Case Presentation

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4/22/2005

Management of Duodenal Tumors

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Duodenal Tumors

- The duodenum is the shortest segment of the small intestine but may be affected by a wide variety of primary and secondary neoplasms
- Benign :
  adenoma, leiomyoma, lipoma
- Malignant :
  adenocarcinoma, carcinoid, lymphoma, leiomyosarcoma

Duodenal Tumors

- Primary tumors of the duodenum are uncommon.
- Peak incidence : 6th and 8th decade of life

The Etiology of DT is unknown

Protective factors ?

- Secretory immunoglobulins
- Small intestinal hydroxylases (could inactivate potential carcinogens)
- Alkalinity in the duodenum (could prevent formation of potential carcinogens)
- Rapid transit of liquid bowel contents
- Lack of bacteria

Symptoms are related to the tumor location

- N ,V
- Pain
- Jaundice
- Anemia
- Pancreatitis
- Hematemesis
- Melena
- Palpable mass
- Obstruction
- Weight loss
- Intussusception
- Cholangitis

Benign Duodenal Tumors

Adenomas

- The most common benign tumors in the duodenum
- Present either as :
  Adenomatous polyps
  Brunner's gland adenomas
  Villous adenomas (high rate of malignant transformation)

Adenomatous polyps

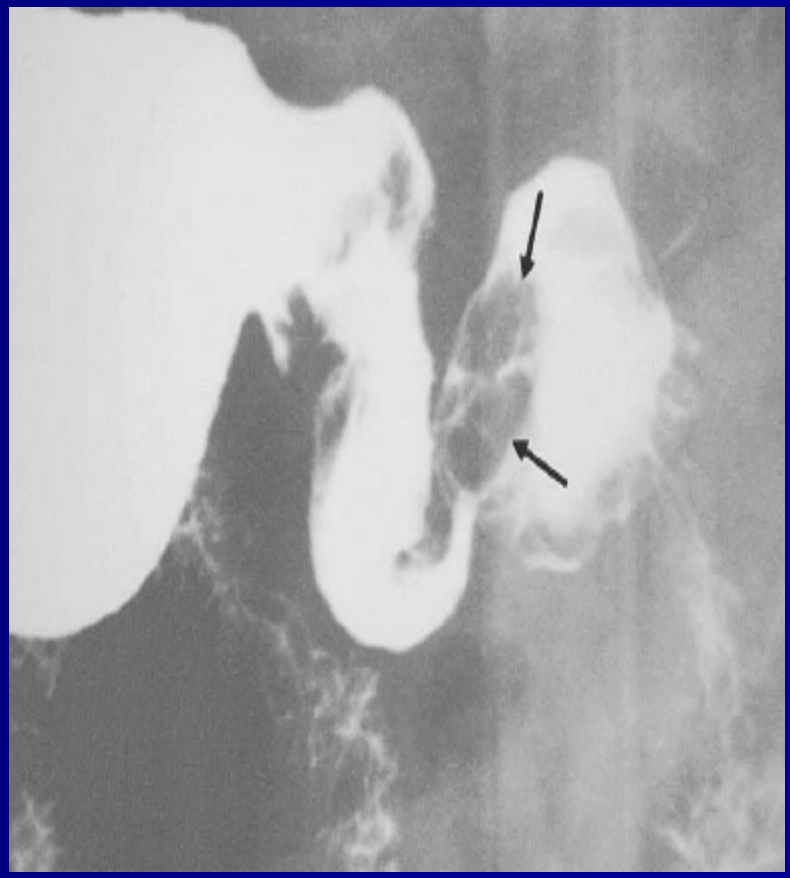
- Sessile, nodular or pedunculated.
- Most are asymptomatic
- Periampullary lesions may cause intermittent jaundice or pancreatitis.
- Anemia may also occur secondary to chronic blood loss.
- Duodenal adenomatous polyps are common in FAP and Gardner's syndrome

Adenomatous polyps

- Adenomatous polyps are most often at the gastroduodenal junction
- For the small polyps (< 2 cm), standard abdominal CT may not be able to demonstrate the lesions clearly

Adenomatous polyps

Sessile or pedunculated intraluminal filling defects are usually depicted on barium studies



Sporadic duodenal adenoma (DA)

- 7% of duodenal polyps (6.9% of 378 duodenal polyps found at 25 000 EGD were adenomatous)
- Predominance at the ampulla and periampullary region

Sporadic DA is associated with colorectal neoplasia

Table 2 Proportion of duodenal adenoma patients and endoscoped controls with identified colorectal neoplasia

Colorectal neoplasia found*	Duodenal adenoma (n = 34)	Controls (n = 102)	p Value
All colorectal neoplasia	19 (56%)	34 (33%)	0.03
Cancer or advanced adenoma	13 (38%)	19 (19%)	0.05 (NS, see text)
Cancer	7 (21%)	8 (8%)	0.05 (NS, see text)
Advanced adenoma	6 (18%)	11 (11%)	0.44 NS
Non-advanced adenoma	6 (18%)	15 (26%)	0.51 NS

Sporadic duodenal adenoma (DA)

- Colonoscopy is indicated for all DA pts.

Approach to benign duodenal polyps

- Any pedunculated lesion is removed by using a snare.
- If tumors < 1 cm → snare polypectomy
- Small single sessile polyps or multiple small polyps → argon plasma coagulator.
- Endoscopic US is helpful in any sessile lesion where invasion is suspected.
- Large flat polyps are removed by piecemeal polypectomy

Laparoscopic resection of a periampullary villous adenoma

- Criteria for endoscopic snare papillectomy of papillary adenomas:

  (1) < 4 cm
  (2) (regular margin, no ulceration, soft consistency)
  (3) benign histologic findings
  (4) absence of intraductal involvement as demonstrated by ERCP/endoscopic ultrasound .

- Recurrence rates of 19%- 26%, within 1 year of polypectomy

Approach to benign duodenal polyps

- Criteria for surgery :

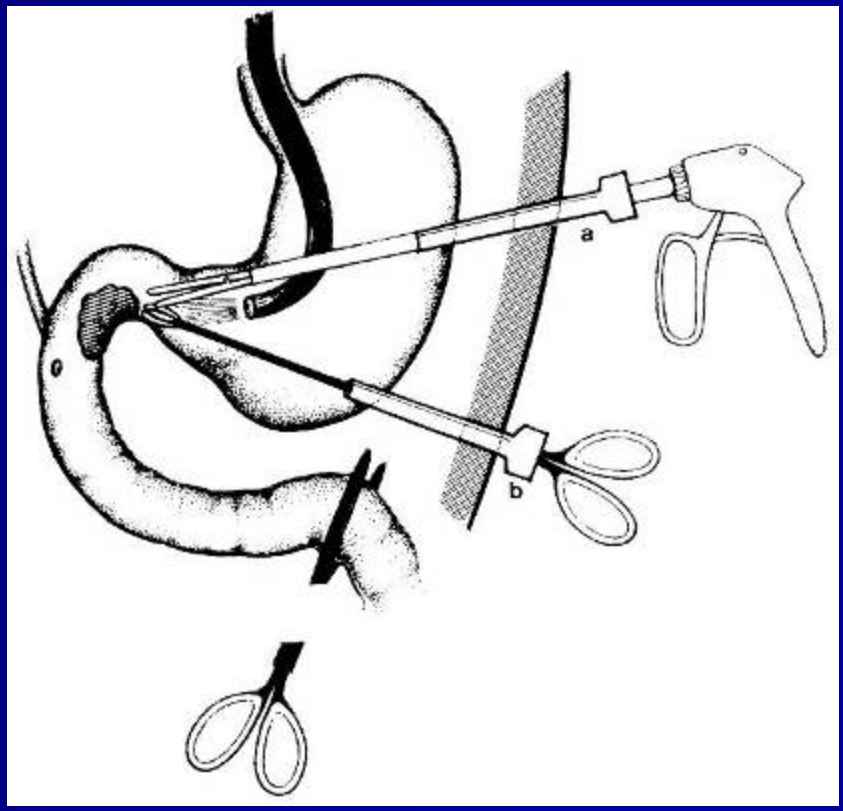
  (1) large polyp (1-3 cm in size)
  (2) a polyp in which EUS shows deeper tumor infiltration
  (4) polyp with severe dysplasia or carcinomatous infiltration
  (5) recurrence of the polyp after complete endoscopic removal.

Approach to benign duodenal polyps

- The surgical procedure of choice should be segmental duodenal resection or transduodenal polypectomy when feasible
- Lesions located in D1 are well suited for transduodenal polypectomy because the duodenum can be closed with pyloroplasty avoiding luminal narrowing
- Segmental resection should be undertaken if simple closure would induce luminal narrowing (D3,D4)

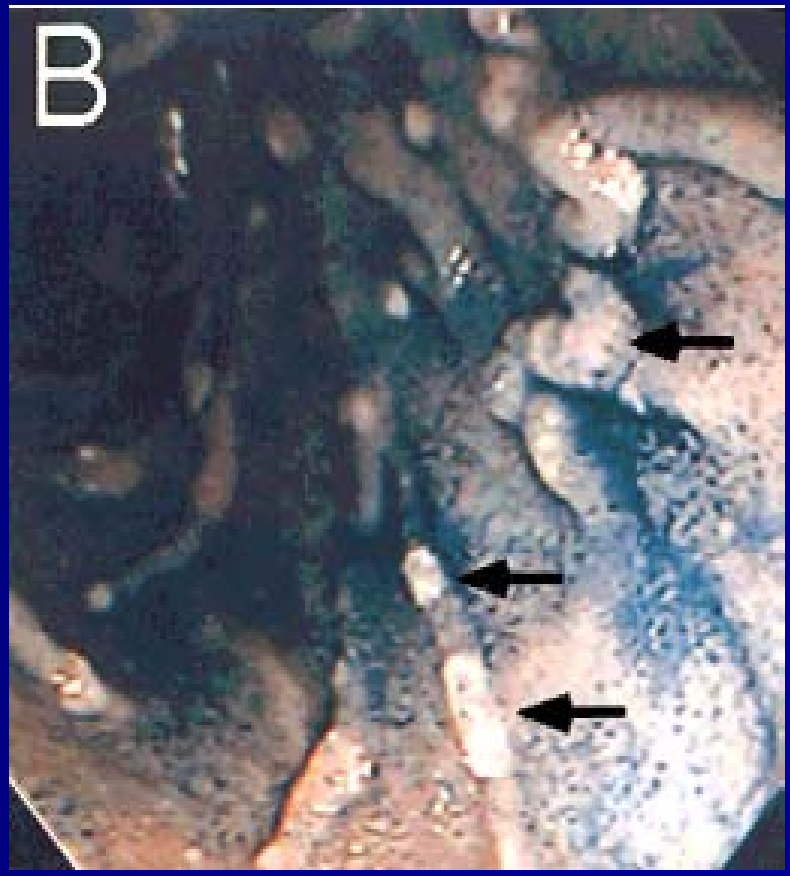
Transgastric Endoluminal Laparoscopic Resection of a Duodenal Polyp

- Laparoscopic procedure used to treat gastric lesions such as polyps and carcinoma in situ.
- This approach can also be used to treat proximal duodenal problems such as bleeding and polyps.



