

6. Tijssen CC, Halprin MR, Endtz UP. Familial aspects of gliomas. *In: Familial Brain Tumors*. Eds. Tijssen CC, Halprin MR, Endtz LJ. Boston, Martinus Nighoff, 1982. pp 411-443.
7. Maroun FB, Jacob JC, Henegahan WD, Mangan MA. Familial intracranial gliomas. *Surgical Neurology*, 1984, 22: 76-78.
8. Chaddock WN, Netsky MG. Familial gliomas: Report of four families with chromosomal studies. *Neurosurgery* 1982, 10: 445-449.
9. Hardman PD, Bell J, Whittle IR, Groeger A. Familial glioma: Report of glioblastoma in identical twins and oligoastrocytoma in siblings. *Br J Neurosurgery* 1989, 3: 709-715.
10. Prasad VSSV, Murthy JMK, Reddy R. Familial Intracranial gliomas. First Report of a family from India. *Neurology India* 1992, 40: 49-55.

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

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adenomatous polyps in colon and rectum. Less than 12% of these patients are prone to develop adeno-carcinoma upto 20 years of the age in the rectal stump after the ileo rectal anastomosis(1,2). The youngest patient encountered in literature was 8 years old(3). The best way of screening them is by mapping the long arm of chromosome number 5(4).

Coffey(5) was the first person to perform the total proctocolectomy and ileostomy for the treatment of this disease while Ravitch *et al.*(6) performed ileoanal anastomosis combined with total proctocolectomy.

The other mode of treating these patients are subtotal colectomy with ileorectal anastomosis alongwith regular follow up with proctoscopy for development of cancer in retained rectal mucosa(3,7). Recently, Stevan *et al.* (8) utilized laproscope for total proctocolectomy as well as ileoanal anastomosis while others performed ileopouch anal anastomosis alongwith total proctocolectomy(9,10). Thus the ideal op-

eration 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 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 dysentery 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 gastro-intestinal endoscopy to rule out the possibility of associated duodenal polyposis.

Total colectomy and endorectal ileo-anal 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

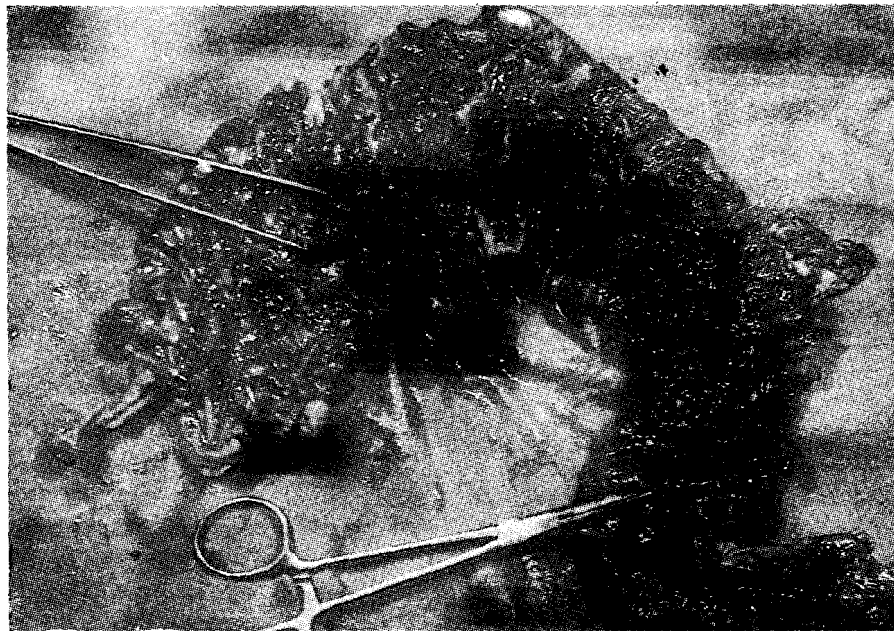


Fig. 1. Excised specimen of colon and rectum of case 3 showing multiple polyps of varying size scattered all over.

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 are quite high(1-3,11,12). The malignant neoplasm can not only develop from polyp but also from nonpolypoid mucosa(11,12). 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 preferred 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 par-

ticipation 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

1. Bulow F. Clinical features of familial polyposis coli. Result of the Danish polyposis register. *Dis Colon Rectum* 1986,29*102-107.
2. Soper RT. Gastrointestinal neoplasms. *In: Pediatric Surgery*. Eds Ashcraft KW, Holder TM. Philadelphia, W.B. Saunders Company, 1993, pp 453-469.
3. Schuchardt WA, Ponsky JL. Familial polyposis and Gardner's syndrome. *Surg Gyne Obst*, 1979,148: 97-103.
4. Bodmer WF, Baily CJ, Boderner J. Localization of the gene for familial adenomatous polyposis on chromosome 5. *Nature* 1987, 328: 614-616.
5. Coffey RC. Colon Polyposis with engrafted malignancy. *Ann Surg*, 1926, 93: 364-369.
6. Ravitch MM. Anal ileostomy with sphincter preservation in patient requiring total colectomy for benign conditions. *Surgery* 1948, 24: 170-187.
7. Coleman ST, Eckert C. Preservation of rectum in familial polyposis of colon and rectum. *Arch Surg* 1956,73: 635-644.

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8. Steven DW, Olaf J, Jauan JM, David GJ. Laproscopic total abdominal colectomy. A prospective trial. *Dis Colon Rectum* 1992, 35: 651-655.
 9. Peter MS, Wynl, Peter JH, David J. One stage restorative proctocolectomy without temporary defuncting ileotomy. *Dis Colon Rectum* 1992, 20: 582-588.
 10. Othon W, Hasan FH, David JS, *et al.* Ileopouch anastomosis. *Dis Colon Rectum* 1991,34:805-809.
 11. Santulli TV, Schullinger JN. Polypoid diseases of the gastrointestinal tract. *In: Pediatric Surgery.* Ed. Welch KJ, Randolph JG, Ravitch NM, O'Neill JA, Rowe MI. Chicago, Year Book Medical Publisher Int, 1986, pp 932-944.
 12. Randal WB, Wade SS. The adenomatous polyp and the hereditary polyposis syndrome. *Gastroenterol Clin North* 1988, 17: 657-678.
 13. Imbembo AL, Fitz Patrick JL. Benign neoplasm of the colon, including vascular malformations. *In: Textbook of Surgery: The Biological Basis of Modern Surgical Practice.* Ed Sabiston DC. Philadelphia, W.B. Saunders Company, 1991, pp 924-925.
 14. Leaper DJ. Tumors of the colon. *In: Maingot's Abdominal Operation,* Eds Schwertz SJ, Elto H. London, Prentice-Hall International Inc, 1990, pp 1033-1047.
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