Acalculous disorders of the biliary tract

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Acalculous Cholecystitis

Definition

Gallbladder inflammation in the absence of cholelithiasis (5–10% of patients with cholecystitis)

Riskfactors

Major surgery
Critical illness
Extensive trauma
Burn-related injury

Male
>50 years
Parenteral nutrition (Risk of sludge formation)

Salmonella or cytomegalovirus infections in immunocompromised hosts

Systemic vasculitides (polyarteritis nodosa, systemic lupus erythematosus)

Pathogenesis/Clinic

A combination of biliary stasis, chemical inflammation and ischemia

“typical” cholecystitis symptoms may be absent, especially in elderly patients

Unexplained fever and/or hyperamylasemia ->exclusion of acalculous cholecystitis

Cave: More and ealier complications in acalculous cholecystitis than in calculous cholecystitis.

50–70% have gangrene, empyema, or perforation of the gallbladder at the time of surgery

Mortality rates of 10–50% are observed

Diagnosis/Therapy

By ultrasonography, gallbladder wall thickening (>4 mm)

CAVE: False-positive results from prolonged fasting

Cholecystectomy, but sometimes contraindicated in severly ill patients. Supportive treatment with antibiotics.

Percutaneous or EUS guided drainage is an alternative

Gallblader Polyps

Epidemiology

Prevalence 1–4%

Gallbladder polyps have been observed in 0.004-3.8% of resected gallbladders, and in 1.5-4.5% of gallbladders assessed by ultrasonography

Majority of the polyps (95%) are benign

Cholesterol polyps>inflammatory polyps>gallbladder adenomas (<1%)

The frequency of association between gallbladder adenomas and carcinoma is unknown.

Gallbladder adenomas greater than 10 mm have a carcinoma risk

High rates of GBC are seen in South American countries, particularly Chile, Bolivia, and Ecuador, as well as India, Pakistan, Japan, and Korea

Diagnosis/Therapy

Ultrasound can help distinguish polyps from cholelithiasis

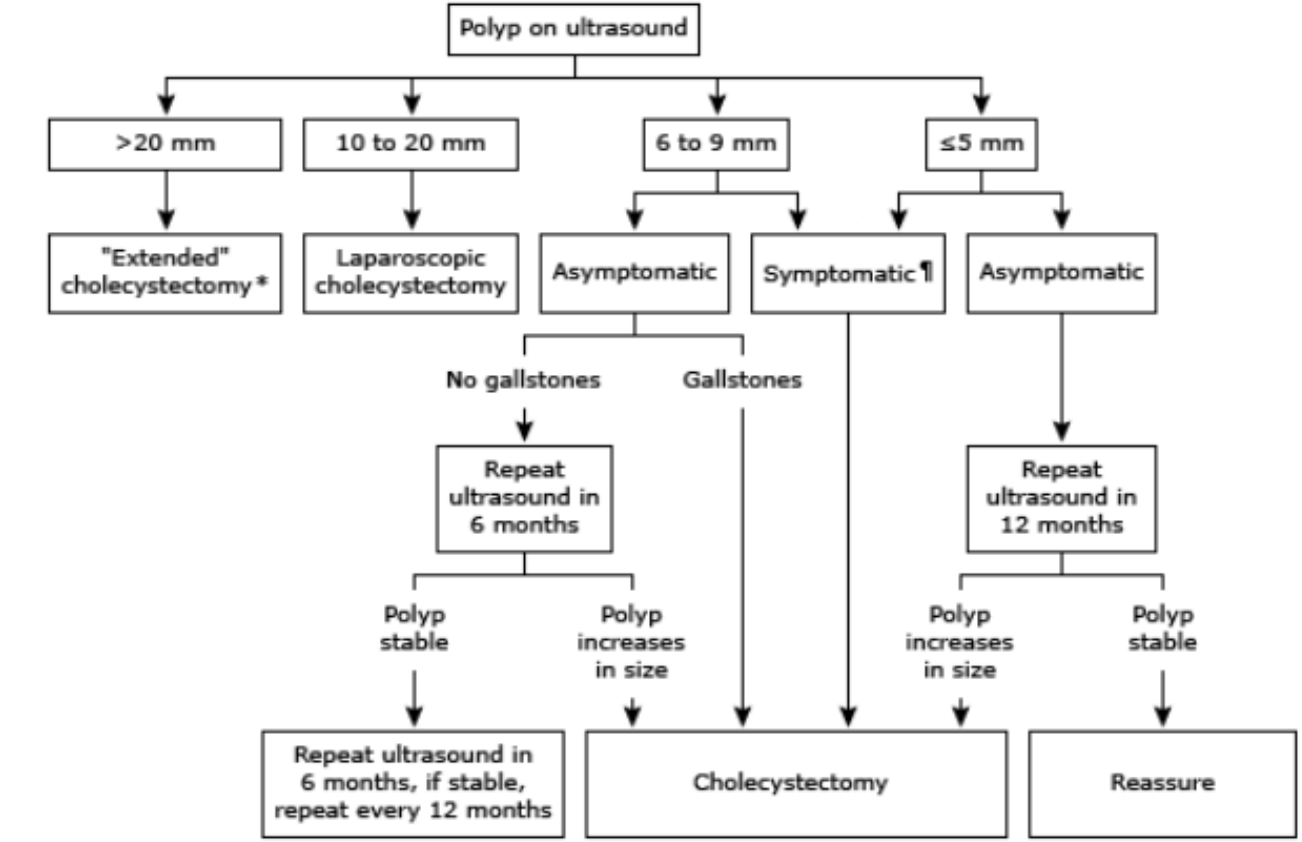
Polyps <10 mm, no associated cholelithiasis, <50 years -> No cholecystectomy

Polyps between 10–18 mm in diameter have a small but appreciable risk of carcinoma -> cholecystectomy is recommended in good operative candidates.