Duodenal adenomatosis in FAP

Up to 95% of pts with FAP develop duodenal polyps



Duodenal adenomatosis in FAP

- Adenoma–carcinoma sequence (like colonic polyps)
- Lifetime risk of 3–5% of duodenal cancer
- Prophylactic colectomy ↓ the incidence of colorectal cancer, duodenal cancer (330 x the population risk) and desmoid disease are now the leading causes of death in FAP.

Duodenal adenomatosis in FAP

- The incidence and severity of adenomatosis increase with age.

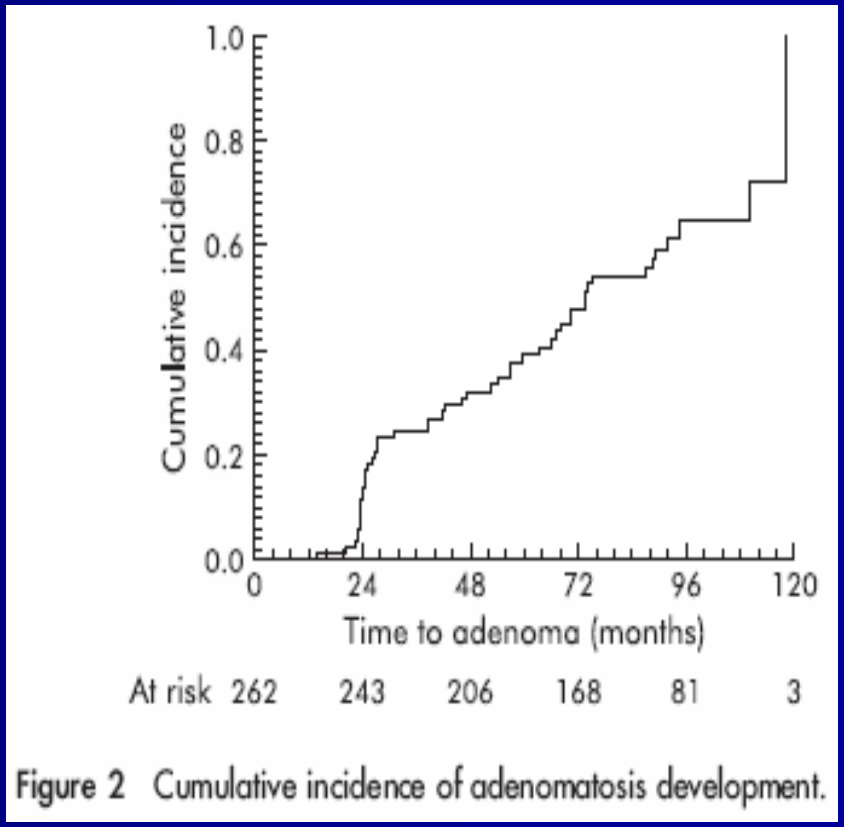


Figure 2 Cumulative incidence of adenomatosis development.

Duodenal adenomatosis in FAP

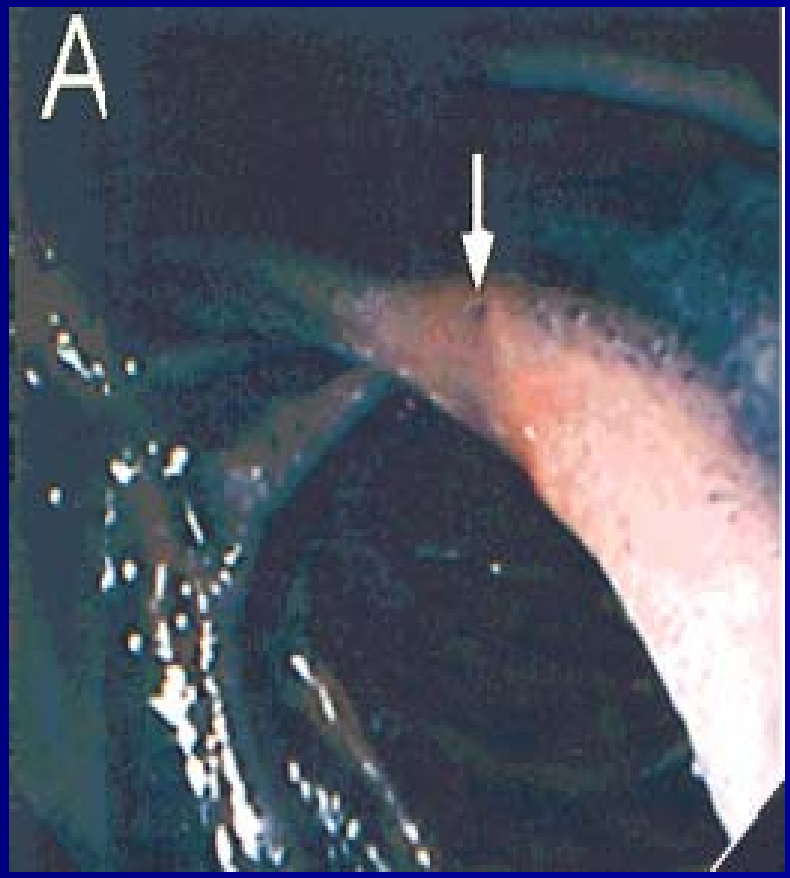
Table 3 Patient series, including results of upper gastrointestinal endoscopy follow up

					Findings at first endoscopy							
Author (ref No)	Year	No of patients	Endoscopy type*	Mean No of endoscopies	Age at diagnosis of FAP	Age at endoscopy	Duodenal adenomas (%)	Spigelman stage IV (%)	Cumul. incidence (%) of duodenal adenomas at 70–75 y	Cumul. incidence (%) of Spigelman stage IV at 70–75 y	Duodenal cancer (%)	Cumul. incidence (%) of duodenal cancer at 75 y
Church[7]	1992	247	SV+FV	3	25	33.5	65					
Nugent[8]	1994	70	SV	2							1.4	
Debinski[9]	1995	200				39	65				2.0	
Heiskanen[12]	1999	98	SV/FV	3			54	2	97†	30†		4†
Björk[14]	2001	180	FV	2			74	8	98	20		10
Kadmon[16]	2001	231	FV				58				1.7	
Groves[17]	2002	114	SV			42					5.0	
Present study		368	FV	4	25	37	65	7	99	52	1.6	4.5‡

*FV, forward viewing endoscope; SV, side viewing endoscope.
†At age 80 years.
‡At age 57 years.

Duodenal adenomatosis in FAP

- Even flat duodenal mucosa in patients with FAP had high proliferative activities



Duodenal adenomatosis in FAP

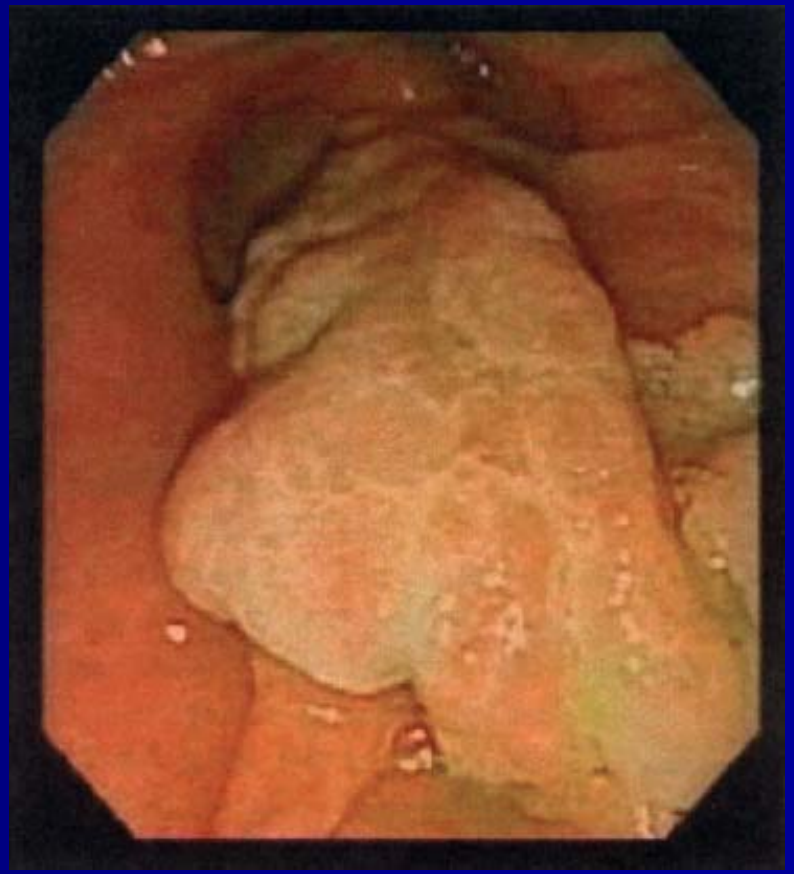
Table 1 Spigelman classification for duodenal familial adenomatous polyposis

	Score		
	1	2	3
No. of lesions	1–4	5–20	> 20
Maximum size (mm)	1–4	5–10	> 10
Histology	Tubular	Tubulovillous	Villous
Dysplasia	Mild	Moderate	Severe

Spigelman stage: 0, score 0; I, score 1–4; II, score 5–6; III, score 7–8; IV, score 9–12.

