Original article

Pancreas-preserving total duodenectomy *versus* standard pancreatoduodenectomy for patients with familial adenomatous polyposis and polyps in the duodenum

S. M. M. de Castro[1], C. H. J. van Eijck[2], J. P. Rutten[3], C. H. Dejong[3], H. van Goor[4], O. R. C. Busch[1] and D. J. Gouma[1]

Departments of Surgery, [1]Academic Medical Centre, Amsterdam, [2]Erasmus Medical Centre, Rotterdam, [3]University Hospital Maastricht, Maastricht, and [4]University Medical Centre, St Radboud, Nijmegen, The Netherlands

Correspondence to: Dr D. J. Gouma, Academisch Medisch Centrum, Meibergdreef 9, 1105 AZ, Amsterdam, The Netherlands (e-mail: d.j.gouma@amc.nl)

Background: **Pancreas-preserving total duodenectomy (PPTD) was introduced as a replacement for pancreatoduodenectomy (PD) for familial adenomatous polyposis (FAP). This study analysed the results of PPTD in the Netherlands and reviewed the relevant literature.**

Methods: **All 26 patients who underwent PPTD for FAP in four centres in the Netherlands between January 2000 and January 2007 were compared with a group of 77 patients who had PD for ampulla of Vater adenocarcinoma at one centre during the same interval.**

Results: **Morbidity rates were similar after PPTD for FAP (16 patients, 62 per cent) and PD for ampulla of Vater adenocarcinoma (44 patients, 57 per cent) ($P = 0{\cdot}694$). One patient (4 per cent) died after PPTD and two (3 per cent) after PD. A review of the literature, including patients from the present study, found that 71 patients had PPTD, with postoperative morbidity in 36 (51 per cent) and one death (1 per cent). In publications containing a total of 94 patients who underwent PD for FAP, 43 (46 per cent) developed complications and three (3 per cent) died.**

Conclusion: **PPTD has similar short-term results to PD in terms of morbidity and mortality.**

Paper accepted 15 September 2008

Published online in Wiley InterScience (www.bjs.co.uk). **DOI:** 10.1002/bjs.6308

Introduction

Patients with familial adenomatous polyposis (FAP) develop multiple colonic adenomatous polyps. The disease is an autosomal dominant inherited condition caused by a germline mutation in the adenomatous polyposis coli (*APC*) gene. Without treatment it invariably progresses to colonic adenocarcinoma by the fourth decade. Treatment consists of a prophylactic or therapeutic proctocolectomy and reconstruction preferably with an ileal pouch–anal anastomosis[1].

After 10 years of follow-up about 94 per cent of patients also have duodenal polyps[2–4]. In contrast to colonic polyps, duodenal polyps do not invariably become malignant, but approximately 5 per cent progress to adenocarcinoma[5]. Overall, this translates into a 300-fold increased risk of patients with FAP developing duodenal adenocarcinoma compared with the general population[6]. Duodenal adenocarcinoma is currently the leading cause of death after proctocolectomy in these patients[7,8].

Prophylactic removal of duodenal polyps has now become a reasonable option as the morbidity and mortality rates associated with pancreatic and duodenal surgery have decreased in recent decades[9]. Two surgical procedures have traditionally been used: duodenotomy and local resection for limited disease and (pylorus-preserving) pancreatoduodenectomy (PD) for more extensive disease. Recently pancreas-preserving total duodenectomy (PPTD) has been introduced as an alternative. Although advocates of this procedure believe that it might result in definitive prevention of duodenal cancer with potentially lower morbidity and mortality rates than standard PD, a

The Editors are satisfied that all authors have contributed significantly to this publication

formal comparison of the two procedures has never been performed. The aim of the present study was to analyse the outcome after PPTD for FAP compared with that after PD for ampulla of Vater adenocarcinoma.

Methods

Patients who underwent PPTD between January 2000 and January 2007 in four centres (Academic Medical Centre (AMC) in Amsterdam, Erasmus Medical Centre in Rotterdam, University Hospital of Maastricht and University Medical Centre St Radboud in Nijmegen) throughout the Netherlands were included. Four patients from the Erasmus Medical Centre have been reported previously[10]. Patients who had PPTD were compared with a group that had PD for neoplasms of the ampulla of Vater at the AMC during the same interval. These patients have been described previously in a separate study[11]. All surgeons had extensive experience in pancreatic surgery.