Polyps >18 mm in diameter have a significant risk of carcinoma and require cholecystectomy.

Patients with PSC and polyps of any size should be considered for cholecystectomy. PSC Patient need yearly sonography

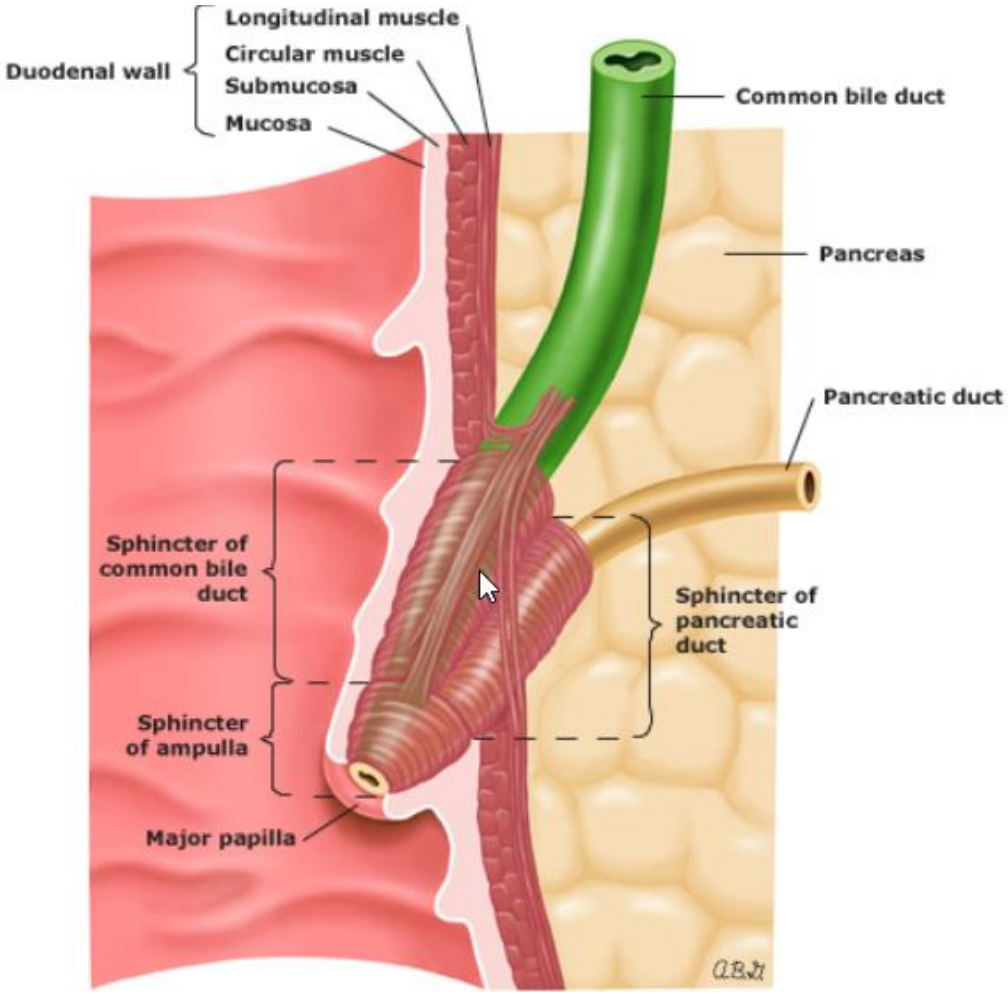
A suggested algorithm for managing gallbladder polyps found on ultrasound



* An extended cholecystectomy includes lymph node dissection and partial hepatic resection in the gallbladder bed.

¶ Symptoms: Biliary type pain, common duct obstruction, cholangitis, or recurrent pancreatitis. Dyspepsia is not an indication for surgery.

Biliary cysts



Epidemiology

Incidence 1:100’000 in western an 1:13’000 in Asian population

Congenital anomalies

Females>Males 3-4x

Cysts are primarily a disease of children and young adults (80% of diagnosis)

Anatomy/Pathophysiology

Often long common channel (insertion of the CBD more than 15 mm from the ampulla of Vater). Less than 2% of the population

Anomalous junction of the pancreaticobiliary duct leads to pancreatic reflux into the biliary tree

Amylase levels in the fluid contained in the gallbladder are typically elevated

Clinical manifestation

In infancy:

- Classical trias: abdominal pain, jaundice and a palpable mass
- Jaundice is the most common sign
- Vomiting
- Hepatomegaly
- Can lead to biliary cirrhosis and portal hypertension
- Presentation may be identical to that of biliary atresia

Adults:

- Most common is epigastric pain
- Intermittent jaundice
- Recurrent cholangitis
- Abdominal masses, cirrhosis, and portal hyper-tension are less common than in the infantile form

Link to other anomalies

Type V can be associated with polycystic kidney disease (autosomal recessive inherited mutation in PKD1)

Sclerosing cholangitis

Congenital hepatic fibrosis

Pancreatic cyst

Congenital cardiac anomalies (31% of pediatric patients with CC)

Biliary, duodenal or colonic atresia

Hemifacial microsomia with extracraniofacial anomalies (OMENS plus syndrome)

Familial adenomatous polyposis

Complications

- recurrent cholangitis
- stone formation
- stricture formation
- pancreatitis
- biliary cirrhosis
- liver abscess
- cyst rupture (rare, often in children)

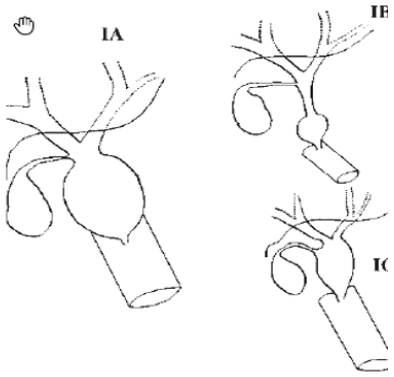
6-30% risk of developing malignancy. The risk is low in childhood (<1%) but increases to about 30% to 40% with age greater than 50 years.