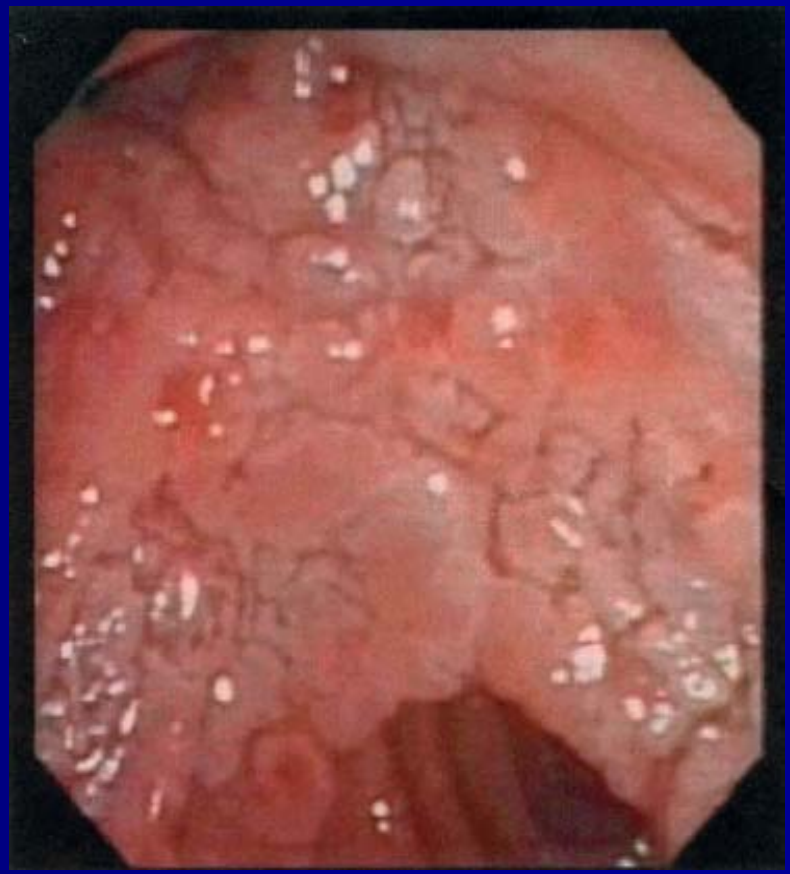
Stage IV duodenal adenomatosis with bulky ampullary neoplasm

- Endoscopic US is recommended for evaluation of pts with Spigelman stage IV



Stage IV duodenal adenomatosis (carpeting disease)

- Pts with Spigelman stage IV should be offered prophylactic surgery
- Risk of malignancy was 36 % > 10 y



Endoscopic Surveillance

- The first endoscopy should be carried out at the age of 30 y and include multiple random biopsies taken from the duodenal mucosa in pts w/o visible polyps.

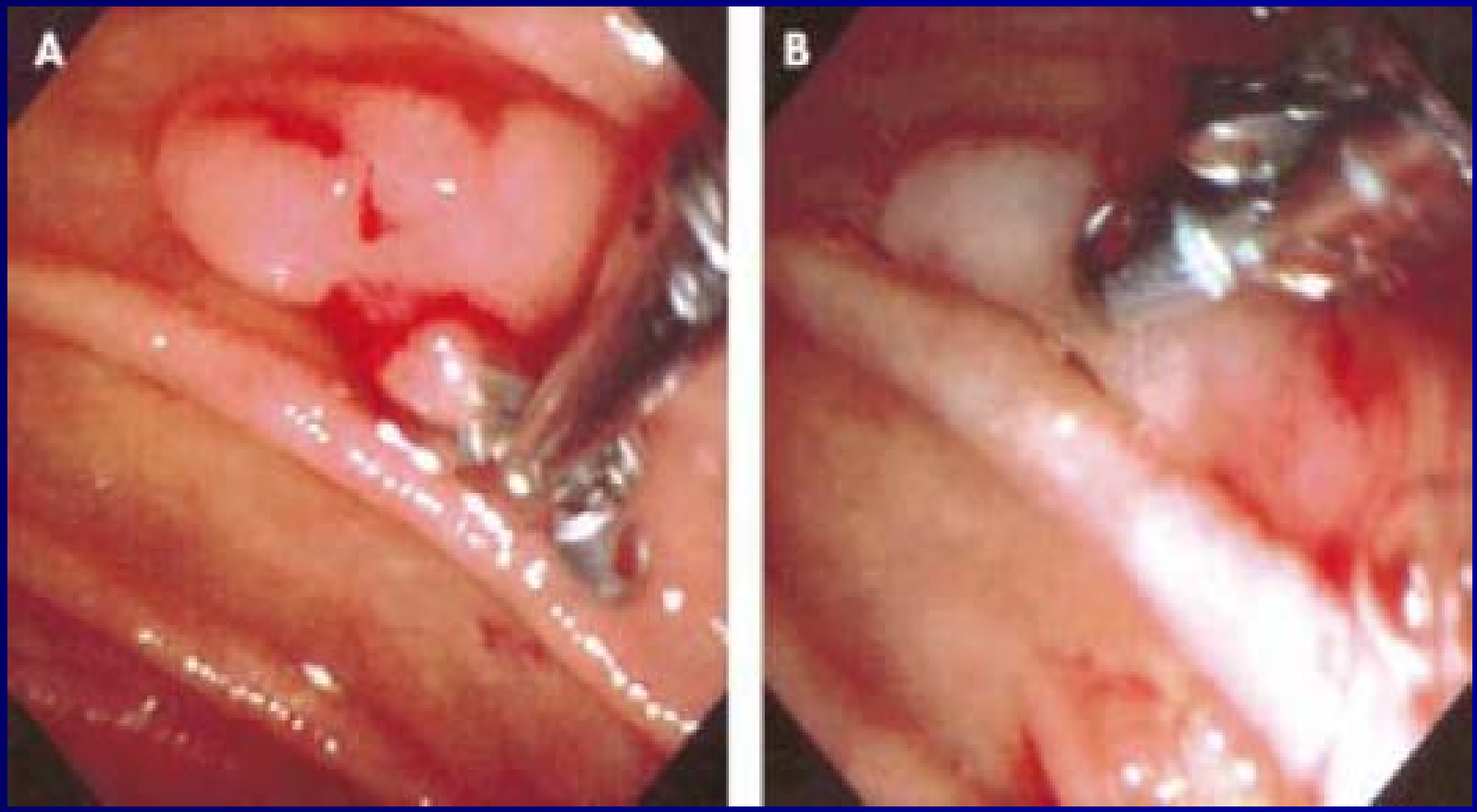
Endoscopic Surveillance

Table 2 Proposed programme for surveillance and treatment of duodenal adenomatosis

Spigelman stage 0	Endoscopy* at intervals of 5 y
Spigelman stage I	Endoscopy† at intervals of 5 y
Spigelman stage II	Endoscopy† at intervals of 3 y
Spigelman stage III	Endoscopy† at intervals of 1–2 y
Spigelman stage IV	Endoscopic ultrasonography Consider pancreas sparing or pylorus sparing duodenectomy

*Including multiple random biopsies from mucosal folds in patients without visible polyps.
†Including multiple biopsies from polyps.

Cyclooxygenase 2 inhibitor celecoxib significantly improves the endoscopic duodenal stage.



Duodenal adenomatosis in FAP

- Snare polypectomy in the FAP duodenum can be difficult because of the flat morphology of most duodenal polyps.

- Thermal ablation techniques
- Photodynamic therapy
- The argon plasma coagulator

Duodenal adenomatosis in FAP

- Surgical approaches include :

- Pancreaticoduodenectomy
- Duodenotomy and surgical polypectomy
- Local excision of the ampulla
- Pancreas-sparing duodenectomy

Duodenal adenomatosis in FAP

- Initial operations to remove duodenal polyps locally were followed by almost universal polyp recurrence within 1 year.
- In the 1990s, with reports of lower mortality rates following pancreatic surgery prophylactic resectional duodenal surgery became a reasonable option.

Duodenal adenomatosis in FAP

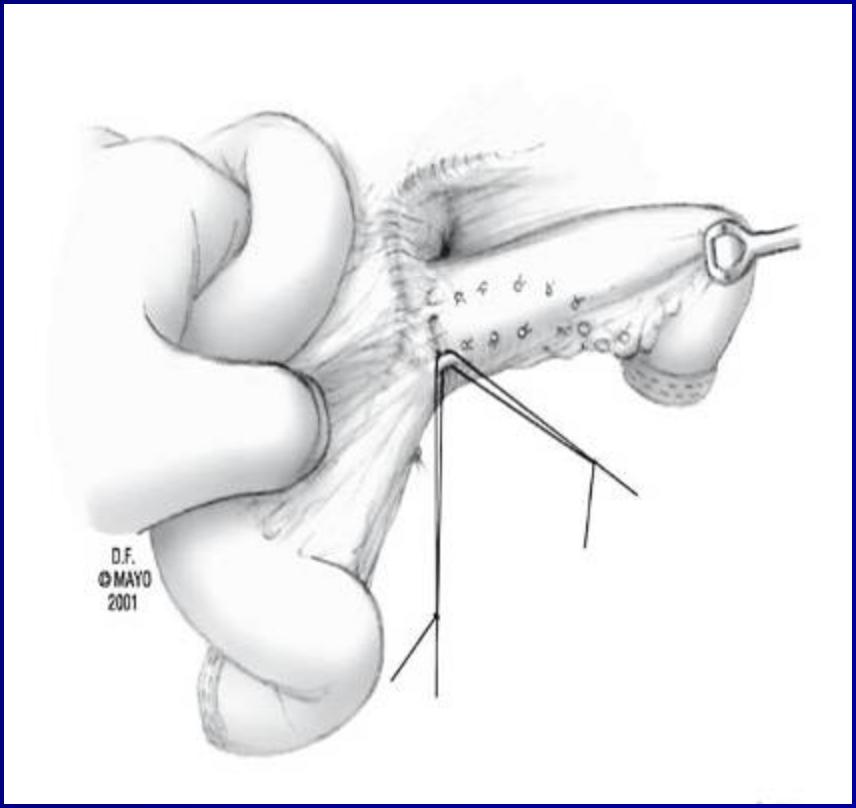
- Duodenotomy and clearance of adenomas in FAP was associated with recurrence in all patients after 6–36 months and progression to stage IV disease after a mean of 53 months.

Duodenal adenomatosis in FAP

- The operation of choice for premalignant duodenal polyposis is a pancreas preserving duodenectomy which leads to good functional outcome and facilitates endoscopic follow up.

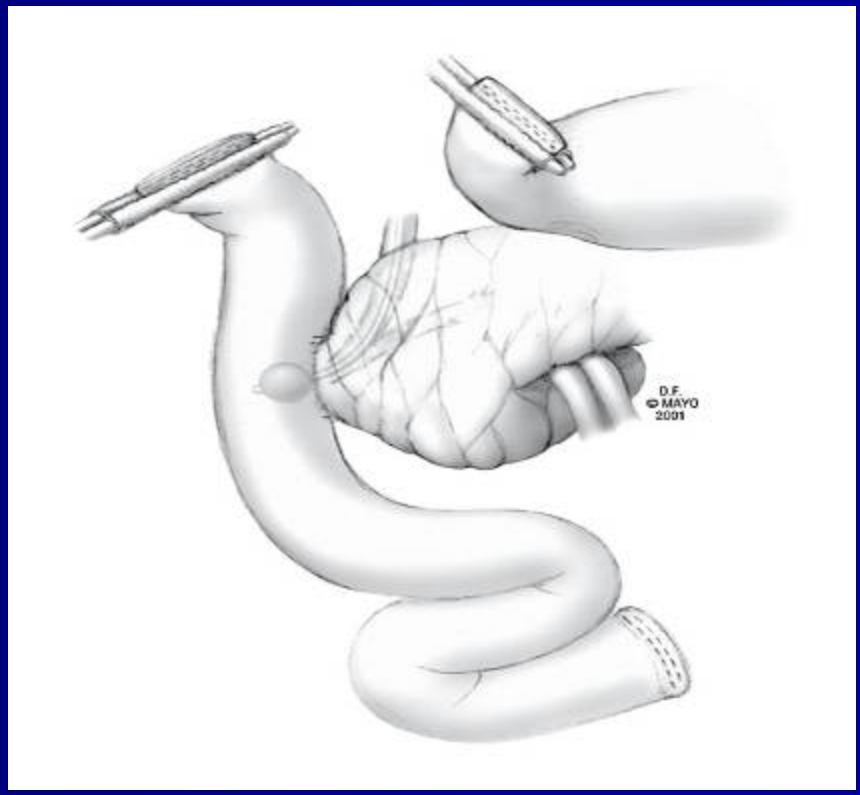
Pancreas-Sparing Duodenectomy for Duodenal Polyposis

- Dissection of the proximal jejunum and distal duodenum beginning 10 cm from the ligament of Treitz.



