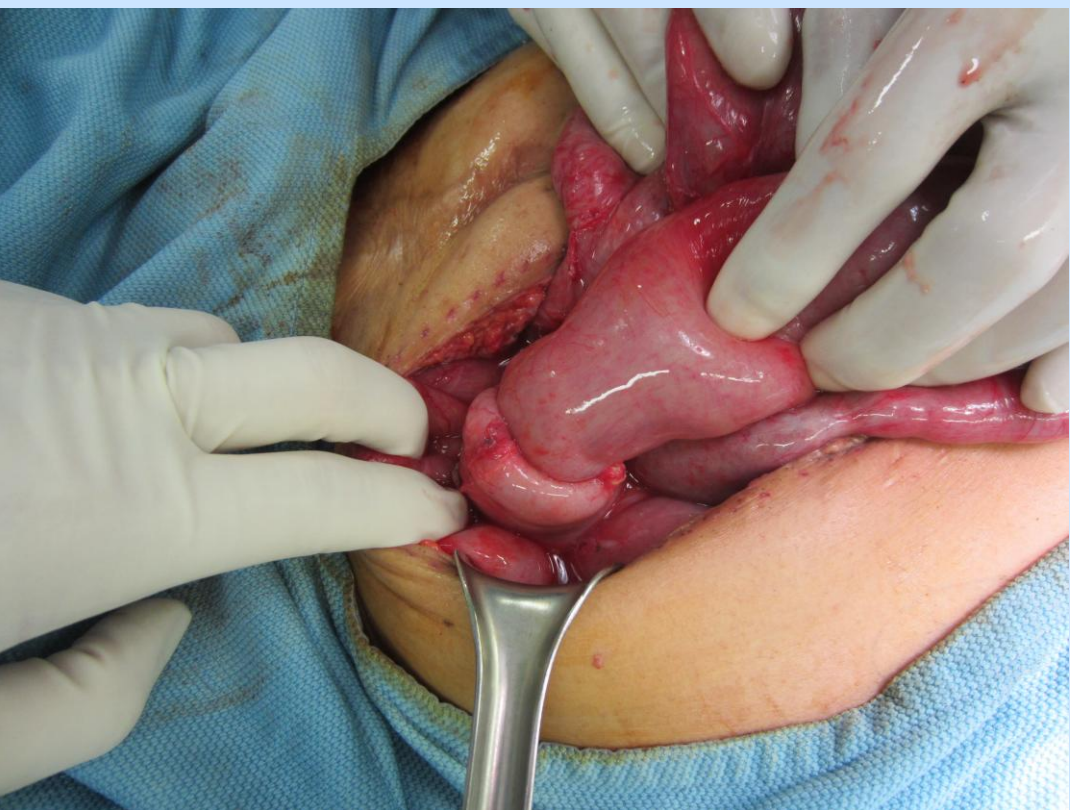


Fig 1
Small bowel intussusception caused by a pedunculated polyp, complicated by obstruction & proximal intestine dilatation



Fig 2
The process of polyp clearance via the combined endoscopic-surgical technique