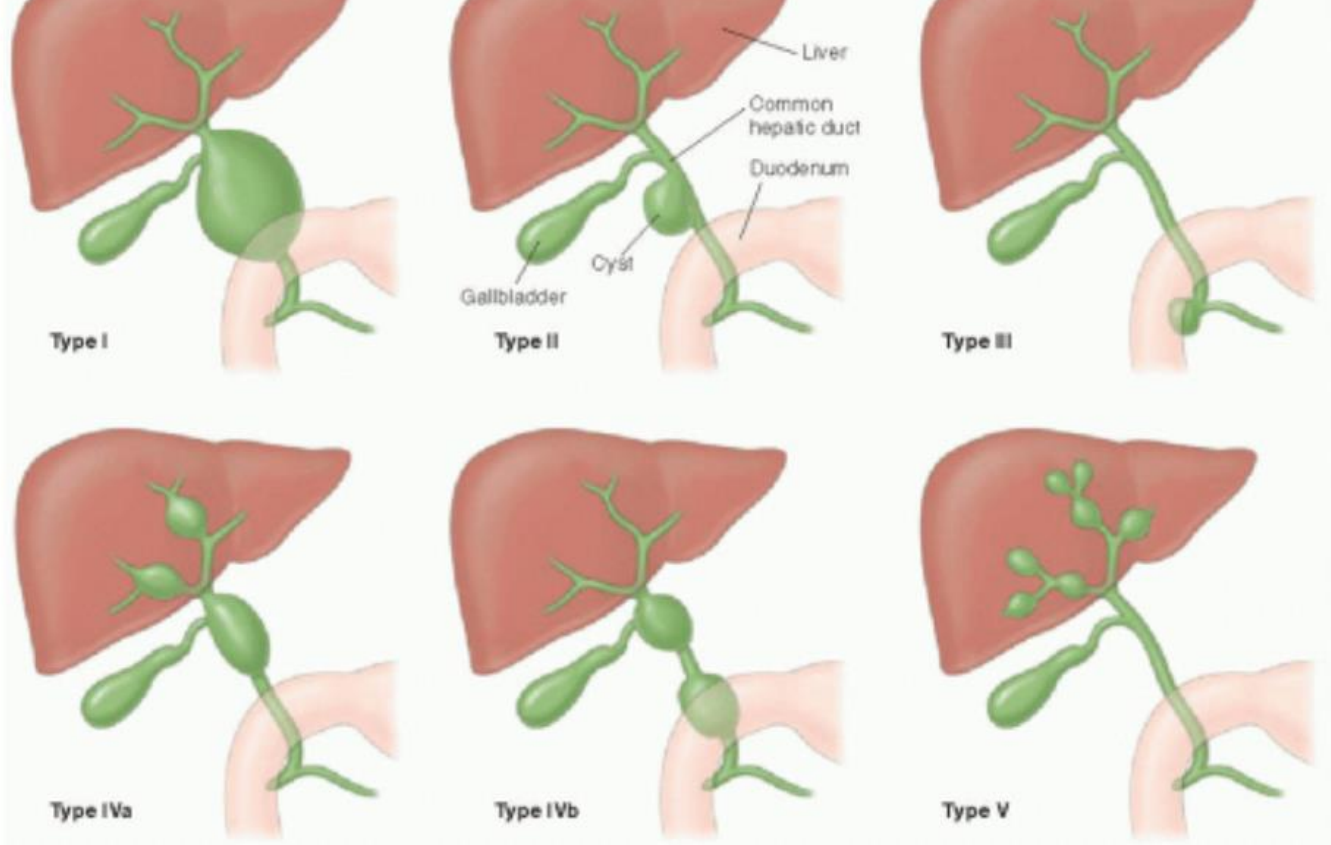
Todani Classification

Type I dilation of the extrahepatic duct
Type II diverticula in the extrahepatic ducts
Type III choledochoceles
Type IVa multiple intra- and extrahepatic cysts
Type IVb multiple cysts of the extrahepatic ducts only
Type V single or multiple cystic dilations of the intrahepatic ducts

Type I (most common, 65-85%)
Type V = Caroli's disease

Classification





Diagnosis

Ultrasonography, CT and MRI may be useful
ERCP more detailed anatomic information for planning operative approaches

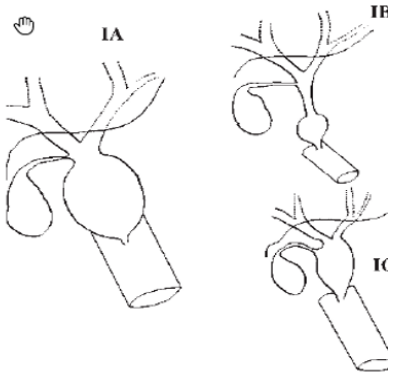
Typ IV can be difficult to distinguish from polycystic liver disease and primary sclerosing cholangitis

