

Prevention of Intussusception in Peutz-Jeghers Syndrome*

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THE PEUTZ-JEGHERS SYNDROME is a familial disease characterized by mucocutaneous melanin pigmentation and by intestinal polyposis.^{3,4} The intestinal polyps are hamartomas and are distributed throughout the gastrointestinal tract. Intussusception of the small intestine is the most common complication present with the polyps of the small intestine.¹

This is a report of a prophylactic small-intestinal plication performed on a patient with Peutz-Jeghers syndrome in whom previous intussusceptions had necessitated several bowel resections.

Report of a Case

A 48-year-old black man with Peutz-Jeghers syndrome was admitted electively to the Georgetown University Surgical Division, D. C. General Hospital in September 1977. Past history included frequent episodes of cramping abdominal pain, attributed to self-reducing intussusceptions, interspersed between asymptomatic periods. Meals were often closely followed by attacks of abdominal pain. The frequency and severity of the attacks seemed to be increasing. Stools had been intermittently guaiac-positive. The patient was taking vitamin B₁₂ and iron supplementation. On three occasions nonreducing intussusceptions had necessitated bowel resections because of vascular compromise (Table 1). Since his most recent operation he had lost 11 pounds in weight. He was not anemic.

In an attempt to prevent future intussusceptions and additional small-intestinal resections, the following operation was performed. After standard mechanical and antibiotic bowel preparation, the abdomen was opened through the previous midline incision. The peritoneal cavity was essentially free of adhesions. Dozens of polyps of widely varying sizes could be palpated throughout the small intestine. An enterotomy was performed in the proximal jejunum and a Baker tube was advanced to the remaining portion of the right colon. With the tube as a splint, adjacent loops of small bowel were then sutured together along their antimesenteric borders with simple seromuscular sutures of 000 silk (Fig. 1). Acute angulation of the bowel was avoided by beginning and ending 3 cm from each turn.

The postoperative course was uneventful. Diet was gradually advanced and tolerated. The Baker tube was removed on the fourteenth postoperative day.

The patient was followed at regular intervals in the Surgery Clinic. He had no further bouts of cramping abdominal pain and

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regained approximately 8½ pounds in weight. Stools remained guaiac-negative. X-ray and endoscopic gastrointestinal surveillance were planned. Eight months after operation he complained of chest pain and died suddenly at home. An autopsy could not be obtained.

Comments

Over 300 patients with Peutz-Jeghers syndrome have been described.² The clinical course is characterized by asymptomatic periods and complications such as intermittent cramping abdominal pain, intussusception, often leading to intestinal obstruction, and gastrointestinal hemorrhage, which is often occult.

The majority of intussusceptions resolve spontaneously but when they do not, the treatment is surgical. The intussusception is reduced and the offending polyp removed. Preservation of small-intestinal length is mandatory to avoid the short-gut syndrome. Ordinarily Peutz-Jeghers syndrome is treated only when symptomatic. In some patients, however, the risk of developing the short-gut syndrome may warrant a prophylactic approach.

TABLE 1. Intussusceptions Necessitating Surgical Treatment

Date	Location of Intussusception	Portion of Bowel Excised
January 1964	Jejunum	80 cm of jejunum
May 1973	Terminal ileum	9 cm of terminal ileum, ileocecal valve, and cecum
May 1977	Ileum	39 cm of ileum

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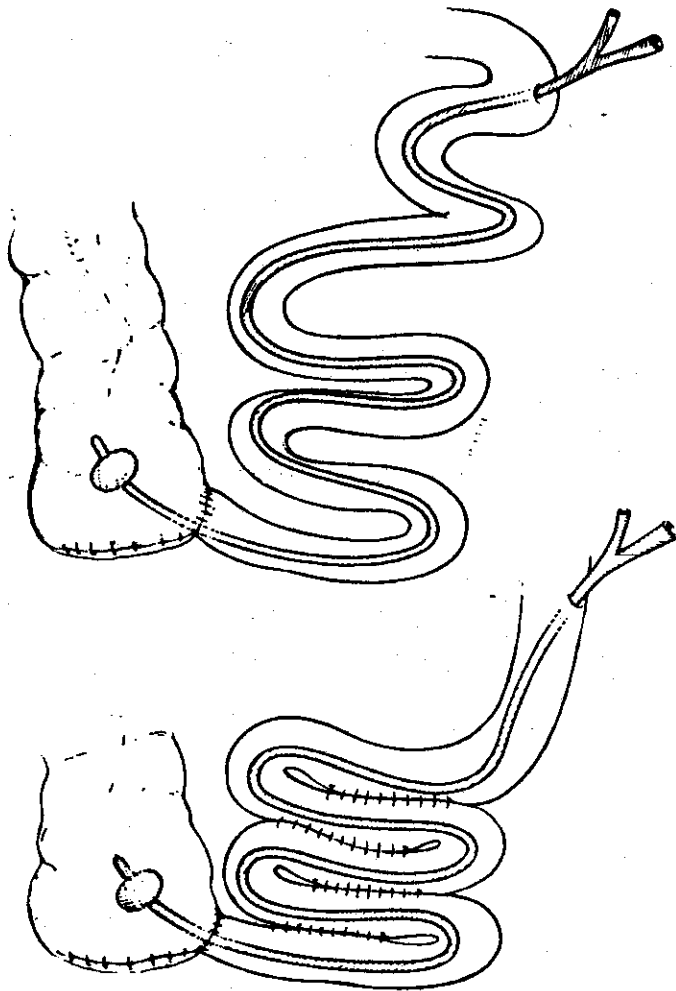


FIG. 1. Plication technique. Baker tube placed via enterotomy in proximal jejunum and threaded down to right colon. Anti-mesenteric borders of adjacent loops of bowel sutured together with seromuscular simple sutures of 000 silk.

Noble has described an operation for chronic adhesional intestinal obstruction in which adjacent loops of small intestine are sutured together.⁵ In this way adhesions, when reformed, may not compromise the lumen of the small intestine. In the patient described here, following a Noble-type plication there was an absence of colicky abdominal pain. Pain was notice-

ably absent after eating, and he gained weight. Quiescent periods have been described in Peutz-Jeghers syndrome but more characteristically the attacks become more frequent and remissions shorter.

Two other possible complications of Peutz-Jeghers syndrome deserve comment. First, the question of malignancy is not completely resolved. Reid found 14 documented cases of metastatic carcinoma in patients with Peutz-Jeghers syndrome with the location of the tumors primarily in the stomach and duodenum.⁶ Periodic x-ray surveillance of the stomach/duodenum and colon is indicated with polyps removed endoscopically. The risk of malignancy is not great enough in the small intestine to warrant prophylactic polypectomy. Second, anemia is a common and difficult problem. Endoscopy may help in visualizing a bleeding polyp. The plication technique described may make any subsequent small-intestinal surgical treatment more difficult.

Summary

Peutz-Jeghers syndrome is a familial disease characterized by mucocutaneous pigmentation and intestinal polyposis. Small-intestinal intussusception is a common complication. Preservation of small-intestinal length is essential to avoid the short-gut syndrome. A plication technique to prevent intussusception is described.

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