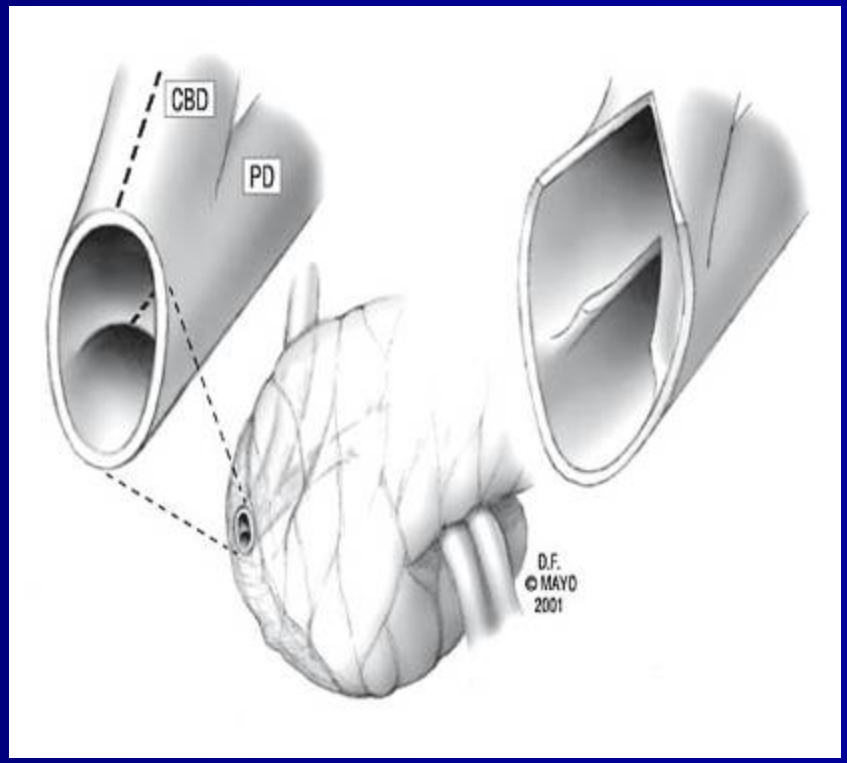
Pancreas-Sparing Duodenectomy for Duodenal Polyposis

- Completion of the duodenectomy. The duodenum is mobilized from above and below the ampulla of Vater, after proximal and distal transection. Note the presence of a Fogarty catheter in the common bile duct with the balloon inflated in the duodenum. The catheter greatly facilitates the identification of the papilla.



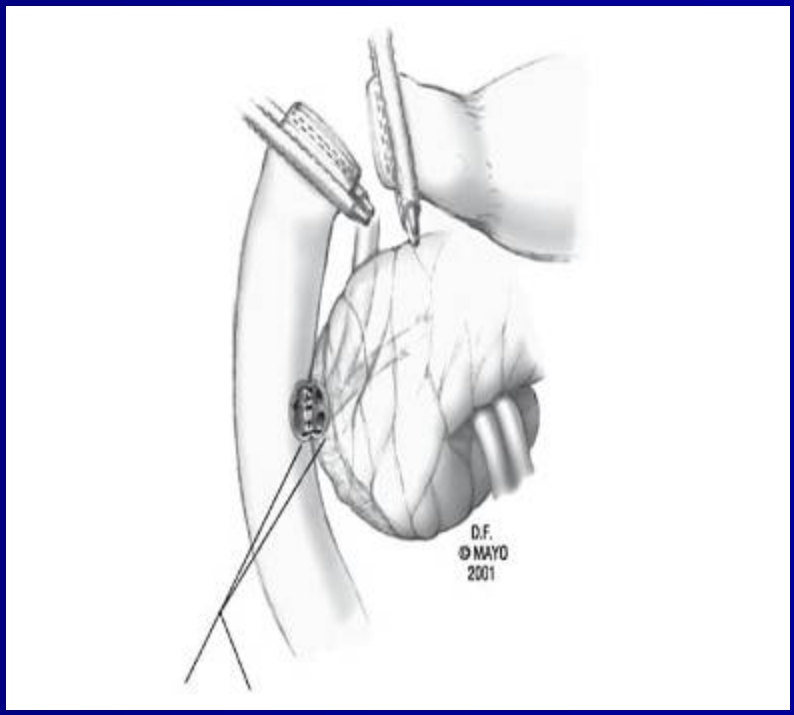
Pancreas-Sparing Duodenectomy for Duodenal Polyposis

- Sphincteroplasty and septoplasty. These 2 procedures increase the size of the ampullary complex, facilitating the posterior anastomosis.



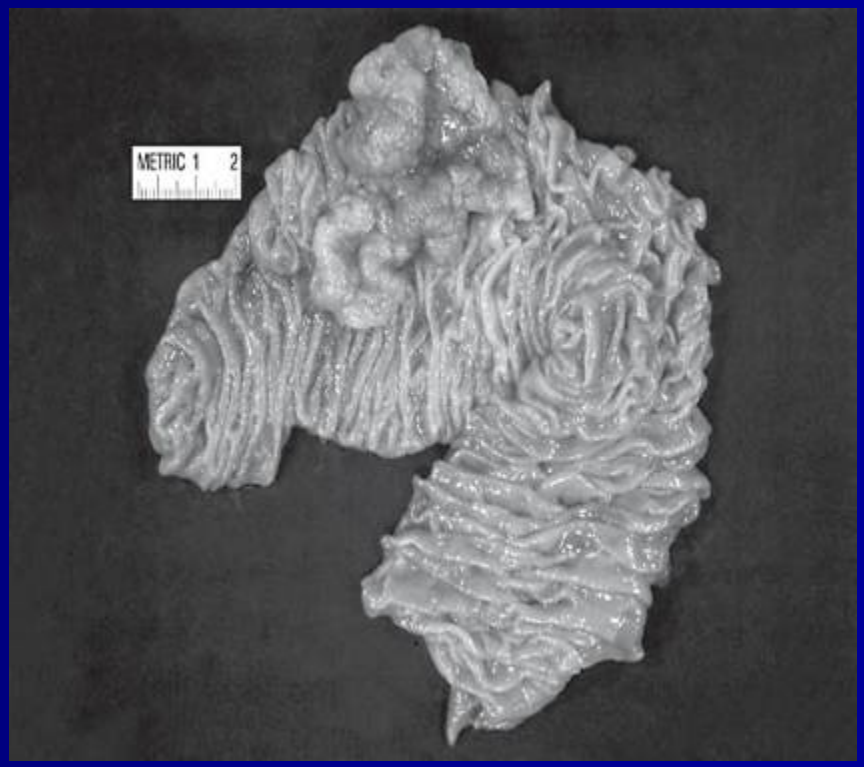
Pancreas-Sparing Duodenectomy for Duodenal Polyposis

- Ampullary-jejunal anastomosis constructed using the neoduodenum. With the jejunal mesentery positioned posteriorly, the anastomosis is fashioned with interrupted 5-0 absorbable suture. The final steps involve ligation of the accessory duct located anterior and superior to the major papilla and construction of the duodenojejunostomy. For the latter, the duodenal cuff is kept short. A duodenal mucosectomy is performed and the pyloric mucosa is incorporated with the duodenal seromuscular layer in preparation for end-to-end anastomosis.



Pancreas-Sparing Duodenectomy for Duodenal Polyposis

- Duodenal specimen in a patient with tubulovillous adenoma. The adenoma occupies more than half of the circumference of the duodenum, a finding that precludes transduodenal excision. Ruler indicates 2 cm.

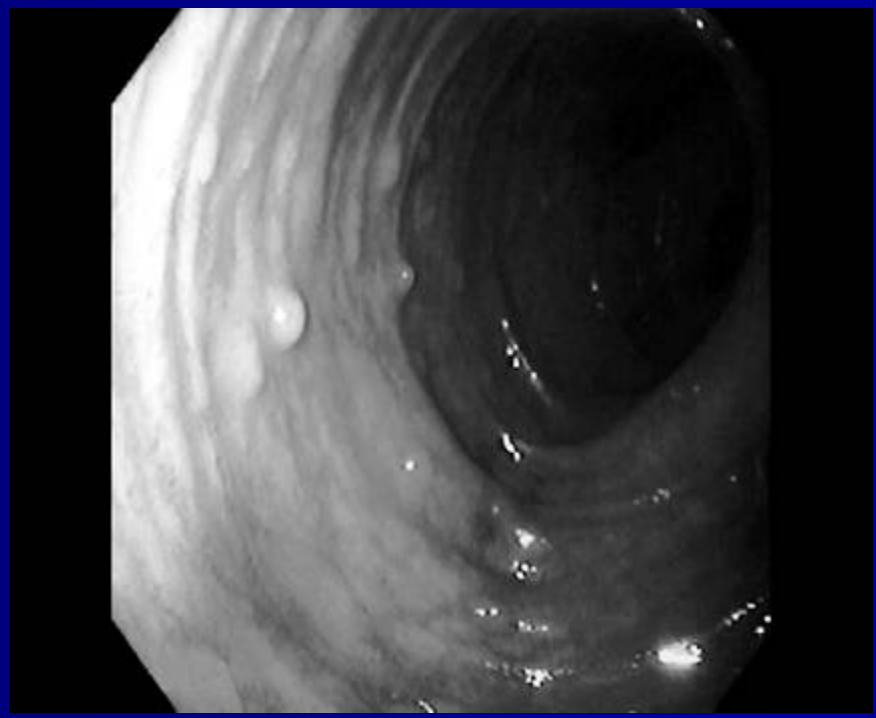


Pancreas-Sparing Duodenectomy for Duodenal Polyposis

- Although technically demanding, eliminates the need for pancreatic resection.
- PSD is associated with good absorptive capacity, weight gain, and quality of life.
- Long-term surveillance, however, is still required.
- Pancreas-sparing duodenectomy is contraindicated in the setting of malignancy.