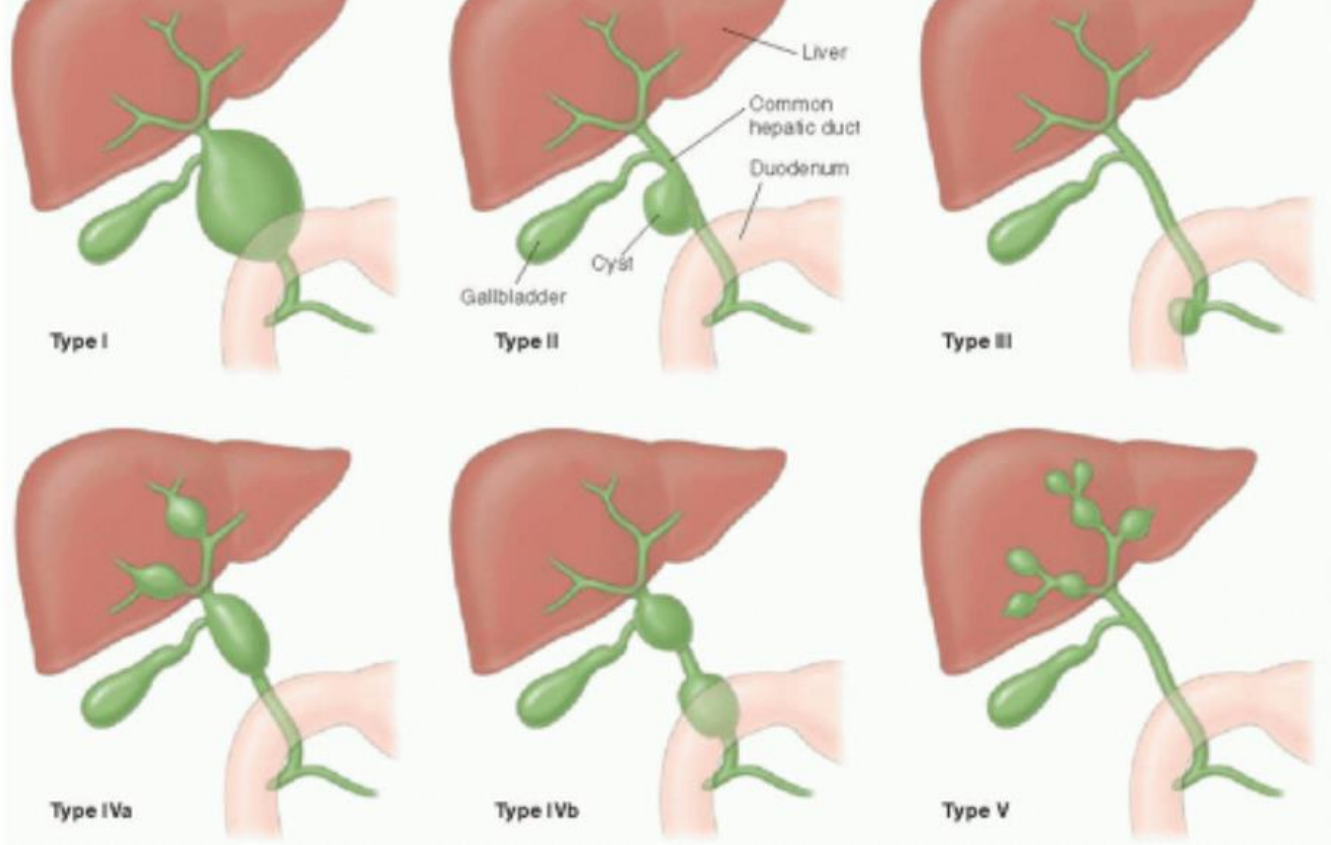
Therapy

Surgery for type I or type IV (cyst excision and reconstruction of the extrahepatic biliary tree by choledochojejunostomy or hepatico-jejunostomy)

Type II cysts should be excised

Classification





Therapy

Surgery for type I or type IV (cyst excision and reconstruction of the extrahepatic biliary tree by choledochojejunostomy or hepatico-jejunostomy)

Type II cysts should be excised

Type III cysts (Choledochoceles) have low premalignant potential. Can be cured by endoscopic biliary sphincterotomy.

Treatment of intrahepatic cysts (type IVa or V) depends upon the extent of liver involvement

Longterm postoperative follow-up is recommended: recurrent cholangitis, stone formation, strictures and pancreatitis

Cyst excision lowers risk of malignancy, but cancer can develop in other portions of the hepatobiliary tree.

HIV Cholangioapthy

HIV Cholangioapthy

HIV-associated biliary tract disease resembles sclerosing cholangitis with papillary stenosis

CD4 T cell count <50-100/mm3 is a risk factor for HIV cholangiopathy. 20% of Patients with AIDS cholangiopathy have CD4 count greater than 100/mm3

Prior to the advent of potent ART, AIDS cholangiopathy occurred in 26 % of AIDS patients.

Cryptosporidium parvum and CMV infection are usually associated with involvement of the large intrahepatic ducts

Sclerosing cholangitis without papillary stenosis, papillary stenosis alone, or long extrahepatic strictures are also possible presentations.

Symptoms

- Diarrhea related to smallintestinal involvement with characteristic pathogens (Cryptosporidium parvum, Cytomegalie virus)
- Epigastric pain
- Right upper quadrant abdominal pain
- Fever

- Often alkaline phosphatase is elevated (>75%) often mild increases in serum transaminases.
BUT 20% have normal laboratory

Therapy

Treat the biliary tree abnormalities AND the identified pathogens.