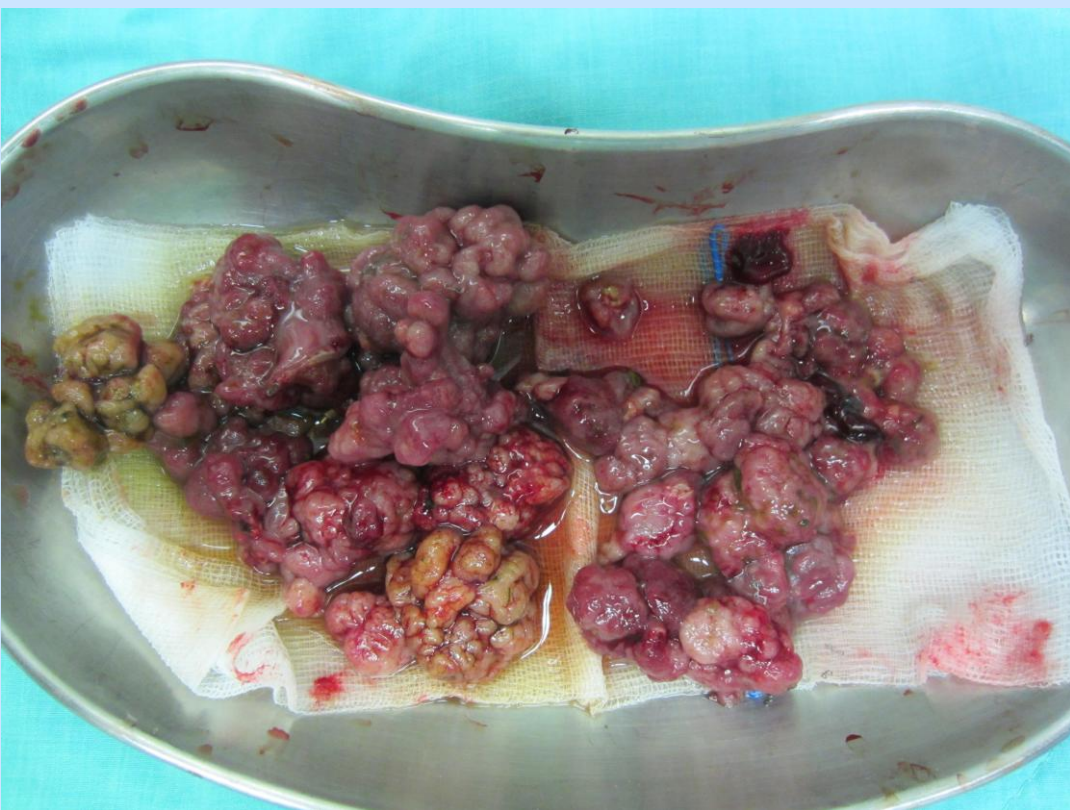


Fig 3
Small bowel polyps removed after the procedure of polyp clearance via the combined endoscopic-surgical technique

Discussion

- **Characteristics of PJS Polyps**
 - **Morphology**
 - Size ranges from a few millimeters to several centimeters
 - Typically lobulated in appearance
 - Larger lesions are frequently pedunculated
 - Smaller lesions often present as sessile polyps
 - **Distribution**
 - Small intestine: 60–90%
 - Colon: 50–64%
 - Stomach: 24–49%
- **Therapeutic Challenges in the Small Intestine**
 - **Limitations of Standard Endoscopy**
 - Push enteroscopy reaches only 70–150 cm beyond the ligament of Treitz
 - Complete visualization of the small bowel is often not feasible with conventional methods
 - **Device-Assisted Enteroscopy**
 - Double-balloon endoscopy improves access and therapeutic reach
 - However, its effectiveness may be reduced in patients with:
 - Dense postoperative adhesions
 - Numerous polyps requiring multiple sessions
 - Procedures can be prolonged and technically demanding
- **Surgical Considerations and Indications**
 - **When to Perform Segmental Resection**
 - Irreducible intussusception
 - Bowel ischemia
 - Dense clustered lesions unsuitable for endoscopic removal
 - Significant serosal compromise
 - **Indications for Open Polypectomy**
 - Large, bulky lesions
 - Thick-stalked polyps not amenable to snare technique
- **Importance of Bowel Preservation**
 - **Risk of Short Bowel Syndrome (SBS)**
 - Repeated resections may lead to progressive loss of functional small intestine.
 - SBS may result in:
 - Chronic malabsorption
 - Electrolyte imbalance
 - Persistent diarrhea
 - Dependence on long-term parenteral nutrition
- **Modern Treatment Philosophy**
 - Prioritize maximal polyp clearance
 - Minimize bowel resection
 - Preserve digestive and absorptive capacity
 - Improve long-term nutritional status and quality of life

Clinical Implications

- **The combined endoscopic–surgical approach:**
 - Allows comprehensive inspection of the entire small intestine
 - Achieves extensive polyp removal in a single session
 - Reduces the need for repeated enterotomies
 - Decreases the likelihood of excessive bowel resection
 - Provides a safe and bowel-preserving strategy for complex PJS cases