Pancreas-Sparing Duodenectomy for Duodenal Polyposis

- Endoscopic picture of a neoduodenum (jejunum). Note the presence of small polyps in the wall. A biopsy specimen of the larger polyp was diagnosed as a tubular adenoma with low-grade dysplasia.

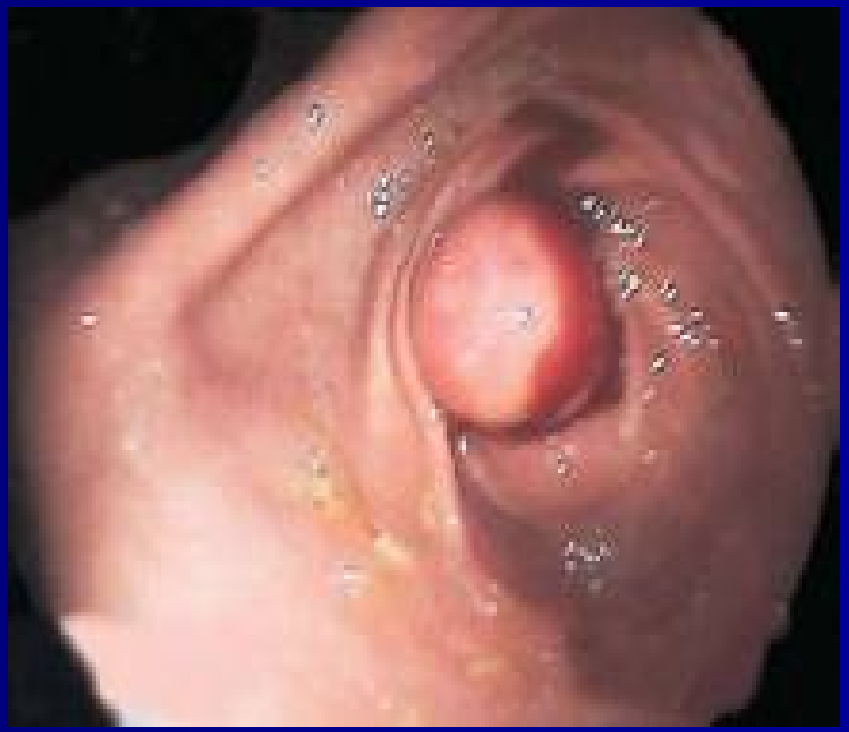


Brunner's gland adenomas

- May present as pedunculated polyps, circumscribed nodular hyperplasia, or diffuse nodular hyperplasia.
- The most common location is the posterior wall of the duodenum near the junction D1-D2 .
- The malignant potential is extremely low.
- Most patients remain asymptomatic
- Endoscopic or local open resection are curative.

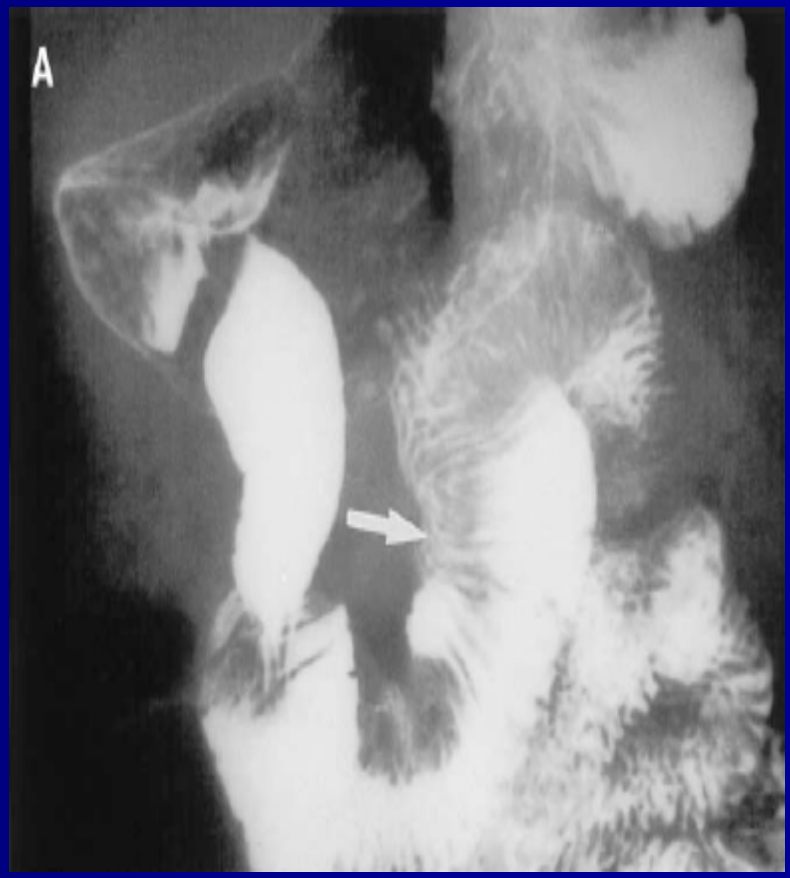
Lipomas

- Lipomas arise from the submucosa, but can be a subserosal lesion.
- Duodenum is the third common location of lipomas, following colon and ileum



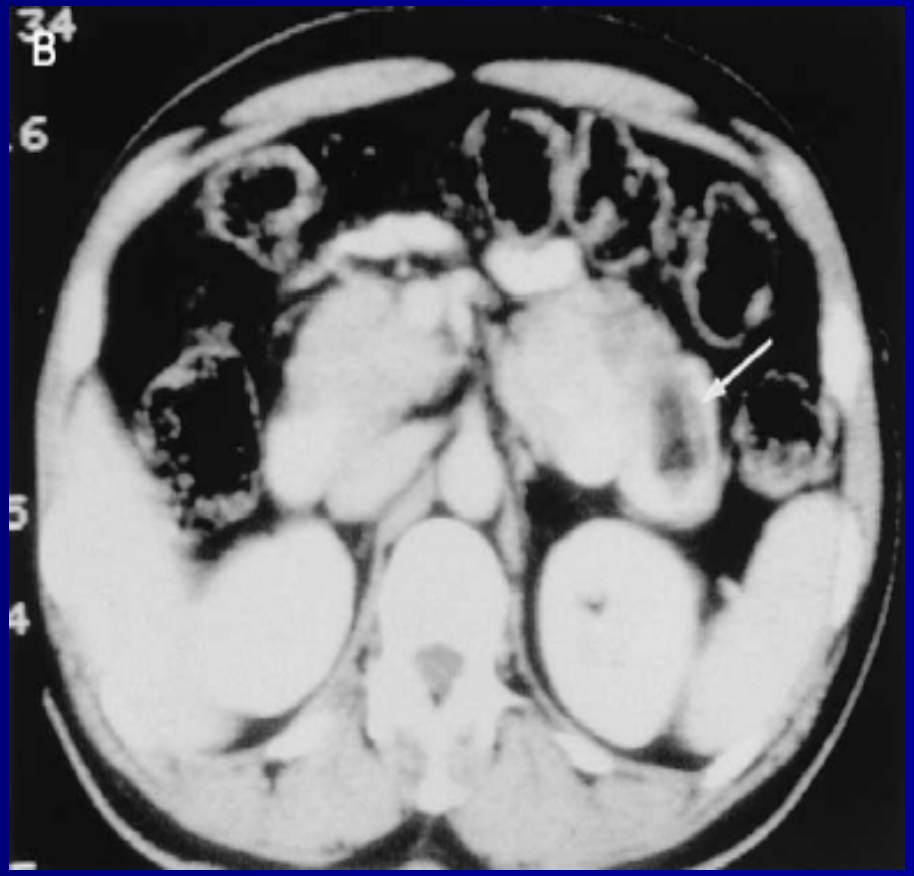
Lipomas

- (< 1 cm) are usually asymptomatic
- (>4 cm) may result in variable degree of intestinal obstruction, hemorrhage or intussusception



Lipomas

- Enucleation or local excision is sufficient treatment for symptomatic lesions.



Haemangiomas and lymphangiomas

- Well circumscribed submucosal masses, composed of blood vessels or lymphatic vessels.
- Bleeding from haemangiomas may be massive enough to require emergency laparotomy.