Biliary probes from the ERCP should be sent for histologic and microbiologic examination, (CMV), cryptosporidium, microsporidium and acid fast bacilli, as well as routine bacterial and fungal organisms)

Blood cultures and CMV-PCR

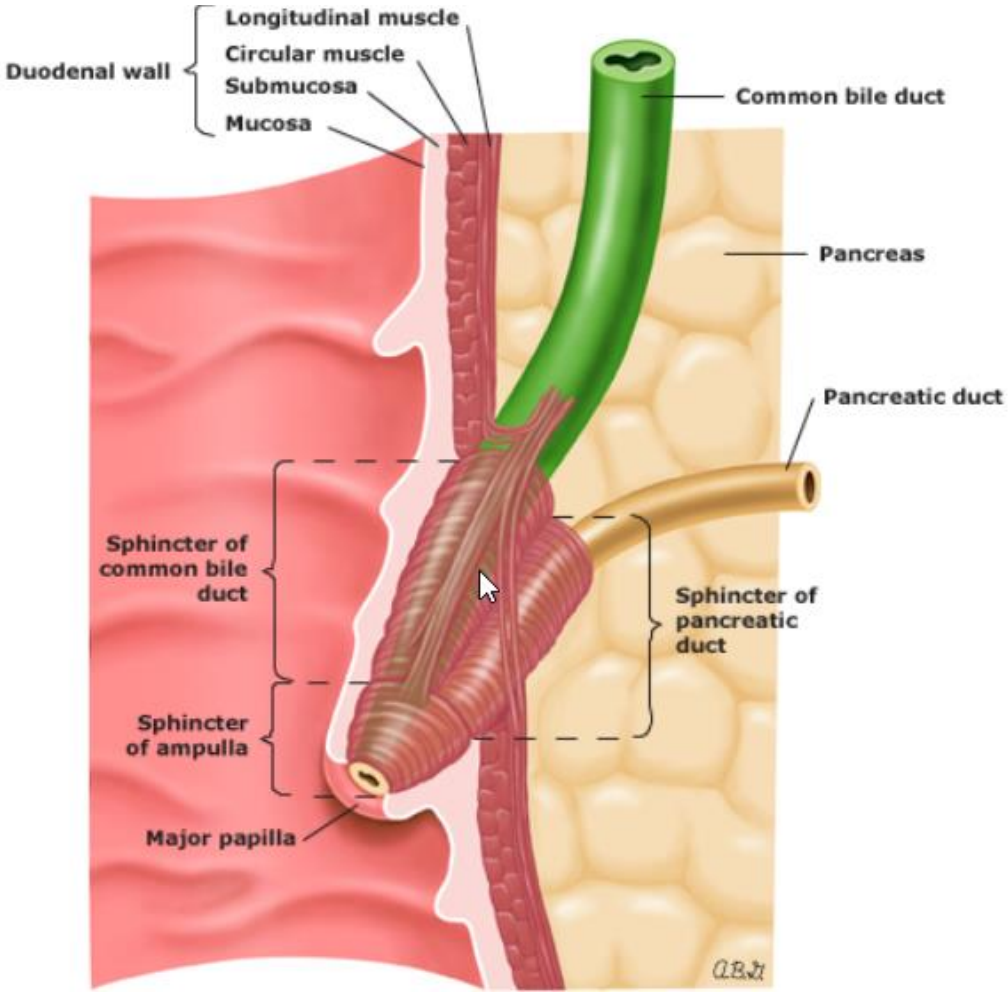
If only papillary stenosis alone (10%), endoscopic sphincterotomy

Up to half of affected patients will not have an identifiable opportunistic Pathogen

Ursodeoxycholic acid for intrahepatic cholestasis

Antiretroviral therapy (Benefit unclear)

Sphincter of Oddi Dysfunction



Definition

Sphincter of Oddi dysfunction (SOD) is an acalculous disorder associated with biliary-type pain with or without abnormal liver tests or recurrent pancreatitis

60% have histologic changes (muscular hypertrophy, inflammation, and fibrosis)

No infectious cause has been identified

Riskfactors: female gender, age in the 4th and 5th decades, recurrent abdominal pain after cholecystectomy

Less than 50% of patients have serum hepatic or pancreatic biochemical parameter elevations.

Diagnosis

Exclude upper gastrointestinal tract diseases. Always exclude choledocholithiasis or hepatic disease in patient with significant elevations in hepatic biochemistries

Bloodanalysis during episodes of biliary or pancreatic-type pain is most informative

SOD is diagnosed manometrically during ERCP when basal sphincter pressures exceed 40 mmHg (Goldstandard)

Nardi test = morphine-neostigmine provocation test (Positiv if amylase or lipase rises 4x and reproduction of pain after intramuscular injection of 10 mg morphine (to induce sphincteric spasm) and 1 mg prostigmine (to stimulate pancreatic exocrine secretions). CAVE: Can be positive in healthy people.

Sekretin MRI

Hepatobiliary scintigraphy

Classification

Type I:
Abnormal liver tests and a dilated common bile duct -> ERCP with endoscopic sphincterotomy (Classification Milwaukee AST or alkaline phosphatase >2–3 times elevated and dilated DHC >12mm)

Type II:
Either abnormal liver tests or a dilated common bile duct AND typical pain. Futher testing or empiric sphincterotomy in such patients (positive cost-effectiveness analysis)

Type III
Only Pain with normal liver tests and bile duct. Functional biliary type pain or other functional bowel diseases, such as dyspepsia or irritable bowel syndrome. Symptomatic treatment.