Pancreas-preserving total duodenectomy

The procedure was performed through a right subcostal incision or median incision. After kocherization, transection of the pylorus or distal stomach was performed, depending on the extent of disease. Subsequently the duodenum was transected near the ligament of Treitz. A transection plane was created between the duodenum and the head of the pancreas. The small vessels were identified and clipped or ligated. The accessory pancreatic duct was identified and ligated, and circumferential stay sutures were placed through the common bile duct (CBD) and the pancreatic duct. The duodenum was removed and reconstruction performed by advancing the jejunum into the position previously occupied by the duodenum. Generally, the pancreatic and bile ducts were sutured together and the combined, newly formed distal pancreaticobiliary junction was anastomosed to the jejunum such that the papilla was removed, and the terminal portions of the CBD and pancreatic duct were exposed and anastomosed to an isolated loop of jejunum. Proton-pump inhibitors were administered only when a gastric resection was performed.

Pancreatoduodenectomy

PD, defined as *en bloc* total removal of the duodenum and pancreatic head, preferably with preservation of the pylorus, was performed as reported previously[12]. Reconstruction was performed by means of a retrocolic jejunal loop with an end-to-side pancreaticojejunostomy, hepaticojejunostomy, and gastrojejunostomy or duodenojejunostomy.

Outcome

All complications during the hospital stay or requiring readmission within 90 days after discharge were recorded. Complications were categorized as surgical (intra-abdominal abscess, wound infection, haemorrhage, anastomotic leakage and delayed gastric emptying) or general (urological, pulmonary and cardiac). The definitions of delayed gastric emptying, pancreatic leakage and postpancreatectomy haemorrhage recently proposed by the International Study Group of Pancreatic Surgery were used[13–15]. All patients who had PPTD were followed up annually by endoscopic surveillance. General practitioners were contacted for definitive follow-up.

Literature review

The medical databases Medline and Embase (1966–2007), and The Cochrane Library (1996–2007) were used to search for relevant studies. Search algorithms combined the medical subject heading (MeSH) terms and keywords. The 'related articles' feature of PubMed was also used. A manual search of the bibliographies of relevant papers was carried out to identify publications for possible inclusion. No unpublished data or abstracts were included.

Statistical analysis

Statistical analysis was performed using SPSS® statistical software (SPSS, Chicago, Illinois, USA). The Mann–Whitney *U* or Student's *t* test was used as appropriate to analyse differences in continuous data. Pearson's χ^2 test was used to examine differences between dichotomous groups. Fisher's exact test was used when a table had a cell with an expected frequency of less than 5. $P < 0{\cdot}050$ was considered statistically significant.

Results

Twenty-six patients with FAP had polyps in the duodenum and underwent PPTD (*Table 1*).The mean(s.d.) age at presentation was 29(9) years. All patients had a proctocolectomy followed by reconstruction with an ileal pouch–anal anastomosis (11 patients), ileorectal anastomosis (nine) or an ileostomy (six). Ten patients received chemopreventive therapy comprising sulindac 75–150 mg orally twice daily. Preoperative endoscopy and histological evaluation confirmed Spigelman III and Spigelman IV disease in one and 25 patients respectively.

Table 1 Characteristics of 26 patients who underwent pancreas-preserving total duodenectomy between 2000 and 2007

	No. of patients*
Sex ratio (M : F)	16 : 10
Mean(s.d.) age at colectomy (years)	29(9)
Procedure	
Ileal pouch–anal anastomosis	11
Ileorectal anastomosis	9
Ileostomy	6
Mean(s.d.) age at PPTD (years)	48(9)
Median (range) interval between duodenoscopy (months)	6 (2–36)
Chemoprevention	10
Preoperative Spigelman classification	
III	1
IV	25

*Unless indicated otherwise. PPTD, pancreas-preserving total duodenectomy.