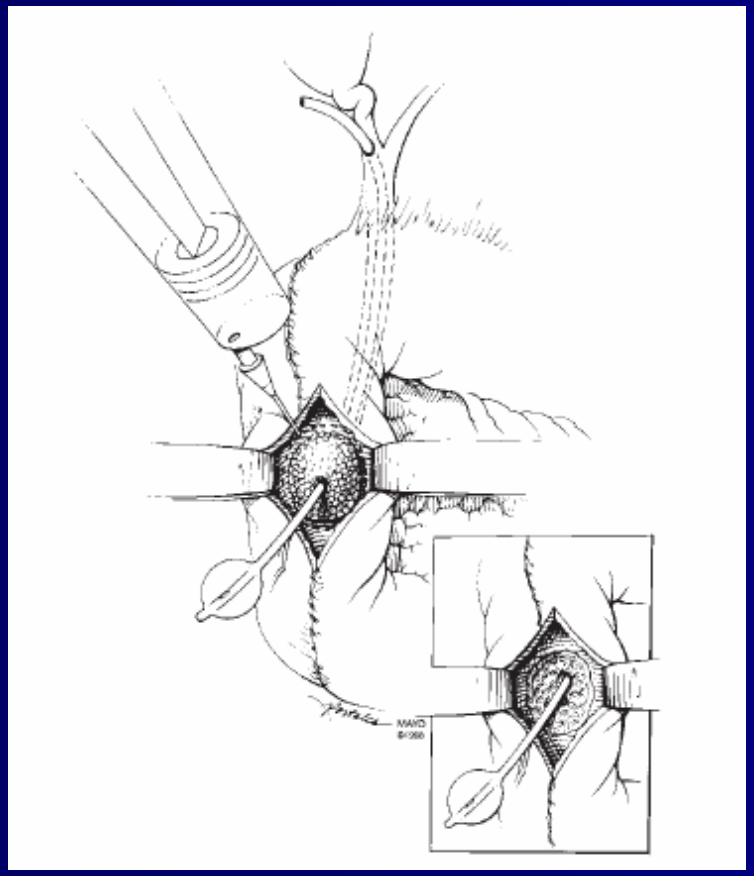
Villous adenoma

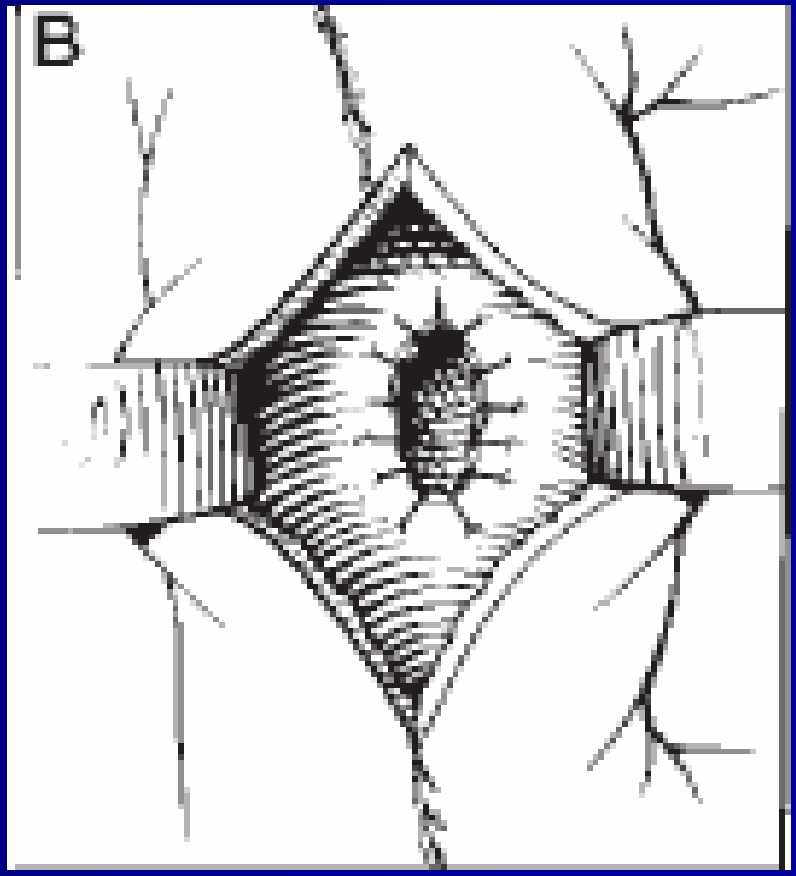
- 40 to 50 % harbour adenocarcinoma.
- Size is not related to their malignant potential
- The majority of tumors are located around the papilla of Vater and therefore present with symptoms earlier than other duodenal tumours.

Villous adenoma

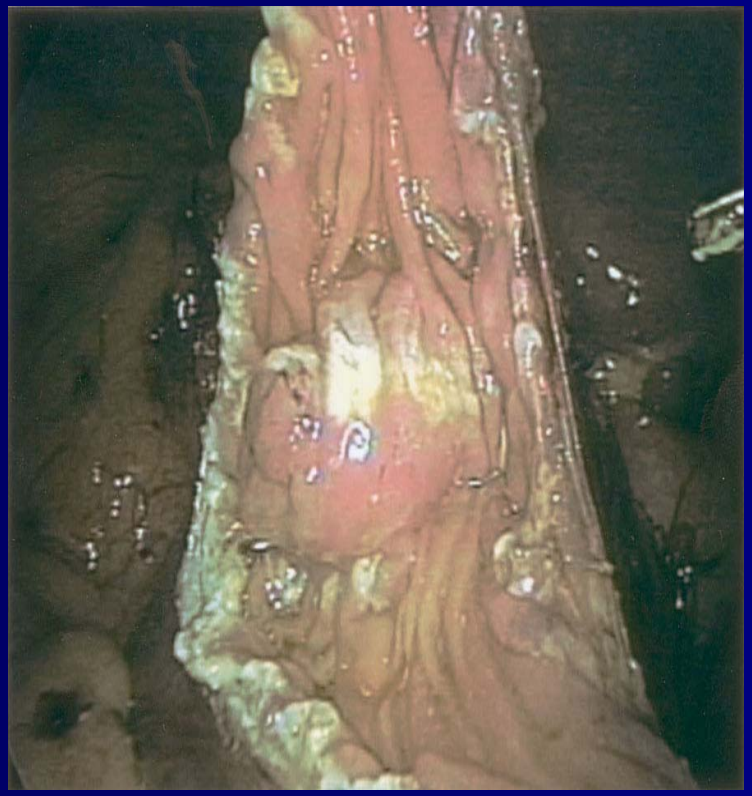
- In the few cases with a tumor which is small and pedunculated, endoscopic resection is possible.
- Benign villous adenomas can be locally excised
- Local recurrence occurs in 20 to 50 % of pts

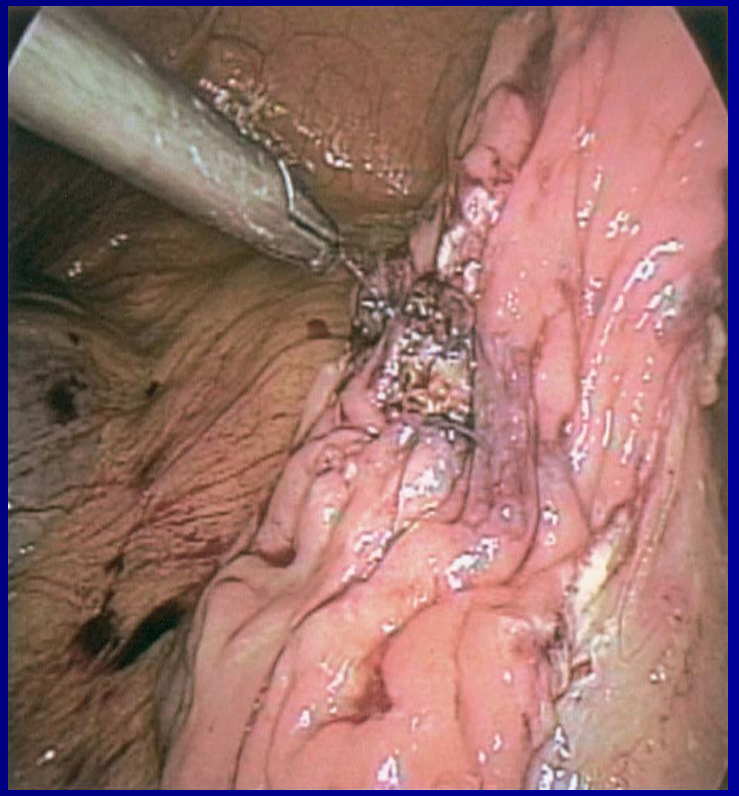
Local excision of periampullary villous tumor of the duodenum





Laparoscopic resection of a periampullary villous adenoma





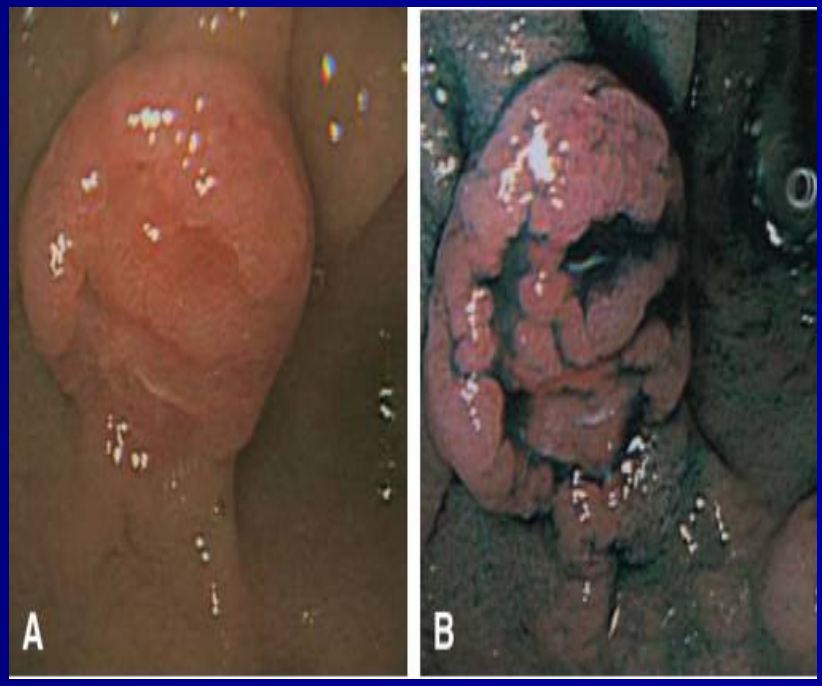
Malignant duodenal tumours

Adenocarcinoma

- Primary adenocarcinomas of the duodenum are rare, accounting for < 0.5 % of all carcinomas of the GIT.
- The duodenum is the most common site of carcinoma in the small bowel, accounting for 50 % of all cases.
- 20 % arise in villous adenomas.

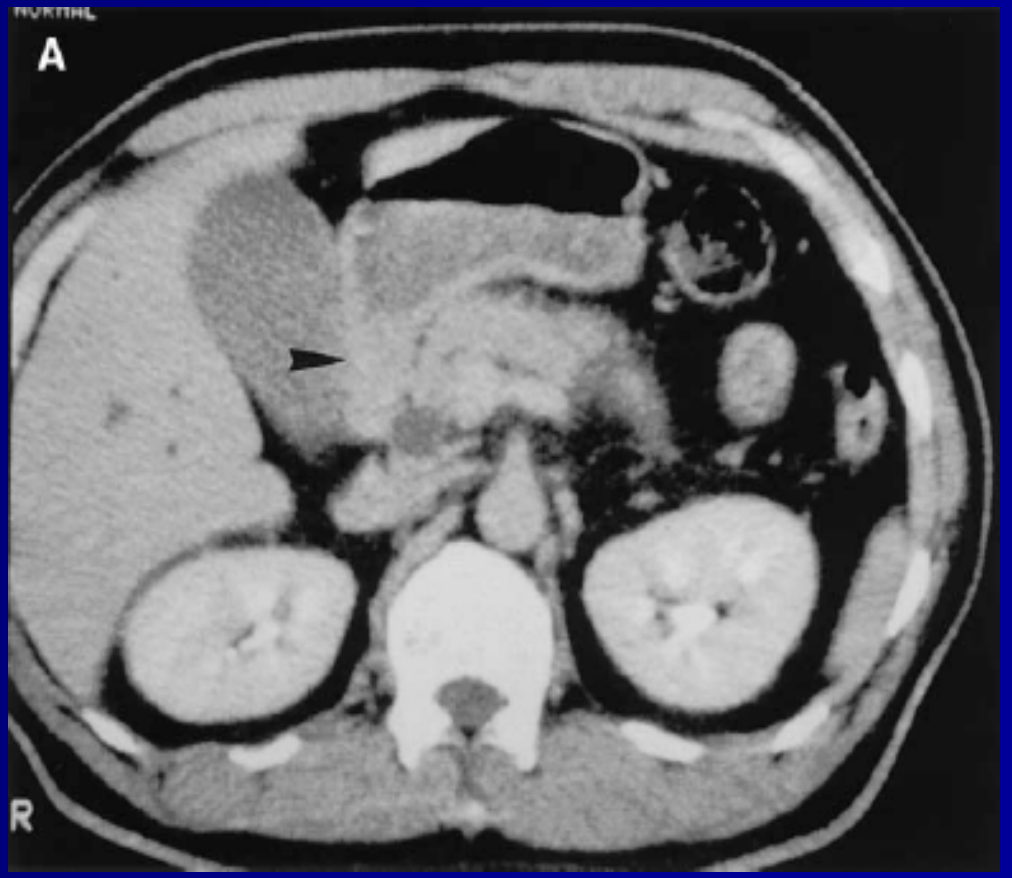
Adenocarcinoma

- Macroscopically their appearance ranges from ulcerating and infiltrating to polypoid.