Rome IV

The following are the Rome IV criteria for biliary pain. In order to fulfill the Rome IV criteria, **all** of the following conditions must be met:

- Pain located in the epigastrium and/or right upper quadrant
- Episodes lasting 30 minutes or longer
- Recurrent symptoms occurring at different intervals (not daily)
- The pain builds up to a steady level
- The pain is severe enough to interrupt the patient's daily activities or lead to an emergency department visit
- The pain is not significantly (<20 percent) related to bowel movements
- The pain is not significantly (<20 percent) relieved by postural change or acid suppression

Functional biliary sphincter of Oddi disorder — The following are the Rome IV criteria for functional biliary sphincter of Oddi disorder:
- Criteria for biliary pain are fulfilled
- Absence of bile duct stones or other structural abnormalities
- Elevated liver enzymes or dilated bile duct, but not both

Supportive criteria include
- Normal amylase/lipase
- Abnormal sphincter of Oddi manometry
- Abnormal hepatobiliary scintigraphy

Functional pancreatic sphincter of Oddi disorder — The following are the Rome IV criteria for functional pancreatic sphincter of Oddi disorder:
- Documented recurrent episodes of pancreatitis (typical pain with amylase or lipase >3 times normal and/or imaging evidence of acute pancreatitis)
- Other etiologies of pancreatitis excluded
- Negative endoscopic ultrasound
- Abnormal sphincter manometry

Therapy

Type I and type II SOD patients with severe symptoms should be referred for ERCP
Type I patients nearly universally respond to endoscopic. Manometry is not required.

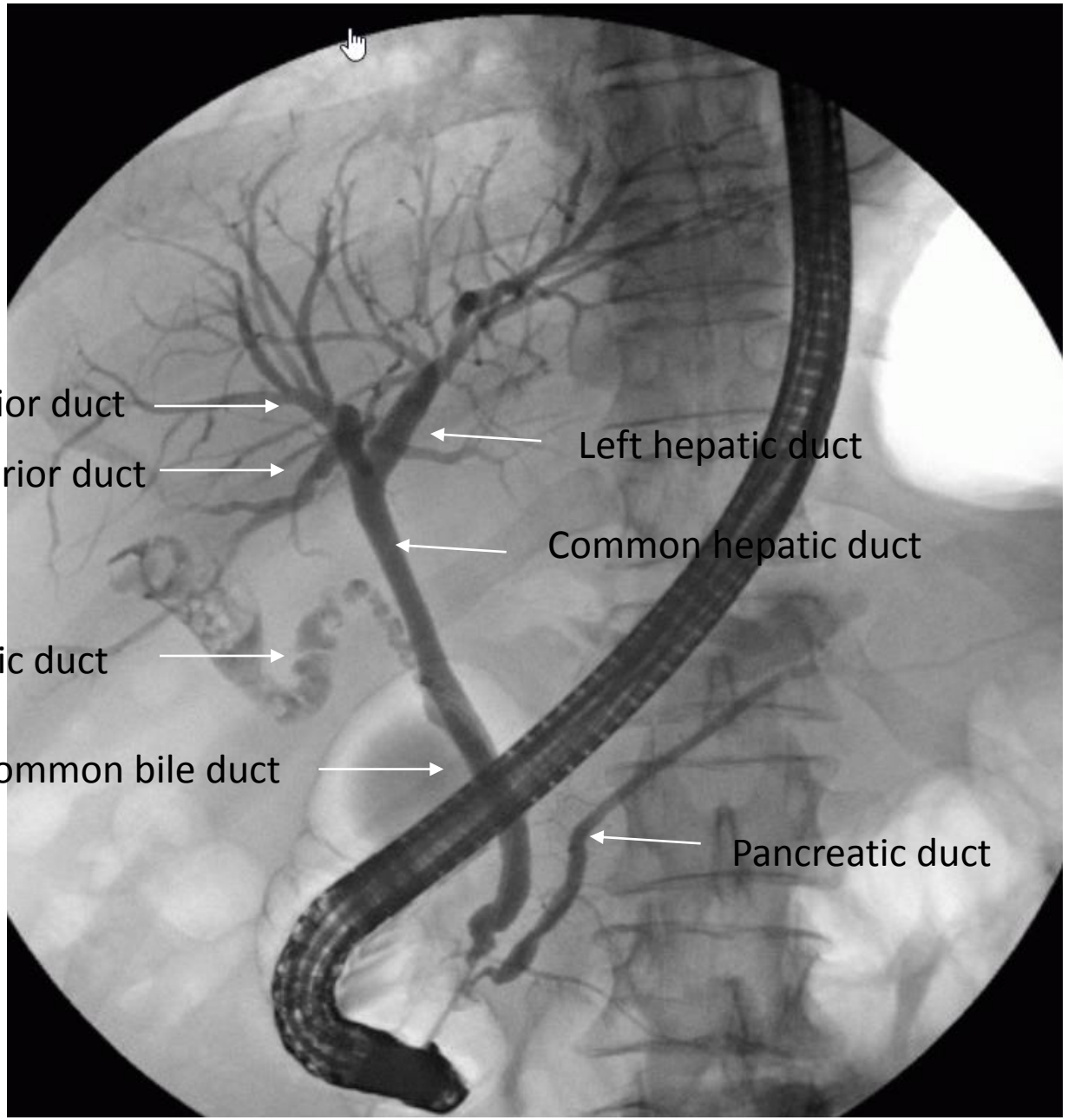
The majority of patients with type II SOD and abnormal manometry will have symptom relief after sphincterotomy. Lower success rates (55–60%) are observed with type III SOD (even with abnormal manometry)

