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ORIGINAL ARTICLE

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Prophylactic pancreaticoduodenectomy for premalignant duodenal polyposis in familial adenomatous polyposis

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Abstract The frequency of duodenal adenomas in patients with, familial adenomatous polyposis is high. Duodenal adenoma has malignant potential, and duodenal adenocarcinoma is one of the main causes of death in patients who have had previous proctocolectomy. A conservative approach to the treatment of duodenal adenomas (nonsteroidal anti-inflammatory drugs, endoscopy, polypectomy through duodenotomy) is inefficient and unsafe. When invasive cancer occurs in duodenal adenomas, the result of surgery is poor. We have performed prophylactic pancreaticoduodenal resection (PDR) for nonmalignant severe duodenal polyposis in five patients since 1991. No operative mortality was observed. One patient developed a pancreatic fistula which was successfully managed by medical treatment. The mean follow-up was 35 months. All five patients are still alive and have a good functional outcome. Prophylactic PDR may be indicated in familial adenomatous polyposis when duodenal polyposis is severe. Stages III and IV of Spigelman's classification, periampullary adenoma, age above 40, and family history of duodenal cancer are factors that may lead to the decision to perform prophylactic PDR.

Key words Familial adenomatous polyposis · Duodenal adenomas · Prophylactic pancreaticoduodenal resection

Résumé L'incidence d'adénomes duodénaux chez des patients atteints de polypose familiale (F. A. P.) est élevéee. Les adénomes du duodénum ont un potentiel de malignité et les adénocarcinomes du duodénum sont une des causes principales de décès chez les patients qui avaient subi au préalable une procto-colectomie. Une approche conservatrice du traitement des adénomes duodénaux (médication anti-inflammatoire non stéroïdienne, endoscopie, polypectomie par duodénotomie) est insuffisante et peu sûre. Si un cancer invasif se développe dans un adénome du duodénum, le résultat de la chirurgie est mauvais. Nous avons réalisé des résections pancréatico-duodénales prophylactiques pour des polyposes duodénales sévères non malignes chez 5 patients depuis 1991. Aucune mortalité opératoire n'est à déplorer. Un patient a développé une fistule pancréatique qui a été traitée avec succès médicalement. Le follow-up moyen est de 35 mois. Les cinq patients sont toujours en vie et ont un bon résultat fonctionnel. En cas de F. A. P., la résection pancréaticoduodénale prophylactique peut être indiquée lorsqu'une polypose duodénale est présente. Les stades III et IV de la classification de Spigelman, un adénome péri-ampullaire, plus de 40 ans d'âge et une histoire familiale de cancer du duodénum sont des facteurs qui peuvent conduire à la décision de réaliser une résection pancréatico-duodénale prophylactique.

Introduction

Familial adenomatous polyposis (FAP) is a premalignant condition characterized by numerous colorectal adenomas. In addition, patients with FAP frequently develop upper gastrointestinal tract adenomas, particularly in the duodenum [1–13]. As with colorectal polyps, duodenal adenomas are premalignant lesions [14]. Upper gastrointestinal endoscopy is required to diagnose and treat duodenal lesions at early stage [4, 6, 8, 15–23], as once invasive cancer has developed, prognosis is poor [23].

We report five cases of prophylactic pancreaticoduodenal resection (PDR) performed in patients with FAP for nonmalignant duodenal polyposis.

Patients and methods

Five women with a past history of FAP have undergone PDR for duodenal polyposis since 1991. Patient characteristics are reported in Table 1.

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Table 1 Patient characteristics: age (years) at diagnosis of different stages of the disease and intestinal status when PDR was performed. (*IRA* Ileo-rectal anastomosis, *IAA* Ileo-anal anastomosis)

Patient no.	FAP	Duodenal disease	PDR	Intestinal status
1	16	38	38	Ileostomy
2	40	52	53	Ileostomy
3	19	27	27	IRA
4	21	38	38	IRA
5	28	33	36	IAA

Table 2 Duodenal polyposis: Spigelman's classification. (*Stage I* score 1–4, *Stage II* score 5, 6, *Stage III* score 7, 8, *Stage IV* score 9–12)