Comparison with pancreatoduodenectomy for ampullary tumours

Characteristics and outcome of patients who had PPTD for FAP and those who had PD for ampullary tumours are summarized in *Table 2*. Those who had PPTD were significantly younger and all had undergone laparotomy previously. The pylorus was involved in 15 patients with FAP; a distal gastric resection was therefore performed and these patients received proton-pump inhibitors after surgery. The operating time was significantly longer for PPTD than for PD. Postoperative morbidity rates were similar and there were no significant differences in the type of postoperative complications. Mortality rates were also comparable.

Literature review

A review of the literature identified eight studies of PPTD for FAP (*Table 3*). A study by Chung and colleagues[24] was not included separately because the patients described in this study were also included in a later study at the same institution. A Dutch survey[22] included patients from a study by Soravia *et al.*[23] and so the latter was not considered. None of the patients from the present study was included in this Dutch survey. Overall, including the present study, a total of 71 patients who underwent PPTD have been reported. Morbidity was described in 36 patients (51 per cent) and there was one death (1 per cent). Seven studies were identified that described the results of PD for patients with FAP. Of 94 patients in total, 43 (46 per cent) developed complications and three (3 per cent) died (*Table 4*).

Discussion

This study has shown that PPTD used as a means of removal of polyps from the duodenum of patients with FAP has postoperative morbidity and mortality rates similar to those of PD for ampulla of Vater disease. These results are comparable with existing data.

Duodenal disease in patients with FAP is becoming increasingly important as a result of the increased life expectancy of these patients in surveillance programmes after prophylactic proctocolectomy together with an overall increase in the number of proctocolectomies performed[31,32]. Surveillance programmes aim to identify patients with severe polyposis of the duodenum before adenocarcinoma is present because the prognosis following malignant differentiation is poor[5,33].

Selection of patients for 'prophylactic' surgical intervention is difficult, as up to a third of patients with Spigelman IV duodenal disease progress to adenocarcinoma of the duodenum[34–36]. Patients with large ampullary lesions represent a particularly high-risk group. Another problem is that endoscopic surveillance is not sufficiently sensitive to detect adenocarcinoma harboured in villous neoplasms of the duodenum[11,37]. Similarly low diagnostic accuracy rates have been reported in patients with FAP[38,39].

Ideally, a prophylactic procedure should carry no risk of death and have low morbidity, while preventing future disease. Clearance of polyps by duodenotomy and local resection is relatively safe, but has a 100 per cent recurrence rate and is no longer advocated[40]. A proctocolectomy for FAP of the colon has a mortality rate of around 1 per cent. PD has a relatively low mortality rate but morbidity remains substantial even in experienced hands[29]. Pancreas-sparing duodenectomy is a relatively new procedure with limited data and follow-up. The procedure was first reported by Chung and colleagues[24] in 1995 as an alternative to PD on the basis of potentially lower morbidity and mortality rates. Depending on the extent of resection, the authors classified PPTD into three types. Type I comprises subtotal duodenectomy that preserves the major and minor papillae, and has been further subdivided into Ia (duodenal resection above the papilla) and Ib (resection below the papilla). In type II, the papilla is left as a button on the pancreatic head after total duodenectomy, with reconstruction using the technique of papillojejunostomy. In type III, the papilla is removed, and the terminal portions of the CBD and pancreatic duct are exposed and anastomosed to an isolated loop of jejunum. The latter procedure was performed in the present study.

PPTD might offer several advantages compared with conventional PD. The procedure involves removal of the

Table 2 Comparison of pancreas-preserving total duodenectomy with pancreatoduodenectomy for ampulla of Vater tumours