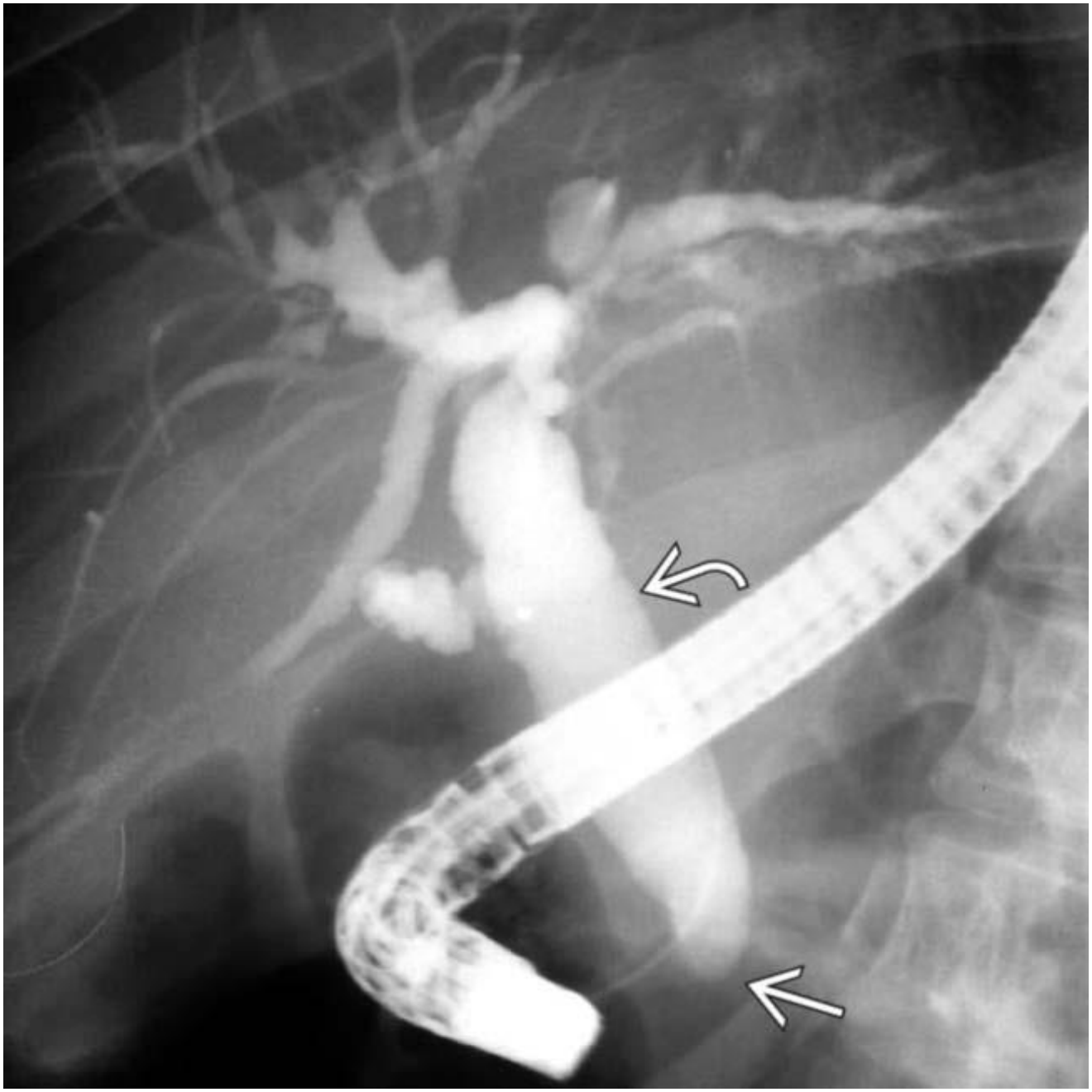
Nifedipine, Duspatalin

Cave

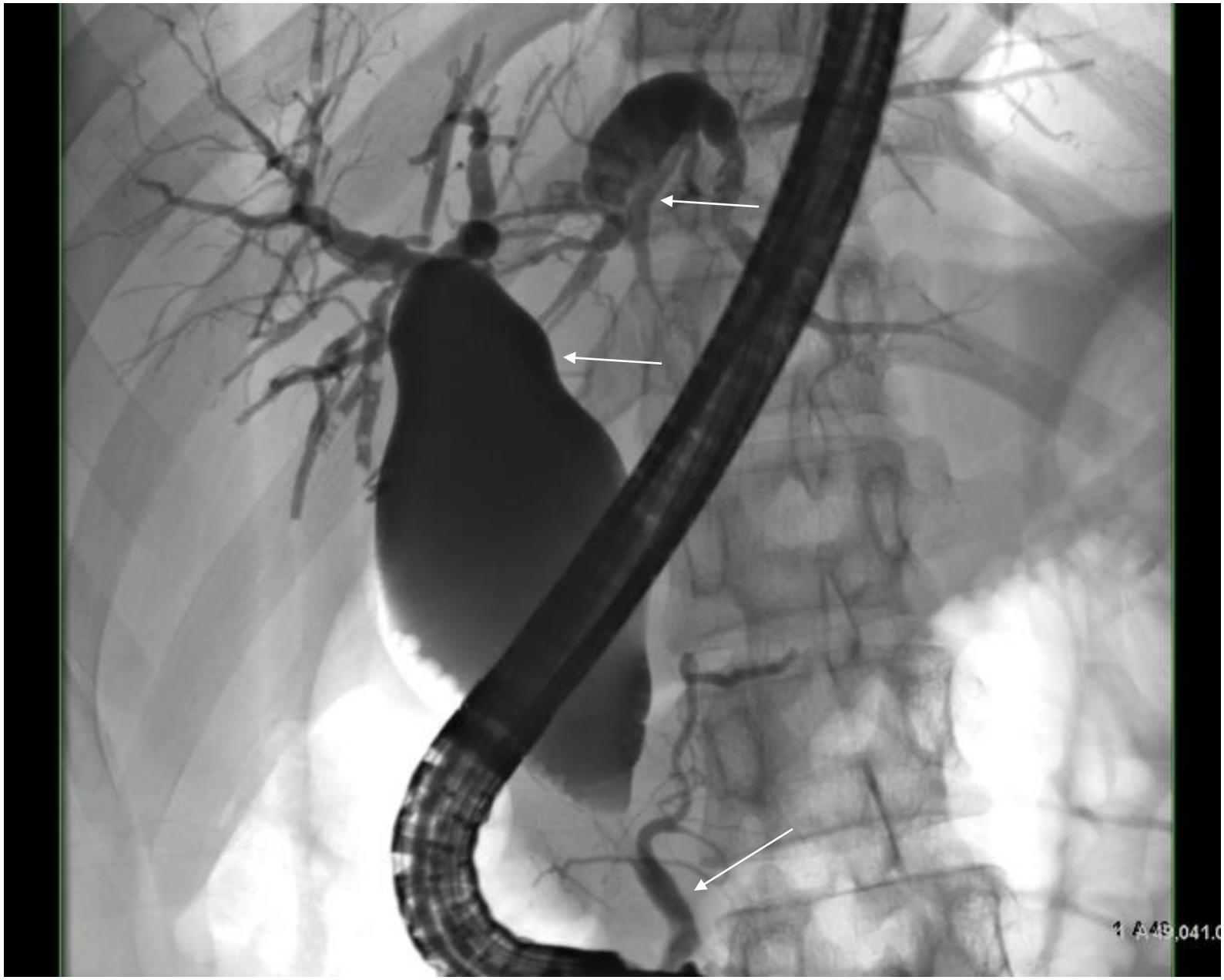
High risk of complications during sphincterotomy (2–33 times higher). 25% Pancreatitis (severe 1–3%)

