Introduction

Diagnosis of FAP is typically established on the basis of typical clinical features and APC (adenomatous polyposis coli) gene test. Teenage patients with FAP family should be followed by annual sigmoidoscopic or colonoscopic examinations depending on the polyp burden. Although surgery is a usual practice at an early time point in the evolution of FAP, it has been possible to defer surgery in these young patients, and all significant sized adenomas should be removed if surgery is delayed.1

The main goals in management of FAP patients are cancer prevention and to maintain good quality of life. Therefore, the timing for prophylactic cancer-preventive surgery can be arranged to be the least inconvenient for the patient and family and delayed if the patient is compliant and polyp burden are relatively low.1 In addition, preventive interventions such as chemoprevention and polypectomy can be applied with right indications. However, we need a more reasonable standard for assessing the severity of polyposis and applying it to specific intervention, and there have been some recent reports for staging system and extension of colonoscopic interventions.

Evaluation of polyposis: New staging system and stage-specific intervention

It is impossible to assess the number and size of adenomas accurately during colonoscopy in FAP patients. Nevertheless, polyp counts are critical in evaluating a patient’s response to intervention like chemopreventive drugs and polypectomy, and many clinicians use the extent of polyp burden for clinical judgment to determine the timing of colectomy. However, these evaluation and judgement are subjective and individual based, thus indicating a need for standardization. There has been no guidance for endoscopists and surgeons in determining the interventions including polypectomy and surgery. Especially, for the approval of new chemopreventive agents and other interventions, they must meet a higher standard of clinical benefit, based on reliable evaluation system.2

Recently, members of the International Society for Gastrointestinal Hereditary Tumors (InSiGHT) developed new staging system with 5 progressive stages based on adenoma number and size, InSiGHT polyposis staging system (IPSS), and stage-specific intervention system that would provide an acceptable measure of clin-
ical benefit in the endoscopic, surgical, and/or chemopreventive interventions considered appropriate to the adenoma burden. These systems can be applied to future clinical trials for interventions for FAP patients, enabling more reliable measures of patients’ response to nonsurgical treatments. Further validation of this scoring system is needed in the course of prospective clinical chemoprevention trials.

Endoscopic management of polyposis

Two major surgical procedures have been used for cancer prevention in FAP patients: colectomy with ileorectal anastomosis, and proctocolectomy with ileal pouch-anal anastomosis. In case of low polyp burden in rectum, the colectomy with ileorectal anastomosis is preferred with strict endoscopic surveillance of rectum. Many studies have shown that adenomas and occasionally even adenocarcinomas have been found in the ileo-anal pouch after restorative proctocolectomy. Therefore, endoscopic annual surveillance of the pouch and transitional anal zone is also essential. Endoscopic techniques are used in surveillance of patients after colectomy and to remove polyps from the rectal stump or ileal pouch. In recent report, endoscopic submucosal dissection (ESD) technique was reported as a useful technique to remove dense adenomas in rectal stump and anastomotic site.

The prophylactic cancer-preventive surgery can affect quality of life and some patients develop desmoid tumor after surgery. In addition, some fraction of polyposis patients with relatively lower polyp burden can be managed conservatively, with periodic multiple polypectomies without surgery. Especially, endoscopic polypectomy can be considered in the management of patients with attenuated FAP, which is characterized by fewer adenomas, a later age of onset for colorectal adenomas and cancer, and a decreased cancer risk compared with typical FAP.

As an alternative to prophylactic colorectal surgery, endoscopic management of FAP with strict indications might be considered with several potential advantages, including no risk associated with surgery, preserved normal bowel function, and no increased risk of desmoid tumor. Their use in FAP patients with preserved colon, as an alternative to colectomy, has not been applied by concerns about the feasibility of removing innumerable polyps and the risk of missing cancer. However, recent report showed that endoscopic management of FAP is feasible and safe in the medium term as an alternative to surgery in FAP patients who decline colectomy or who want to postpone colectomy by a few years. In this management, polyp clearance has to be achieved by multiple treatment sessions and removal of numerous polyps. However, further studies are needed to investigate the long term outcome, optimal intervals and proper indications.

Conclusions

In FAP patients, to develop more effective cancer preventive intervention and improve the quality of life, we need more objective evidence for staging system and stage-specific interventions to be applied to cancer preventive intervention trials and to provide an acceptable measure of clinical benefit.
References