FAPA – Familial Adenomatous Polyposis Association
The Belgian Polyposis Registry

A. Kartheuser[1,2], S. Laurent[1,3], D. Franchimont[1,4], E. Van Cutsem[1,5], on behalf of the FAPA[3]

(1) Belgian Polyposis Project, Familial Adenomatous Polyposis Association (FAPA), Brussels ; (2) General Secretary of the FAPA, Colorectal Surgery Unit, Department of Abdominal Surgery and Transplantation, Cliniques Universitaires St-Luc, Brussels ; (3) Treasurer of the FAPA, Digestive Oncology, Ghent University Hospital, Gent ; (4) Vice-President of the FAPA, Gastroenterology Department, Erasme University Hospital, Brussels ; (5) President of the FAPA, Digestive Oncology Unit, University Hospital Gasthuisberg, Leuven.

Familial adenomatous polyposis (FAP) is an autosomal dominant disease that affects about 1 in 10 000 to 1 in 8 000 individuals. The underlying genetic defect is a mutation in the APC (Adenomatous Polyposis Coli) gene. If left untreated, this condition inevitably leads to colorectal cancer and a nationwide and systematic screening for asymptomatic gene carriers is therefore of paramount importance.

In May 1993, the Belgian "Familial Adenomatous Polyposis Association" (FAPA) was founded during a gathering of representatives of the seven Belgian universities of all disciplines involved in FAP.

The objectives of the FAPA are the following :

- to inform patients and relatives about FAP
- to stimulate informal contacts between patients via a Polyposis Contact Group
- to set up and run the Belgian Polyposis Registry including all families with FAP
- to support physicians in tracing families and guaranteeing regular screenings and follow-ups
- to contribute to national and international scientific research.

The main aim of a national registry is the surveillance of families with FAP in order to reduce the incidence of colorectal cancer from 65% to less than 5%. Following the example of the first registry in 1925 by Lockhart-Mummery at St Mark's Hospital in London, several national registries are now established in Europe and all around the world.

In Belgium, patient registration started in September 1995 thanks to the FAPA. Fifteen years later, the registration of affected FAP patients has been a great success, with 529 patients belonging to 237 families of 77 collaborating hospitals (Flanders, 55, Brussels, 11 and Wallonia, 11). More than 234 medical doctors have participated to this registration effort.

These figures compare favourably with other European registries such as those of the UK, The Netherlands, Denmark or Finland. For instance, for a population of 5 126 400, the Danish Polyposis Registry, established in 1971, included in 1996, i.e 25 years later, 321 histologically verified polyposis patients in 123 families (1). Moreover, except for the Italian registry led by L. Bertario (604 patients), all other European registries have fewer than 450 FAP patients (2).

For its 15th anniversary, the results of the Belgian Polyposis registry have been presented at national and international meetings (3,4,5).

For this special issue of Acta Gastroenterologica Belgica, the gastro-enterological, genetic, surgical and psychological issues of FAP have been reviewed.

In their gastro-enterological aspect review, S. Laurent et al. recall the different clinical presentations of the disease with special attention to extra-colonic manifestations such as congenital hyperthrophy of the retinal pigment epithelium (CHRPE), upper GI tract polyps and cancer, desmoid tumours or thyroid cancer. Different variants of FAP are also revisited, such as the Gardner Syndrome, Turcot Syndrome, Attenuated FAP (AFAP) and MutYH-associated polyposis (MAP). Screening and surveillance programmes as well as medical treatment and prevention are also proposed.

The genetic background of FAP and MAP is clearly explained in the paper by K. Claes *et al*. Beside the description of the underlying genetic defects, important clinical and practical implications of molecular screening and phenotype-genotype correlations are pointed out. The sensitive issue of attitude toward predictive and prenatal genetic testing is also raised.

After all these years of clinical studies and publications about prophylactic surgery for FAP, the choice between total proctocolectomy (TPC) with ileal pouch-anal anastomosis (IPAA) and total colectomy (TC) with ileo-rectostomy (IR) is still a difficult one to make.

The role of total colectomy with ileo-rectal anastomosis in the treatment strategy of FAP is discussed by A. Wolthuis *et al*. in terms of "low" and "high" risk rectum with regard to rectal cancer risk, fertility, desmoid tumour phenotype, functional results and quality of life, which have all to be taken in account.

Correspondence to : Prof. A. Kartheuser, M.D., Ph.D., M.Sc., General Secretary of the FAPA, Colorectal Surgery Unit, Department of Abdominal Surgery and Transplantation, Cliniques Universitaires St-Luc, 1200 Brussels. E-mail : alex.kartheuser@uclouvain.be

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The restorative coloproctectomy (RCP) with ileal pouch-anal anastomosis (IPAA) is advocated by D. Léonard *et al*. When considering IPAA, new technical issues have to be added to the discussion, such as double stapling technique, mesenteric lengthening, omission of temporary protective stoma and laparoscopic approach.

And last but not least, E. Claes and colleagues extensively address the problem of psychological implications of living with FAP, including response to diagnosis and distress, illness representation – perceived seriousness and perceived control –, knowledge and information needs, prophylactic surgery and surveillance behaviours, genetic testing of adults and reproductive decision-making, genetic testing of children and finally, family communication.

All these extensive reviews of the different aspects of the FAP syndrome underline once again the overwhelming importance of nationwide screening and surveillance programmes and stress the need for complete central registration of every affected and "at risk" FAP patient in Belgium.

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