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# Restorative Proctocolectomy with Handsewn Ileal J Pouch–anal Anastomosis and Mucosectomy for a Patient with Familial Adenomatous Polyposis

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Restorative proctocolectomy with ileal pouch–anal anastomosis is a common method of treatment for patients with ulcerative colitis and familial adenomatous polyposis. Since originally proposed in 1978, a few modifications of this operation have been implemented. In Lithuania this is still a rare procedure. We report our first prophylactic restorative proctocolectomy with mucosal proctectomy and hand sewn ileal J pouch–anal anastomosis for a 27-year-old female with familial adenomatous polyposis. Both stages of her operative treatment have been uncomplicated; after ileostomy closure and returning to normal diet her immediate bowel function was 5 stools per day and 1 per night. Her continence was normal. The necessity of not leaving any colorectal mucosa behind during such prophylactic procedures for polyposis patients is stressed, therefore mucosal proctectomy and handsewn ileal pouch–anal anastomosis is recommended.

**Key words:** familial adenomatous polyposis, ileal pouch–anal anastomosis, restorative proctocolectomy

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Familial adenomatous polyposis (FAP) is inherited in an autosomal dominant fashion disease, which due to a defect on chromosome 5 results in a development of up to a several thousand of colorectal polyps and a number of extracolonic manifestations. All untreated patients develop colorectal cancer. The only reliable treatment of FAP today is surgical, and operative procedures for prophylactic surgery are subtotal colectomy with ileorectostomy and restorative proctocolectomy, the latter being the main method of treatment (1). For a few suitable cases, subtotal colectomy with caecorectostomy has been proposed (2). Total proctocolectomy with permanent ileostomy for prophylactic purposes now has mostly a historical meaning.

A number of medications have been demonstrated to have an effect on colorectal adenomas in FAP: ascorbic acid has shown to have a temporary effect of reduction of adenoma growth (3); non-steroid antiinflammatory drugs, especially sulindac (Clinoril), have been proven to effect colorectal polyps as well (4, 5); at the moment, European trial is still conducted to evaluate a possible effect of resistant starch and/or aspirin on FAP patients with intact

colons (6). To date, none of the medical options are able to substitute or even postpone surgery, even though they might be of help in different clinical situations.

Restorative proctocolectomy with S-shaped ileal reservoir–anal anastomosis was first described by Parks AG in 1978 (7). Only the modification of Utsunomiya (8) has been used so far in the few procedures of this type done in Lithuania (1, 9); mucosal proctectomy has not been performed. Recently Kartheuser (10) has again very illustratively demonstrated the safety of mucosal proctectomy and hand-sewn ileal pouch–anal anastomosis, stressing the importance of mucosectomy for FAP patients. To the best of our knowledge, this is the first restorative proctocolectomy with handsewn ileal J pouch–anal anastomosis and mucosal proctectomy performed in Lithuania.

## CASE REPORT

Patient D. R., age 27, female, was referred for prophylactic surgery from another hospital where familial adenomatous polyposis had been diagnosed. On

admission (03 11 1999) she complained of diarrhoea, general weakness and occasional mild abdominal pains. Duration of these symptoms was 2 years. Previously she had never had colorectal examination and was not aware of familial predisposition to colorectal cancer. Her mother had been operated on for metastatic rectal cancer one year ago, and Hartman's operation had been performed.

Patient D. R. was reexamined. Upper GI endoscopy revealed 15 to 20 polyps 2 to 3 mm in diameter in the duodenal bulb and descending duodenum. Biopsy was taken, and tubular adenomas were found. Orthopantomography showed one 4 mm large osteoma in the mandibulae. On ophthalmoscopy, seven loci of congenital hypertrophy of retinal pigment epithelium were found in the right eye and 13 in the left eye. Proctoscopy showed appr. 50 polyps (Fig. 1) in the rectum from 4 to 8 mm in diameter; histology: tubular adenomas. As colonoscopy had been performed a month ago and no malignancy had been found, no further examinations were made.

The patient has 2 sisters and a brother, none of them yet screened. Her third sister died from pancreatic cancer 4 years ago at the age of 22. Prior to palliative excision of pancreatic tumor, her colonic barium enema examination was normal.

Patient D. R. was proposed prophylactic operation – restorative proctocolectomy. She agreed.

The operation was performed on the 8<sup>th</sup> of November 1999. Restorative proctocolectomy was done, fully mobilizing and transecting the rectum appr. 4 cm above the dentate line. While mobilizing the right colon, blood supply was ligated close to the bowel wall, thus not sacrificing the marginal branch from the right colic to the ileocolic artery. To gain mobility for the pouch to reach the anus, the ileocolic artery and the end of the lower mesenteric artery were ligated. A 20 cm long ileal J pouch was performed with an Ethicon linear cutting stapler.



Fig. 1. Endoscopic view on the rectum with multiple polyps seen

Through the anal approach, rectal stump has been everted (Fig. 2) and mucosectomy performed, using 20 ml of 1: 20000 diluted adrenaline solution for submucosal injection. After mucosectomy was completed up to the dentate line, the rectal stump was returned and the anal channel exposed with separate stitches. While advancing the ileal pouch through the anus with an Alice clamp (Fig. 3), a two-layer 3.0 vicryl ileal J pouch–anal anastomosis was performed at the level of the dentate line. A covering Turnbull loop ileostomy was used. Postoperative specimen revealed appr. 2000 polyps large 0.2–1.2 cm in the whole colorectum (Fig. 4), histologically–tubular adenomas with moderate and severe dysplasia. The postoperative course was uneventful.