	PPTD ($n = 26$)	PD ($n = 77$)	*P*
Preoperative characteristics			
Male	16 (62)	58 (75)	0·117†
Age (years)*	51 (33–63)	62 (33–88)	< 0·001‡
Previous laparotomy	26 (100)	14 (18)	< 0·001†
Diabetes	0 (0)	8 (10)	0·089§
Intraoperative characteristics			
Pylorus preservation	15 (58)	73 (95)	< 0·001†
Duration of procedure (min)*	336 (225–661)	271 (180–542)	0·002¶
Postoperative complications			
Overall	16 (62)	44 (57)	0·694†
Surgery related	12 (46)	29 (38)	0·444†
Pancreatic leakage	7 (27)	19 (25)	0·820†
Delayed gastric emptying	2 (8)	17 (22)	0·145§
Bleeding	1 (4)	10 (13)	0·283§
Intra-abdominal abscess	2 (8)	12 (16)	0·509§
Wound infection	3 (12)	5 (6)	0·413§
Non-surgical	5 (19)	22 (29)	0·349†
Cardiac	1 (4)	6 (8)	0·676§
Pulmonary	3 (12)	14 (18)	0·551§
Urinary tract infection	1 (4)	6 (8)	0·676§
Relaparotomy	4 (15)	15 (19)	0·775§
Death	1 (4)	2 (3)	1·000§
Postoperative hospital stay (days)*	14 (6–225)	15 (9–140)	0·509¶
Pathology			
Invasive malignancy	2 (8)	77 (100)	< 0·001†
Tumour size (cm)*	2·8 (0·3–8)	2·0 (0–22)	< 0·001¶
Follow-up			
Postoperative diabetes mellitus	0 (0)	9 (12)	0·107§
Ulceration of jejunal limb	5 (19)	0 (0)	0·001§
Recurrent disease	4 (15)	23 (30)	0·146†

Values in parentheses are percentages unless indicated otherwise; *values are median (range). PPTD, pancreas-preserving total duodenectomy; PD, pancreatoduodenectomy. †χ^2 test; ‡Student's *t* test; §Fisher's exact test; ¶Mann–Whitney *U* test.

duodenum without loss of the adjacent pancreas, thereby avoiding unnecessary resection of the pancreas. It also avoids hepaticojejunostomy and a pancreaticojejunostomy to the cut surface of a soft pancreas with a non-dilated duct, each with its inherent problems. A friable and non-dilated pancreas is associated with a high rate of leakage from the anastomosis[41]. PPTD allows optimal postoperative endoscopic surveillance. This is important in patients with a good long-term prognosis and the need for lifelong frequent follow-up.

A high rate of jejunal ulceration was found in the present study, after which a decision was made to perform a Roux-en-Y reconstruction routinely, to reduce this problem. The advantage of optimal postoperative surveillance is unfortunately lost with this reconstruction method. One recent report stated that a drawback of PPTD is the possibility of residual disease in the ampulla of Vater[29]. This would be difficult to detect during surveillance and so the use of PD was advocated[29]. The problem of potential residual disease in the ampulla could be circumvented by performing a type III procedure, which ensures that the duodenal mucosa is removed completely from the ampulla[42].

Prophylactic resection should carry virtually no risk for the patient, but this was not the case in the present study as one patient died and others experienced substantial morbidity. This clearly shows problems with the new procedure. As regards PD, leakage from the pancreaticojejunostomy and subsequent major delayed haemorrhage is generally the cause of death[41]. Leakage has also been reported in patients with FAP, often attributed to the presence of pancreas divisum[43]. Failure to recognize this aberrant duct and subsequent ligation or clipping will have detrimental consequences and may even cause death, as in the present study.

Although comparison between FAP and ampulla of Vater disease has well known limitations and is subject to bias, ampullary disease mimics duodenal FAP very closely. Both diseases originate in the duodenum, and are frequently associated with a soft pancreas and a non-dilated

Table 3 Patients who underwent pancreas-preserving total duodenectomy for familial adenomatous polyposis in the literature, including the present study

Reference	Year	Country	*n*	Morbidity	Death	Follow-up (months)	Recurrence
Alarcon *et al.*[16]	1999	USA	3	0	0	40–50 (mean 45·7)	No recurrence. Two patients had tubular adenoma in duodenal bulb
Farnell *et al.*[17,18]	2000	USA	5	4	0	4–192 (mean 67)	No recurrence
Sarmiento *et al.*[19]	2002	USA	5	2	0	6–44 (mean 23)	Two patients had polyps in jejunum
Kalady *et al.*[20]	2002	USA	3	2	0	6–60 (mean 26)	One patient had polyp recurrence in jejunum at 5 years
Lundell *et al.*[21]	2002	Sweden	2	0	0	All patients had follow-up after 6 months	No recurrence
de Vos tot Nederveen Cappel *et al.*[22,23]	2003	International†	6	4	0	2–15 (mean 11)	No recurrence
Mackey *et al.*[24,25]	2005	USA	21	8	0	3–152 (mean 79)	Two patients developed polyps in jejunum
Present study	2007	The Netherlands	26	16	1	4–81 (mean 52)	Four patients developed polyps in jejunum
Total			71	36 (51)*	1 (1)*		

*Values in parentheses are percentages. †The Netherlands, Germany, Austria, Finland, Sweden, Denmark, South Africa and Canada.