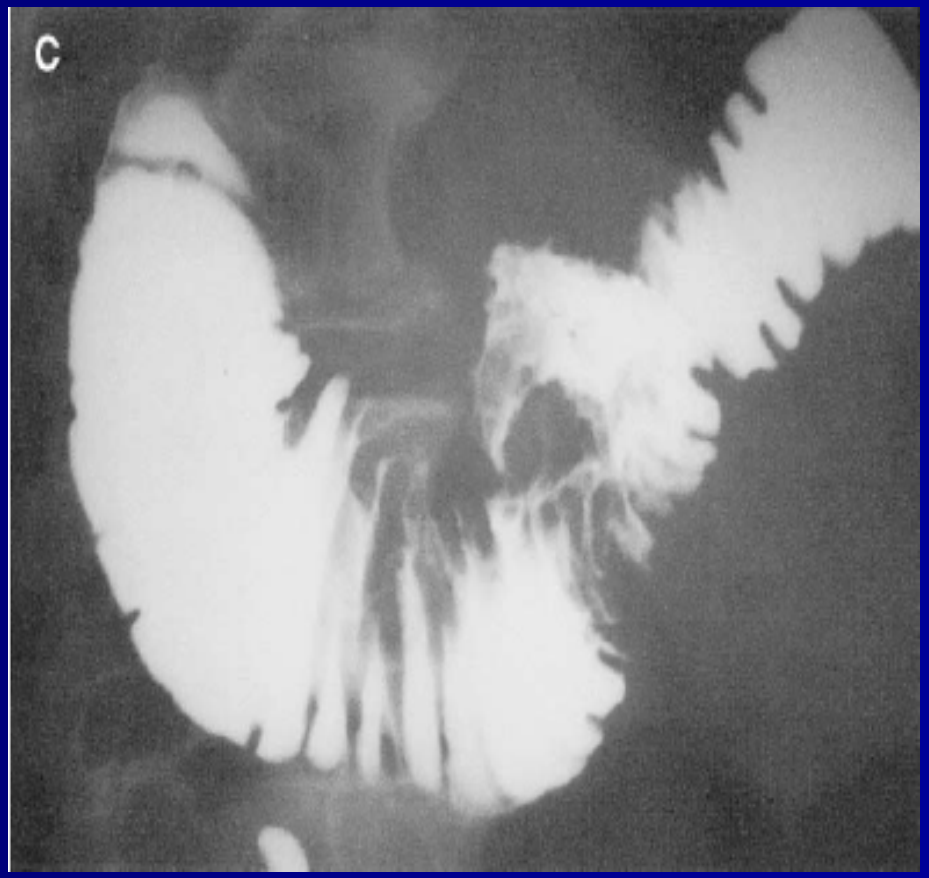
Adenocarcinoma

- On CT, concentric or asymmetrical thickening of the bowel wall is typical findings for duodenal adenocarcinoma



Adenocarcinoma

- Upper gastrointestinal barium studies show irregular filling defects with mucosal derangement of the third portion of duodenum caused by duodenal cancer.



Adenocarcinoma

- Endoscopic resection for early duodenal carcinoma can be an effective treatment.
- Transduodenal resection is an inadequate operation for invasive duodenal carcinoma (significant risk for recurrence)
- The treatment of choice is pancreaticoduodenectomy.
- Only small tumours in D4 should be treated with distal duodenectomy and duodenojejunostomy.

Adenocarcinoma

- Curative resection in these patients carries a 50 to 70 % chance of 5-year survival.
- In pts with resectable lymph node involvement, the 5-year survival is 20 %.
- Up to half of all patients with adenocarcinoma of the duodenum have unresectable lesions, and only occasionally survive for more than 1 year.

Adenocarcinoma

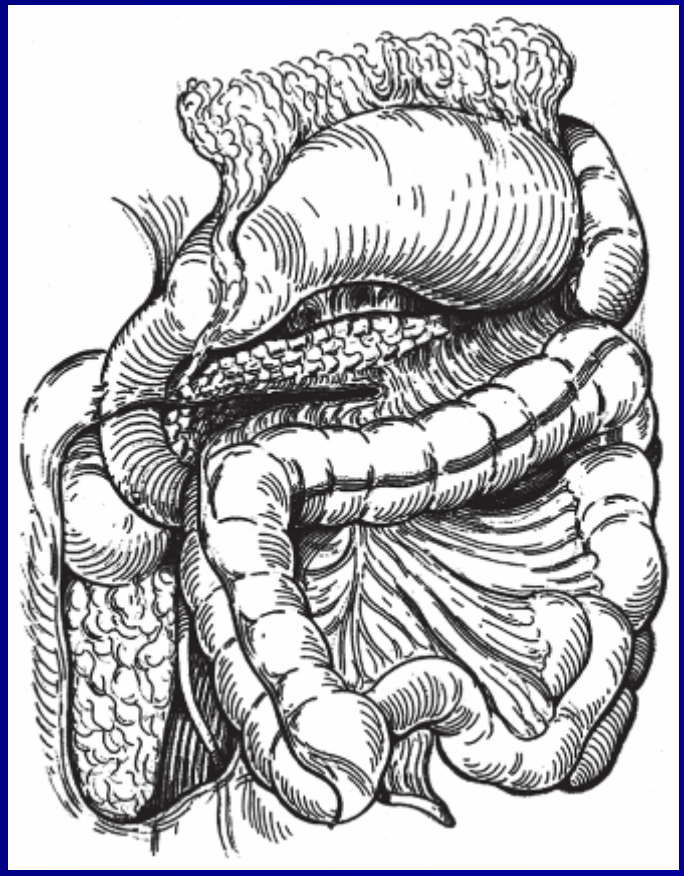
Table 6. Published Results Including More Than 50 Patients With Duodenal Adenocarcinoma

Source	Study Years	No. of Patients	Resectability, %	Lymph Node Positive, %	5-Year Survival After Resection, %
Alwmark et al[2]	1958-1973	66	43	. . .*	. . .
Joesting et al[9]	1937-1977	104	51	. . .	46
Rotman et al[11]	1978-1988	66	71	37	45
Barnes et al[5]	1967-1991	67	61	36	54
Sexe et al[12]	1987-1991	85	44	. . .	23
Santoro et al[10]	1980-1994	89	73	23	25
Rose et al[4]	1983-1994	79	63	38	60
Sohn et al[3]	1984-1996	55	87	56	53
Present study	1976-1996	101	67	32	54

* *Ellipses indicate values not stated in the study cited.*

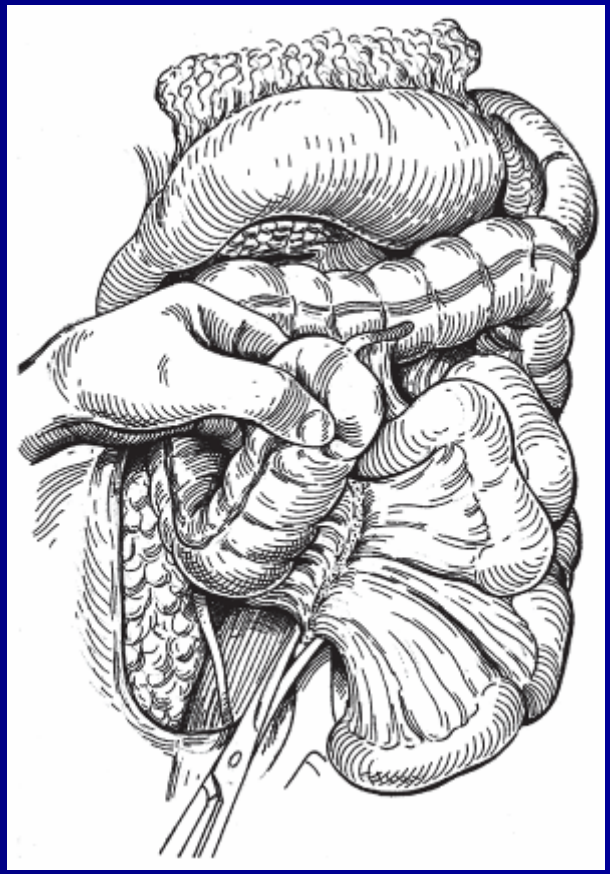
Description of Intestinal Derotation Procedure

- The duodenojejunal flexure is identified, and the proximal jejunum retracted caudally while the peritoneum is incised along its left side.
- The large intestine from the cecum to the midpoint of the transverse colon is extensively mobilized to allow complete rotation of the ileal loops .