Three months later she was admitted for the ileostomy occlusion. The preoperative pouchogram was normal (Fig. 5). On February 9, 2000 ileostomy occlusion was performed. The postoperative course was uneventful. Immediate bowel function after retur-



Fig. 2. Rectal stump is everted through the anal approach and mucosal proctectomy is going to be performed



Fig. 3. Separate stitches are applied from the anal channel (and internal sphincter) to perianal skin, thus exposing the dentate line and allowing safe handsewn ileal J pouch–anal anastomosis

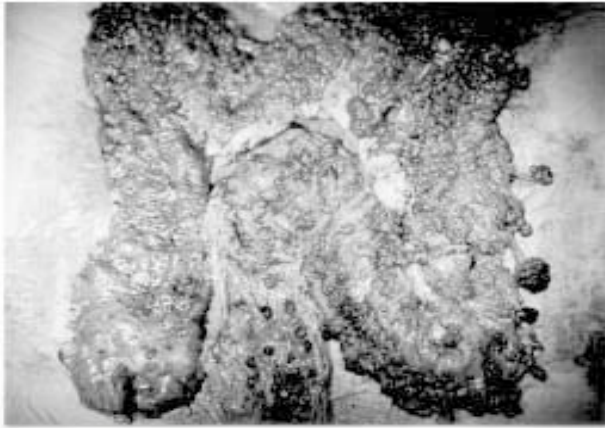


Fig. 4. Specimen of the large bowel after proctocolectomy; a total of appr. 2000 polyps have been counted



Fig. 5. "Pouchogram" – barium enema examination – prior to ileostomy closure, showing normal ileal pelvic pouch

ning to normal diet was 5 stools per day and 1 per night with absolutely normal continence.

**Discussion.** Various types of ileal reservoirs have been proposed: those earlier mentioned S- and J-shaped were followed by W pouch (11), which aims to combine excellent evacuation characteristics of J pouch with greater compliance and capacity which might lower the evacuation frequency. The H-shaped reservoir (12) has not been widely spread. Some authors were able to demonstrate a relation between the pouch volume and frequency of stools (13–15), which would make S and W pouches superior to J pouch, but this relation is not that much straightforward. We chose the J-shaped pouch for our first 10 restorative proctocolectomies because of its widespread recognition and simplicity in construction rather than because of any theoretical consideration. In this particular case, the most important detail was anal mucosectomy, for which we used an eversion technique offering us very good control of the completeness of mucosal removal up to the dentate

line. Even though mucosal proctectomy and further handsewn ileal pouch–anal anastomosis probably exposes internal anal sphincter to damage more than stapling (single or double) technique, it is reasonable to agree that this effect might be associated with long-term sphincter recovery and favourable outcome (16, 17). Several randomized trials have not found stapled technique to be of advantage in terms of function (18, 19), though eversion technique with handsewn anastomosis has been documented as inferior to stapled (20); on the other hand, a certain risk of increasing the number of septic complications in ulcerative colitis patients has been demonstrated with handsewn pouch–anal anastomosis (21). It would be reasonable to support those authors who have pointed out that stapled technique has a great disadvantage of leaving some of the disease behind (10, 18), which might be especially confusing in polyposis patients. The anal transitional zone, a subject for long-lasting speculations, as shown by a recently implemented new technique of Alcian blue staining and computerized histology mapping (22), seems to be much shorter than previously believed. That might affect decision making in restorative proctocolectomy in ulcerative colitis patients, too. Stapled technique might have another disadvantage as well – greater risk of stenosis of the pouch anal anastomosis (23). Authors of this article do believe that good mobilization of the pouch down to the pelvis by alternative ligation of the vasculature and preservation of the branch from right colic artery and tension-free anastomosis could be the key element in avoiding the stricture, regardless of the anastomotic technique used.

**Conclusion.** It is very important not to leave any colorectal mucosa behind during restorative proctocolectomy for familial adenomatous polyposis patients, therefore mucosal proctectomy and handsewn ileal pouch–anal anastomosis is recommended.

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**REKONSTRUKCINĖ PROKTOKOLEKTOMIJA SU GLEIVINĖS PROKTEKTOMIJA BEI RANKINĖ KLUBINĖS ŽARNOS REZERVUARO ANALINĖ JUNGTIŠ LIGONEI, SERGANČIAI ŠEIMINE ADENOMINE POLIPOZE**

**S a n t r a u k a**

Rekonstrukcinė proktokolektomija ir klubinės žarnos rezervuaro ir analinė jungtis yra pripažintos ligonių, sergančių šeimine adenomine polipoze ir nespecifiniu opiniu kolitu, chirurginio gydymo metodas. Jis buvo pasiūlytas 1978 m. Šiuo metu yra keletas originalios operacijos modifikacijų. Lietuvoje tokio tipo operacijos dar retos. Aprašoma pirmoji Lietuvoje rekonstrukcinė proktokolektomija su gleivinės proktektomija bei rankine klubinės žarnos rezervuaro–analine jungtimi, kuri buvo atlikta dėl vėžio profilaktikos 27 metų ligonei, sergančiai šeimine adenomine polipoze. Abu operacijos etapai buvo nekomplikuoti; likvidavus dirbtinę klubinės žarnos išangę, ligonės tuštinimosi dažnis buvo 5 kartai dieną ir 1 kartas naktį, kontinencija normali. Pabrėžiama būtinybė nepalikti nė trupučio storosios žarnos gleivinės atliekant profilaktines šio tipo operacijas sergantiesiems šeimine adenomine polipoze, todėl rekomenduojama gleivinės proktektomija bei rankinė klubinės žarnos rezervuaro ir analinė jungtis.