Table 4 Patients who underwent pancreatoduodenectomy for familial adenomatous polyposis in the literature

Reference	Year	Country	*n*	Morbidity	Death	Follow-up	Recurrence
Balladur *et al.*[26]	1993	France	2	0	0	24–28 (mean 26)	No recurrence
Penna *et al.*[27]	1993	France	10	2	0	9–108 (mean 42)	No recurrence
Farnell *et al.*[17]	2000	USA	20	8	1	4–192 (mean 67)	No recurrence
Ruo *et al.*[28]	2002	USA	8	1	0	37–192 (median 71)	One patient developed polyps in jejunum
de Vos tot Nederveen Cappel *et al.*[22]	2003	International†	35	22	1	7–96 (mean 47)	Recurrence in 15 and nine died from metastatic disease
Gallagher *et al.*[29]	2004	UK	15	8	1	3–105 (mean 35)	No recurrence
Morpurgo *et al.*[30]	2004	USA	4	2	0	Unknown	No recurrence
Total			94	43 (46)*	3 (3)*		

*Values in parentheses are percentages. †The Netherlands, Germany, Austria, Finland, Sweden, Denmark, South Africa and Canada.

duct. In this study, a major difference between patients who had PPTD for FAP and those who had PD for ampullary tumours was that the former had all undergone abdominal surgery previously. Consequently adhesions had to be taken down during the PPTD, prolonging the operating time, with a risk of bowel complications. Furthermore, nearly a quarter of these patients had an ileostomy, which also carries risks, such as wound infection. The present findings were similar to the results in the literature. Because similar techniques were used in these studies, published data and those from the present study were pooled; no obvious advantage for either procedure was found.

PPTD in patients with FAP is a major procedure. There are no clear practical advantages for this new procedure and, in contrast to some reports, PPTD is not associated with lower morbidity or mortality rates than conventional PD.

References

1 Fazio VW, Ziv Y, Church JM, Oakley JR, Lavery IC, Milsom JW *et al.* Ileal pouch–anal anastomoses complications and function in 1005 patients. *Ann Surg* 1995; **222**: 120–127.

2 Spigelman AD, Talbot IC, Penna C, Nugent KP, Phillips RK, Costello C *et al.* Evidence for adenoma–carcinoma sequence in the duodenum of patients with familial adenomatous polyposis. The Leeds Castle Polyposis Group (Upper Gastrointestinal Committee). *J Clin Pathol* 1994; **47**: 709–710.

3 Domizio P, Talbot IC, Spigelman AD, Williams CB, Phillips RK. Upper gastrointestinal pathology in familial adenomatous polyposis: results from a prospective study of 102 patients. *J Clin Pathol* 1990; **43**: 738–743.

4 Spigelman AD, Williams CB, Talbot IC, Domizio P, Phillips RK. Upper gastrointestinal cancer in patients with familial adenomatous polyposis. *Lancet* 1989; **2**: 783–785.

5 Jagelman DG, DeCosse JJ, Bussey HJ. Upper gastrointestinal cancer in familial adenomatous polyposis. *Lancet* 1988; **1**: 1149–1151.

6 Offerhaus GJ, Giardiello FM, Krush AJ, Booker SV, Tersmette AC, Kelley NC *et al*. The risk of upper gastrointestinal cancer in familial adenomatous polyposis. *Gastroenterology* 1992; **102**: 1980–1982.

7 Arvanitis ML, Jagelman DG, Fazio VW, Lavery IC, McGannon E. Mortality in patients with familial adenomatous polyposis. *Dis Colon Rectum* 1990; **33**: 639–642.

8 Belchetz LA, Berk T, Bapat BV, Cohen Z, Gallinger S. Changing causes of mortality in patients with familial adenomatous polyposis. *Dis Colon Rectum* 1996; **39**: 384–387.