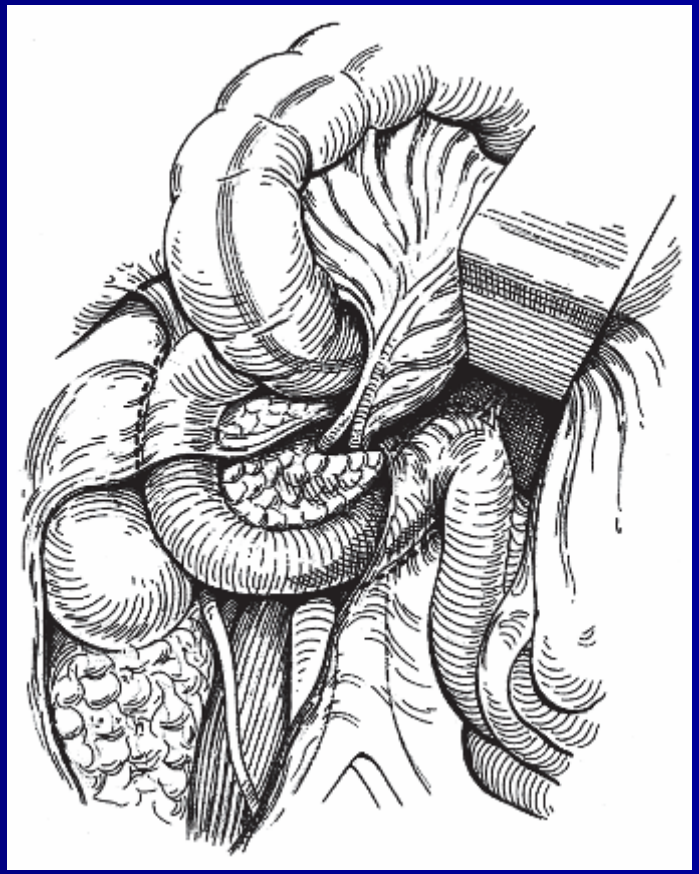
Description of Intestinal Derotation Procedure

- The secondary root of the small-bowel mesentery is totally mobilized upward as far as the third portion of the duodenum



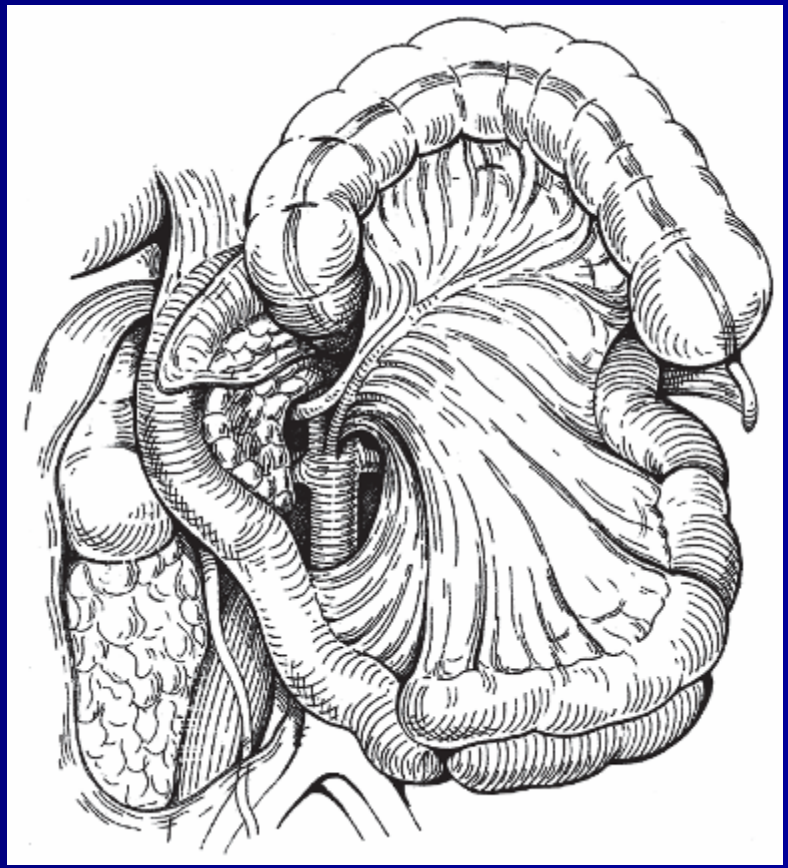
Description of Intestinal Derotation Procedure

- The duodenum is exposed by division of the peritoneum lateral to its second and third part and the ligament of Treitz is divided along the anterior cranial aspect of the 3-4 DC



Description of Intestinal Derotation Procedure

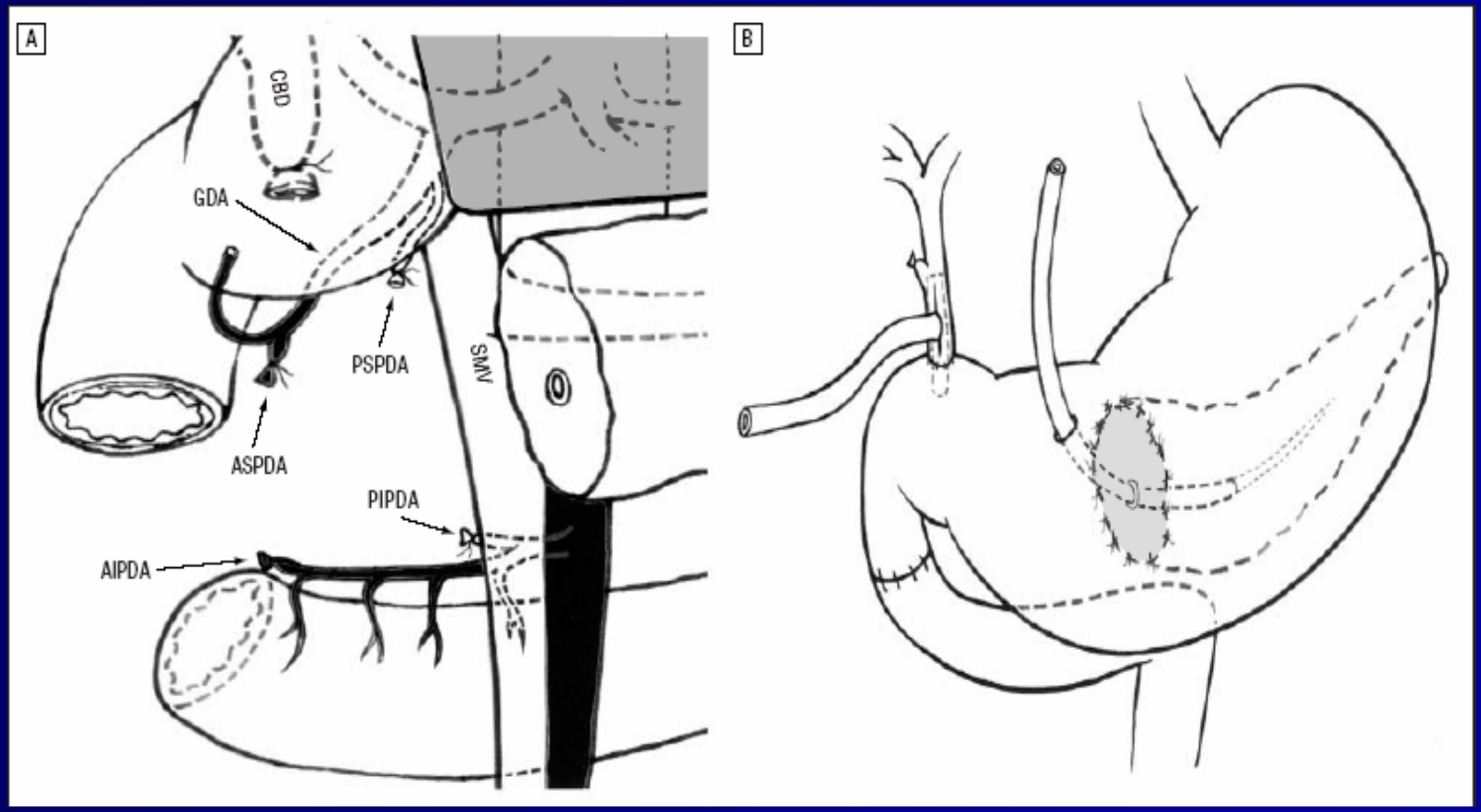
- The 3-4 DC and related mesentery can then be easily moved to the right of the superior mesenteric artery
- Intestinal continuity is restored by an end-to-end duodenojejunal anstomosis. Drainage of the anastomotic site is always provided



Adenocarcinoma of the Third and Fourth Portions of the Duodenum

- DS associated with intestinal derotation represents an approach that is radical, but is associated with negligible rates of morbidity and mortality.

PHRSD



PHRSD

- Indications for doing PHRSD should include benign or lowgrade malignant ampullary and parapapillary duodenal lesions as well as benign pancreatic head lesions.

- It is believed that the preserved relatively short segment may play a significant role in absorbing the iron, calcium, fat, folic acid, and so on.

Carcinoid

- Carcinoid tumours are the second most common malignant lesion in the duodenum after adenocarcinomas.
- Most tumours measure < 1 cm in diameter

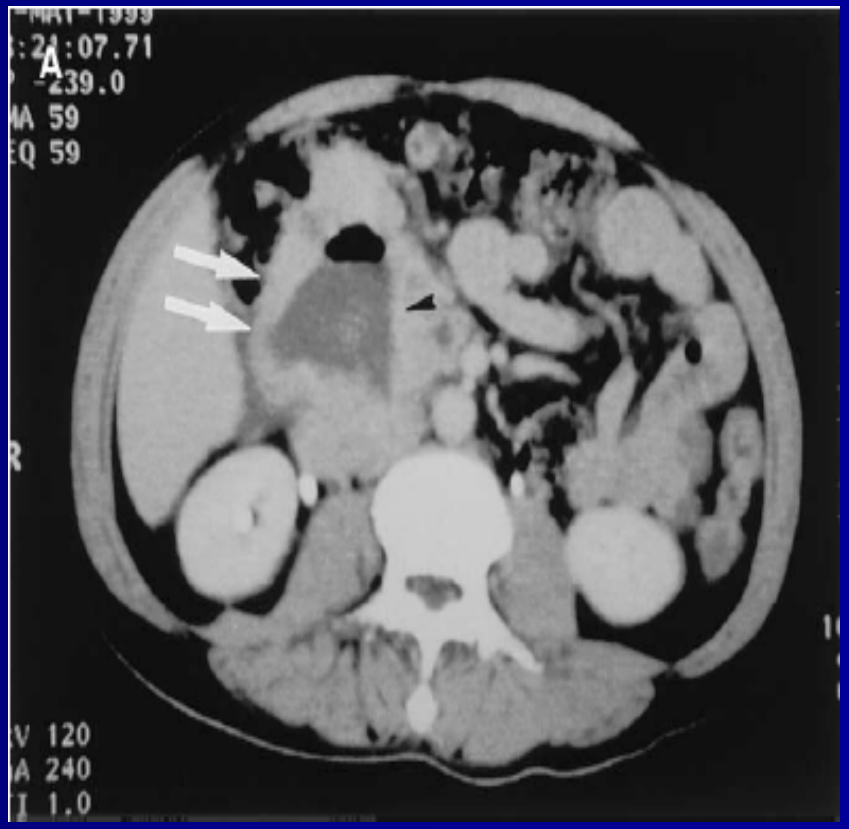
- Local excision is sufficient for benign tumours less than 1.5 cm in diameter
- For larger or invasive tumors the rules for resection of adenocarcinoma apply.
- The prognosis is better than for patients with adenocarcinomas, with overall 5-year survival rates 50 to 75 %.

GIST

- They are least common in duodenum.
- They are more common in males than females and are also more common in the fifth and sixth decades.

GIST

- GIST tumors typically appear exophytic and can be bulky .
- Central necrosis or ulceration is also common.



Lymphoma

- Only 5 % of all lymphomas are primary intestinal lymphoma and less than 10 % of these are located in the duodenum.
- Thickening of bowel wall is a characteristic CT finding for the lymphoma .