	1	2	3
Adenomas	1–4	5–20	>20
Size (mm)	1–4	5–10	>10
Histology	Tubulous	Tubulovillous	Villous
Dysplasia	Mild	Moderate	Severe

Duodenal polyposis was diagnosed by routine gastroduodenoscopy in three patients; another patient presented with a history of gastroesophageal reflux and another with an episode of cholangitis. Gastroduodenoscopy was performed with forward and side-viewing endoscopes. The endoscopic appearance of the papilla was noted, and biopsies were taken systematically from the polyps. The histopathological examination was performed by the same pathologist. To assess the severity of the duodenal polyposis the classification of Spigelman was used (Table 2), and endoscopic aspect of the papilla was noted (Table 3).

Before PDR the duodenum was inspected directly two patients by duodenotomy. In one of them enteroscopy was used to identify possible adenomas in the small intestine. Resection of the first jejunal loop was performed in four cases because of distal duodenal involvement. The reconstruction, according to Child, started with the pancreatic anastomosis. In all cases pancreatic tissue was friable, and the diameter of the Wirsung duct was small (2–3 mm) in all cases but one. Wirsungojejunostomy was performed in four cases and was intubated by a unexteriorised silastic catheter. Two anterior and posterior layers secured the pancreatic section to the jejunal serosa. The fifth patient, in whom no Wirsung duct was seen, underwent a terminolateral pancreato-jejunostomy. The common bile duct was normal in diameter except in one patient in whom it was enlarged. A terminolateral cholangio-jejunostomy was performed in all cases. The pylorus was retained in all patients, and digestive tract continuity was restored by a terminolateral duodeno-jejunostomy.

Results

Histopathological examination of the specimens confirmed the absence of cancer and revealed a more severe polyposis (as regards number of polyps, size, and dysplasia) than expected preoperatively. Four patients were classified stage IV postoperatively compared to only two preoperatively (Tables 3, 4).

No mortality was observed. The postoperative course was uneventful in three patients. In one it was complicated by a delay in bowel transit that required nasogastric suction for 16 days. A pancreatic fistula, with a daily output up to 500 ml, occurred in one patient; this was successfully managed by suction, inhibition of pancreatic secretion (Sandostatine for 23 days), and total parenteral nutrition. The five patients were discharged respectively on the 13th, 14th, 14th, 28th, and 66th postoperative days.

Late outcome

The mean follow-up of the five patients was 35 months (respectively 2, 19, 42, 42, and 69 months). All five patients are still alive and have undergone regular endoscopy of the upper digestive tract; this has also been performed of the retained rectum in two patients. After PDR no gastric adenomas were observed. On the other hand, the rectum displayed recurrent polyposis which was dealt with either by endoscopic electrocoagulation (in one patient) or by proctectomy and a J ileal pouch (in another).

Table 5 shows the functional digestive assessment, need for special died and weight before and after PDR.

Table 3 Duodenal polyposis (endoscopic and histological feature) and stage (Spigelman's classification)

Patient no.	Duodenal adenomas (*n*)	Size (mm)	Papilla aspect	Histology	Dysplasia	Stage
1	3	>10	Abnormal	Tubulovillous	Moderate	III
2	6	>10	?	Tubulovillous	Moderate	IV
3	3	>10	Abnormal	Tubulovillous	Moderate	III
4	3	>10	Abnormal	Tubulovillous	Moderate	III
5	>20	>10	Abnormal	Tubulovillous	Severe	IV

Table 4 Histopathological examination of the specimens and stage (Spigelman's classification)

Patient	Adenomas (*n*)	Max. size (mm)	Papilla aspect	Ampulla localization	Histology	Dysplasia	Stage
1	6	40	Tumor	Yes	Tubulovillous	Moderate	IV
2	3	18	Normal	No	Tubulovillous	Moderate	III
3	5	30	Tumor	Yes	Tubulovillous	Severe	IV
4	15	30	Tumor	Yes	Tubulovillous	Moderate	IV
5	>20	70	Tumor	Yes	Tubulovillous	Severe	IV

Table 5 Functional digestive assessment, requirements for special diet and weight, before and after PDR

Patient	Bowel movement (*n*/24 h)		Ileostomy output (l/24 h)		Special diet		Weight (kg)	
	Before	After	Before	After	Before	After	Before	After
1			<1	<1	No	No	51	51
2			<1	<1	No	No	65	58
3	1, 2	1, 2			No	Yes	66	69
4	1–3	1–3			No	No	44	45
5	4, 5	5, 6			No	No	50	44

Comments

Many rationales have been put forth for PDR in cases of duodenal polyposis in patients with FAP.