9 van Heek NT, Kuhlmann KF, Scholten RJ, de Castro SM, Busch OR, van Gulik TM *et al*. Hospital volume and mortality after pancreatic resection: a systematic review and an evaluation of intervention in the Netherlands. *Ann Surg* 2005; **242**: 781–788.

10 Konsten JL, Kazemier G, Dees J, van Eijck CH. [Pancreas-preserving total duodenectomy due to familial adenomatous polyposis in 4 patients.] *Ned Tijdschr Geneeskd* 2002; **146**: 621–624.

11 de Castro SM, van Heek NT, Kuhlmann KF, Busch OR, Offerhaus GJ, van Gulik TM *et al*. Surgical management of neoplasms of the ampulla of Vater: local resection or pancreatoduodenectomy and prognostic factors for survival. *Surgery* 2004; **136**: 994–1002.

12 Pedrazzoli S, Beger HG, Obertop H, Andren-Sandberg A, Fernandez-Cruz L, Henne-Bruns D *et al*. A surgical and pathological based classification of resective treatment of pancreatic cancer. Summary of an international workshop on surgical procedures in pancreatic cancer. *Dig Surg* 1999; **16**: 337–345.

13 Reid-Lombardo KM, Farnell MB, Crippa S, Barnett M, Maupin G, Bassi C *et al*. Pancreatic anastomotic leakage after pancreaticoduodenectomy in 1507 patients: a report from the Pancreatic Anastomotic Leak Study Group. *J Gastrointest Surg* 2007; **11**: 1451–1458.

14 Wente MN, Veit JA, Bassi C, Dervenis C, Fingerhut A, Gouma DJ *et al*. Postpancreatectomy hemorrhage (PPH): an International Study Group of Pancreatic Surgery (ISGPS) definition. *Surgery* 2007; **142**: 20–25.

15 Wente MN, Bassi C, Dervenis C, Fingerhut A, Gouma DJ, Izbicki JR *et al*. Delayed gastric emptying (DGE) after pancreatic surgery: a suggested definition by the International Study Group of Pancreatic Surgery (ISGPS). *Surgery* 2007; **142**: 761–768.

16 Alarcon FJ, Burke CA, Church JM, Van Stolk RU. Familial adenomatous polyposis: efficacy of endoscopic and surgical treatment for advanced duodenal adenomas. *Dis Colon Rectum* 1999; **42**: 1533–1536.

17 Farnell MB, Sakorafas GH, Sarr MG, Rowland CM, Tsiotos GG, Farley DR *et al*. Villous tumors of the duodenum: reappraisal of local *vs*. extended resection. *J Gastrointest Surg* 2000; **4**: 13–21.

18 Tsiotos GG, Sarr MG. Pancreas-preserving total duodenectomy. *Dig Surg* 1998; **15**: 398–403.

19 Sarmiento JM, Thompson GB, Nagorney DM, Donohue JH, Farnell MB. Pancreas-sparing duodenectomy for duodenal polyposis. *Arch Surg* 2002; **137**: 557–562.

20 Kalady MF, Clary BM, Tyler DS, Pappas TN. Pancreas-preserving duodenectomy in the management of duodenal familial adenomatous polyposis. *J Gastrointest Surg* 2002; **6**: 82–87.

21 Lundell L, Hyltander A, Liedman B. Pancreas-sparing duodenectomy: technique and indications. *Eur J Surg* 2002; **168**: 74–77.

22 de Vos tot Nederveen Cappel WH, Jarvinen HJ, Bjork J, Berk T, Griffioen G, Vasen HF. Worldwide survey among polyposis registries of surgical management of severe duodenal adenomatosis in familial adenomatous polyposis. *Br J Surg* 2003; **90**: 705–710.

23 Soravia C, Berk T, Haber G, Cohen Z, Gallinger S. Management of advanced duodenal polyposis in familial adenomatous polyposis. *J Gastrointest Surg* 1997; **1**: 474–478.

24 Chung RS, Church JM, vanStolk R. Pancreas-sparing duodenectomy: indications, surgical technique, and results. *Surgery* 1995; **117**: 254–259.

25 Mackey R, Walsh RM, Chung R, Brown N, Smith A, Church J *et al*. Pancreas-sparing duodenectomy is effective management for familial adenomatous polyposis. *J Gastrointest Surg* 2005; **9**: 1088–1093.

