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Original Article

Surgical complications and metachronous rectal cancer risk in patients with classic familial adenomatous polyposis

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ABSTRACT

Introduction: Familial adenomatous polyposis (FAP), an autosomal dominant disease characterized by development of numerous adenomatous polyps in the colon and rectum, is caused by germline mutations in the *Adenomatous Polyposis Coli* (*APC*) gene.
Methods: To determine the surgical morbidity in patients with classical familial adenomatous polyposis and determine the incidence of metachronous colorectal cancer (CRC) in those undergoing total colectomy (TC) with ileorectal anastomosis or restorative total proctocolectomy (TPC) and ileal pouch anal anastomosis. We analyzed patients with familial adenomatous polyposis who received treatment and regular follow-up at the A.C. Camargo Cancer Center from 1994 to 2013.
Results: Operative complications occurred in 22 patients (34.3%), 16 (25%) being early complications and 8 (12.5%) late complications. No mortality occurred as a result of postoperative complications. The incidence of metachronous rectal cancer after total proctocolectomy was 2.3% and after total colectomy 18.18% ($p = 0.044$).
Conclusions: In order to provide better quality of life for individuals with familial adenomatous polyposis, total colectomy is commonly offered, as this simple technique is traditionally associated with lower rates of postoperative complications and better functional outcomes. However, it has become a less attractive technique in patients with familial adenomatous polyposis in its classical or diffuse form, since it has a significantly higher probability of metachronous rectal cancer.

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Complicações cirúrgicas e risco de câncer retal metacrônico em pacientes com polipose adenomatosa familiar clássica

Palavras chave:
Polipose adenomatosa familiar
Câncer colorretal
Colectomia total
Pólipos adenomatosos
Proctocolectomia restaurativa

R E S U M O

Introdução: Polipose adenomatosa familiar (PAF), uma doença autossômica dominante caracterizada pela formação de numerosos pólipos adenomatosos no cólon e reto, é causada por mutações da linha germinativa no gene da polipose adenomatosa do cólon (PAC).
Métodos: Para determinar a morbidade cirúrgica em pacientes com PAF clássica e determinar a incidência de câncer colorretal (CCR) metacrônico naqueles pacientes submetidos à colectomia total (CT) com anastomose íleo-retal ou submetidos à proctocolectomia restaurativa (PCT) e anastomose bolsa ileal-anal, foram analisados pacientes com PAF que foram tratados e tiveram acompanhamento periódico no A. C. Camargo Cancer Center de 1994 até 2013.
Resultados: Ocorreram complicações cirúrgicas em 22 pacientes (34,3%); 16 (25%) tiveram complicações precoces e 8 (12,5%) complicações tardias. Não houve mortes como resultado de complicações pós-operatórias. A incidência de câncer de reto metacrônico após PCT foi de 2,3% e após CT foi de 18,18% (p = 0,044).
Conclusões: A fim de proporcionar melhor qualidade de vida para os pacientes com PAF, CT é comumente oferecida, pois esta técnica simples está tradicionalmente associada com menores percentuais de complicações pós-operatórias e melhores resultados funcionais. No entanto, CT se tornou uma técnica menos atraente em pacientes com PAF em sua forma clássica ou difusa, uma vez que traz consigo uma probabilidade significativamente maior de câncer retal metacrônico.

Introduction

Familial adenomatous polyposis (FAP), an autosomal dominant disease characterized by the development of hundreds to thousands of adenomatous polyps in the colon and rectum, is caused by germline mutations in the *Adenomatous Polyposis Coli* (*APC*) gene, which is in chromosomal region 5q21-22.[1,2] This mutation is present in 1 in 10,000 live births, and accounts for 1% of the cases of colorectal cancer (CRC).[3]

In classical polyposis polyps, mainly in the distal colon (rectosigmoid), often develop during childhood and increase in size and number in adolescence, when they characteristically develop throughout the colon. Half of these patients develop adenomas by the age of 15 years and 95% by the age of 35.[1] Genomic penetrance is approximately 100%. The median age of diagnosis of CRC is 39 years; however, 7% of patients develop CRC before the age of 21.

Essential steps in the management of patients with FAP include early diagnosis of affected individuals, performance of prophylactic colectomy when appropriate, genetic counseling, recognition of various extracolonic manifestations and adequate postoperative follow-up.[1]

Surgery is the most effective means of preventing CRC, mainly in the form of total colectomy with ileorectal anastomosis (IRA) or total proctocolectomy (TPC) followed by ileal pouch and ileoanal anastomosis.[4] Preservation of the rectum is associated with better functional outcomes and less morbidity, but carries a risk of metachronous tumor in the stump remnant.[5,6] Decisions concerning the best procedure for each patient should be based on factors such as age, location and number of polyps, location of genetic mutation and patient acceptance of undergoing regular postoperative follow-up.

Until 1980, prophylactic restorative proctocolectomy and ileal pouch anal anastomosis was the procedure of choice for the treatment of FAP. Thereafter, TPC followed by ileal pouch and ileoanal anastomosis became the gold standard for treatment of the classic or diffuse form of this disease.[3]

Our objectives were to assess surgical morbidity in patients with classical FAP and ascertain the risk of metachronous CRC in the anorectal region in patients who had undergone TC with IRA or TPC with ileal pouch, these being the most commonly performed procedures in most institutions.

Patients and methods

Relevant data of patients with FAP who were added to the hereditary CRC registry of the A.C. Camargo Cancer Center (HACC) from 1994 to 2013 were retrospectively analyzed. The diagnoses of FAP had been established by clinical history and colonoscopy with histological analysis of some resected polyps.

The study analyzed 86 patients (from 34 families) with FAP who had received genetic counseling and regular follow-up treatment. Eighty-two of these patients underwent surgical treatment. Studied variables were epidemiological and surgical procedure was performed. The aim being to ascertain the incidence of metachronous rectal cancer after TC or TPC, ten patients who had undergone noncurative surgical treatments or procedures other than TC and TPC were excluded,