Duodenal polyposis is a precancerous condition

FAP is known to involve the upper digestive tract [1 – 14]. Recent studies [1, 4, 6, 7] have shown that nine of ten patients with FAP experience duodenal involvement during the course of the disorder. Gastric lesions consist of glandulocystic polyposis and are not at risk of cancer [3, 4, 6, 7, 10]. In Western countries the risk of gastric cancer is not higher in FAP patients than in the general population [6, 7, 20]. On the other hand, duodenal polyps are adenomas and consequently may advance to dysplasia and cancer [23]. The frequency of duodenal cancer in FAP is 2% [2, 20, 22]. The risk of cancer increases with age and is higher in cases of familial history of duodenal cancer [24]. The two main causes of death in patients who have undergone prophylactic proctocolectomy are currently desmoid tumors and duodenal cancer [15, 19].

When cancer arises in duodenal adenomas, the result of resection is poor [2, 22]. Beckwith et al. [22] report a mean a survival of 13 months in 4 patients (three radical pancreatico-duodenectomies and one palliative bypass).

Patients with duodenal involvement require regular endoscopic examination. The periampullary area is at risk of adenomas [6, 7]. Duodenoscopy with side-viewing endoscope is mandatory as well as systematic biopsies even in cases with normal macroscopic appearance [3, 16].

Conservative approach is inefficient or unsafe

Nonsteroidal anti-inflammatory drugs such as Sulindac have no role in treating severe duodenal polyposis [25, 26]. They may slow the evolution to malignancy [26], and any effect seems to be restricted to polyps less than 2 mm in diameter [25].

Endoscopy allows electrocoagulation of adenomas, but the size, site, and number of polyps limit its therapeutic application. Ampullary localization may risk biliary or pancreatic duct injury, and even if polyps are destroyed, electrocoagulation does not prevent recurrence. Photodynamic laser, a new endoscopic technique, is still under evaluation [27]. Endoscopic methods appear generally to be restricted to small, few, and easily accessible lesions.

Polypectomy through duodenotomy has the same limits as endoscopic treatment. Penna et al. [28] studied 12 patients and reported recurrence of duodenal polyposis in all patients after a mean follow-up of 13 months.

PDR is an available option

Pancreaticoduodenectomy is an aggressive surgical approach [29] that allows radical resection of the ampullary area. Although morbidity and mortality of PDR may be acceptable, this procedure is not to be undertaken lightly and should be restricted to patients with a high risk of cancer and with lesions preventing full resection by using conservative procedures. The risk of malignancy increases with severity of polyposis, as assessed by the Spigelman's classification in cases of periampullary lesions, with age, and with a familial history of duodenal cancer [16, 24]. Preoperative staging of duodenal involvement seems to be underestimated. In our series three patients in whom polyposis had been initially classified as stage III were reclassified as stage IV postoperatively (Tables 3, 4). Spigelman's classification should not be the only method for assessing severity. The site of polyps is also important since periampullary adenomas are at higher risk of cancer, and endoscopic resection is difficult.

In FAP biliary secretion has a promoting effect on adenomatosis, which involves mainly the periampullary area [30, 31]. Retaining the pylorus may be of relevance; pylorus preservation prevents biliary reflux and may avoid its harmful impact on gastric mucosa. Furthermore function of the stomach may be less impaired. In the present series the functional status of the digestive tract was not impaired after PDR. All patients regained their preoperative weight (Table 4), and no was malnutrition was observed. No patient developed diabetes or signs of pancreatic secretion impairment.

Conclusion

PDR in duodenal polyposis must not be a routine procedure but rather should be reserved for patients at high risk of cancer. This risk is related to the severity of polyposis

(stages III and IV of Spigelman's classification) and to the periampullary localization. Age above 40 years and family history of duodenal cancer are also to be taken into account and may indicate the need for PDR.

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