26 Balladur P, Penna C, Tiret E, Vaillant JC, Gailleton R, Parc R. Pancreatico-duodenectomy for cancer and precancer in familial adenomatous polyposis. *Int J Colorectal Dis* 1993; **8**: 151–153.

27 Penna C, Phillips RK, Tiret E, Spigelman AD. Surgical polypectomy of duodenal adenomas in familial adenomatous polyposis: experience of two European centres. *Br J Surg* 1993; **80**: 1027–1029.

28 Ruo L, Coit DG, Brennan MF, Guillem JG. Long-term follow-up of patients with familial adenomatous polyposis undergoing pancreaticoduodenal surgery. *J Gastrointest Surg* 2002; **6**: 671–675.

29 Gallagher MC, Shankar A, Groves CJ, Russell RC, Phillips RK. Pylorus-preserving pancreaticoduodenectomy for advanced duodenal disease in familial adenomatous polyposis. *Br J Surg* 2004; **91**: 1157–1164.

30 Morpurgo E, Vitale GC, Galandiuk S, Kimberling J, Ziegler C, Polk HC Jr. Clinical characteristics of familial adenomatous polyposis and management of duodenal adenomas. *J Gastrointest Surg* 2004; **8**: 559–564.

31 Galle TS, Juel K, Bulow S. Causes of death in familial adenomatous polyposis. *Scand J Gastroenterol* 1999; **34**: 808–812.

32 Vasen HF, Bulow S, Myrhoj T, Mathus-Vliegen L, Griffioen G, Buskens E *et al*. Decision analysis in the management of duodenal adenomatosis in familial adenomatous polyposis. *Gut* 1997; **40**: 716–719.
33 Beckwith PS, van Heerden JA, Dozois RR. Prognosis of symptomatic duodenal adenomas in familial adenomatous polyposis. *Arch Surg* 1991; **126**: 825–827.
34 Bulow S, Bjork J, Christensen IJ, Fausa O, Jarvinen H, Moesgaard F *et al*. Duodenal adenomatosis in familial adenomatous polyposis. *Gut* 2004; **53**: 381–386.
35 Groves CJ, Saunders BP, Spigelman AD, Phillips RK. Duodenal cancer in patients with familial adenomatous polyposis (FAP): results of a 10 year prospective study. *Gut* 2002; **50**: 636–641.
36 Burke CA, Beck GJ, Church JM, van Stolk RU. The natural history of untreated duodenal and ampullary adenomas in patients with familial adenomatous polyposis followed in an endoscopic surveillance program. *Gastrointest Endosc* 1999; **49**: 358–364.
37 Cahen DL, Fockens P, de Wit LT, Offerhaus GJ, Obertop H, Gouma DJ. Local resection or pancreaticoduodenectomy for villous adenoma of the ampulla of Vater diagnosed before operation. *Br J Surg* 1997; **84**: 948–951.
38 Stolte M, Pscherer C. Adenoma–carcinoma sequence in the papilla of Vater. *Scand J Gastroenterol* 1996; **31**: 376–382.
39 Kadmon M, Tandara A, Herfarth C. Duodenal adenomatosis in familial adenomatous polyposis coli. A review of the literature and results from the Heidelberg Polyposis Register. *Int J Colorectal Dis* 2001; **16**: 63–75.
40 Brosens LA, Keller JJ, Offerhaus GJ, Goggins M, Giardiello FM. Prevention and management of duodenal polyps in familial adenomatous polyposis. *Gut* 2005; **54**: 1034–1043.
41 de Castro SM, Busch OR, van Gulik TM, Obertop H, Gouma DJ. Incidence and management of pancreatic leakage after pancreatoduodenectomy. *Br J Surg* 2005; **92**: 1117–1123.
42 Spigelman A, Gani J. Pylorus-preserving pancreaticoduodenectomy for advanced duodenal disease in familial adenomatous polyposis. *Br J Surg* 2005; **92**: 253.
43 Koninger J, Friess H, Wagner M, Kadmon M, Buchler MW. [Technique of pancreas-preserving duodenectomy.] *Chirurg* 2005; **76**: 273–281.