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The surgical risks of Peutz-Jeghers syndrome and the recommended timing of prophylactic enteroscopic intervention

Peutz-Jeghers syndrome (PJS) is a rare disorder characterised by intestinal hamartomatous polyps and an increased risk of enteroenteric intussusception that requires surgery. To explore the incidence and age distribution of surgical procedures in PJS, as well as the timing of enteroscopic interventions and their impact on the incidence of surgery, we conducted a web-based questionnaire survey. A total of 442 PJS cases were included in the analysis. Of these cases, 301(68.1%) experienced 506 surgeries before enteroscopic intervention. The mean age of the first surgery was (15.4±7.71) years, and 388 (76.68%) of the surgeries were performed on patients aged between 6 and 25 years. The annual growth rate of the cumulative incidence of surgery was higher for patients aged 6-25 years (3.4% per year) than for patients aged 1-5 years (1.0% per year) or 26-35 years (1.7% per year). The PJS patients were divided into the enteroscopic intervention group (EIG, n=374) and non-enteroscopic intervention group (nEIG, n=68) based on whether or not they accepted enteroscopic intervention. The mean age at the first enteroscopic intervention was (22.78±10.07) years. No statistically significant difference was observed between the two groups in terms of the incidence of surgical treatment (74.87% vs 67.65%, χ^2 =1.549, P=0.213). We concluded that the incidence of surgery in PJS is high and characterized by an early age at initial treatment. The age distribution curve for surgical treatment steeply ascends from the age of six, with the majority of procedures occurring between ages 6 and 25. Importantly, delayed enteroscopic intervention failed to reduce the overall incidence of surgical treatment in PJS patients. Consequently, early prophylactic enteroscopic intervention starting at age six is recommended.

Keywords

Peutz-Jeghers syndrome, enteroscopic intervention, intussusception, surgery

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Biography

SPECIALTY: endoscopic minimally invasive treatment of digestive diseases, especially good at endoscopic treatment of pancreatic and biliary diseases (ERCP +EUS); endoscopic treatment of early gastrointestinal cancer (ESD combined with laparoscopy to treat difficult gastrointestinal tumors. INNOVATION: the first EUS-guided iodine-125 seed implantation for the treatment of esophageal cancer and duodenal papilla cancer; the first endoscopic ESD for the treatment of recurrent duodenal papilla tumor after endoscopic resection. He was the first to carry out the experimental and clinical study of natural orifice translumenal endoscopic surgery (NOTES) in China, and proposed the concept of endoscopic surgery for the first time.

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