FAMILIAL ADENOMATOUS POLYPOSIS COLI

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Familial adenomatous polyposis coli is genetically transmitted disease, as a Mendelian dominant trait with high degree of penetrance, characterized by multiple chromosomal studies. Neurosurgery 1982, 10: 445-449.


Operation for familial polyposis is to remove all potentially malignant mucosa, preserving the anus and avoiding the hazards of pelvic dissection (2,6).

Case Reports

Two brothers and one sister of familial polyposis coli were treated in our children's hospital. The age range of presentation was 4-7 years. Both male children presented as bleeding per rectum while the female was diagnosed incidentally on screening the family. However, a detailed enquiry revealed that she was suffering from intermittent dysentry and prolapse of rectal mucosa studded with polyps.

All the three patients underwent double contrast barium enema and colonoscopy which revealed multiple polyps of varying size throughout the colon, rectum and cecum but mostly in the rectum (Fig. 1). Histology of snared polyp through colonoscope showed adenomatous polyp. They were also subjected to upper gastrointestinal endoscopy to rule out the possibility of associated duodenal polyposis.

Total colectomy and endorectal ileoanal anastomosis was done in all the three patients. In the immediate post-operative period, all of them had diarrhea (10-15 stool/day) and perianal excoriation which subsided within 3-6 months of follow-up. No patient had incontinence of urine or stool. The first case developed cuff abscess which was drained successfully. The hospital stay was three weeks in the first case, and 2 weeks in the remaining two cases in order to reduce the frequency of stool before discharge. All the three patients had normal development and nutritional status comparable to that age group. Recent follow-up revealed frequency of stool in all the

![Fig. 1. Excised specimen of colon and rectum of case 3 showing multiple polyps of varying size scattered all over.](image)
three cases ranging between 2-4 per day without nocturnal soiling.

**Discussion**

Familial polyposis of colon is primarily a disease of the mucosa. The incidence of adenocarcinoma of the colon in these patients is quite high (1, 3, 11, 12). The malignant neoplasm can not only develop from polyp but also from nonpolypoid mucosa (1, 12). It can no longer, therefore, be assumed that eradication of rectal polyps in patients treated by total colectomy and ileorectal anastomosis provides an effective prophylaxis against carcinoma genesis. Two out of three cases in our series presented with bleeding per rectum. Similar observation had also been noted by others (2, 11, 12).

Considering the high possibility of malignant changes in mucosa, we believe that cancer prevention operation should remove nearly all potentially malignant mucosa. To obviate the necessity of permanent ileostomy, we propose total colectomy with endorectal mucosal stripping and ileoanal anastomosis as the operation of choice for familial adenomatous polyposis coli in children. It eliminates life long attention to the retained rectal segment and at the same time avoids permanent ileostomy. Though controversy exist about the timing of the operation and most of the literature advocates that it should be performed at puberty (13), we prefered to do it earlier because bowel adaptation is better and faster if operated early (2).

The only significant disadvantage of this operation is that it causes prolonged hospitalization and postoperative morbidity to the patient. However, a teenage patient is often psychologically not prepared to accept a permanent ileostomy, as marriage and participation in sports are significant considerations in future. Considering this social aspects, it is quite justified to have the discomforts of prolonged hospitalization in the post operative period with infrequent follow up, rather than to subject these cases for long term regular follow up till adulthood for detection of malignancies, which is not practicable in our socio-economic status. Although ideal and still a standard practice, long term follow up is not practical in our set up. Neither the incidence of nocturnal incontinence nor the association of upper gastrointestinal polyposis or any other associated malformation were encountered in our series as reported in the literature (3, 14).

**REFERENCES**