Pancreatic duct stent placement is recommended in all patients undergoing SOD manometry.


Right anterior duct
Right posterior duct
Left hepatic duct
Common hepatic duct
Cystic duct
Common bile duct
Pancreatic duct

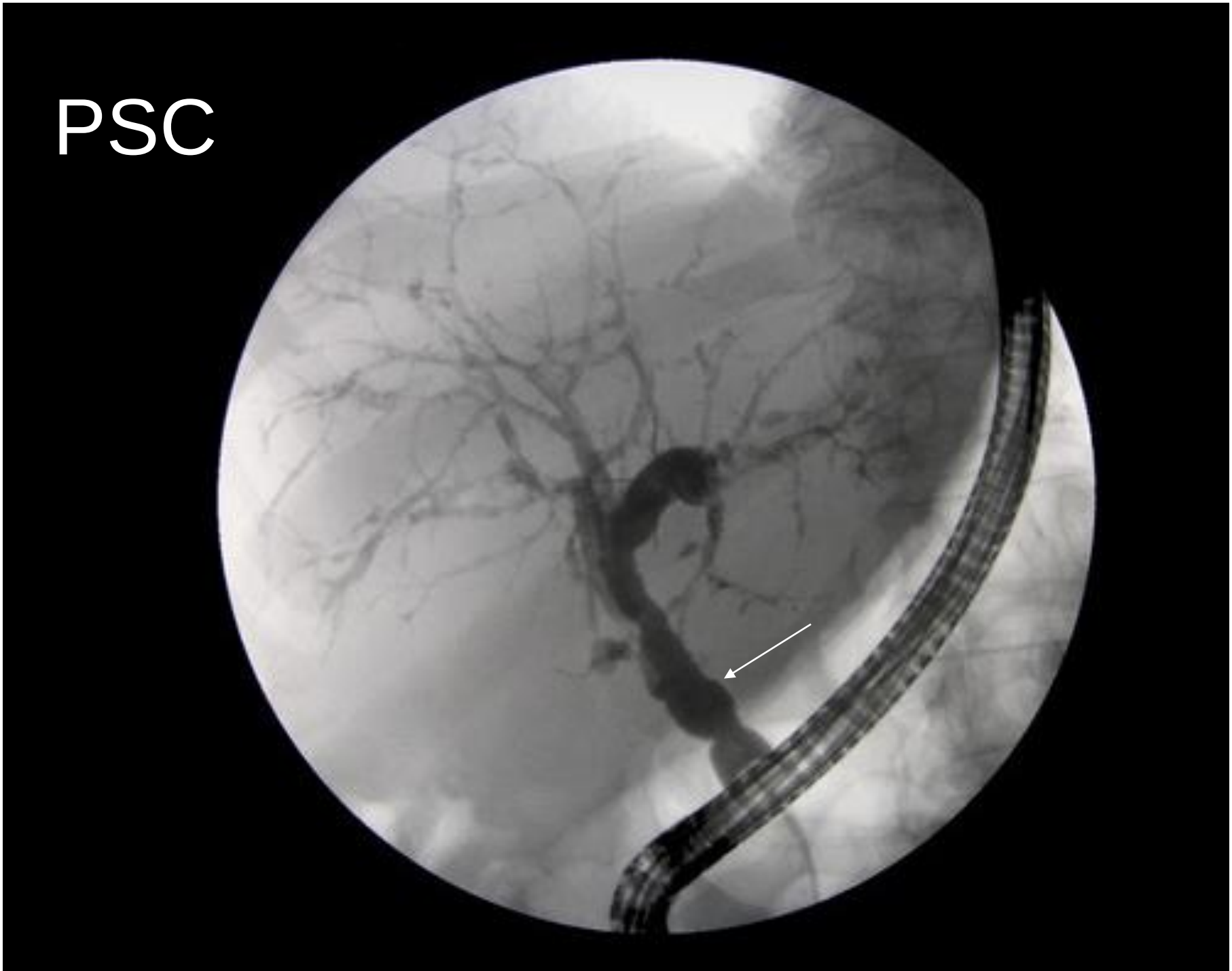


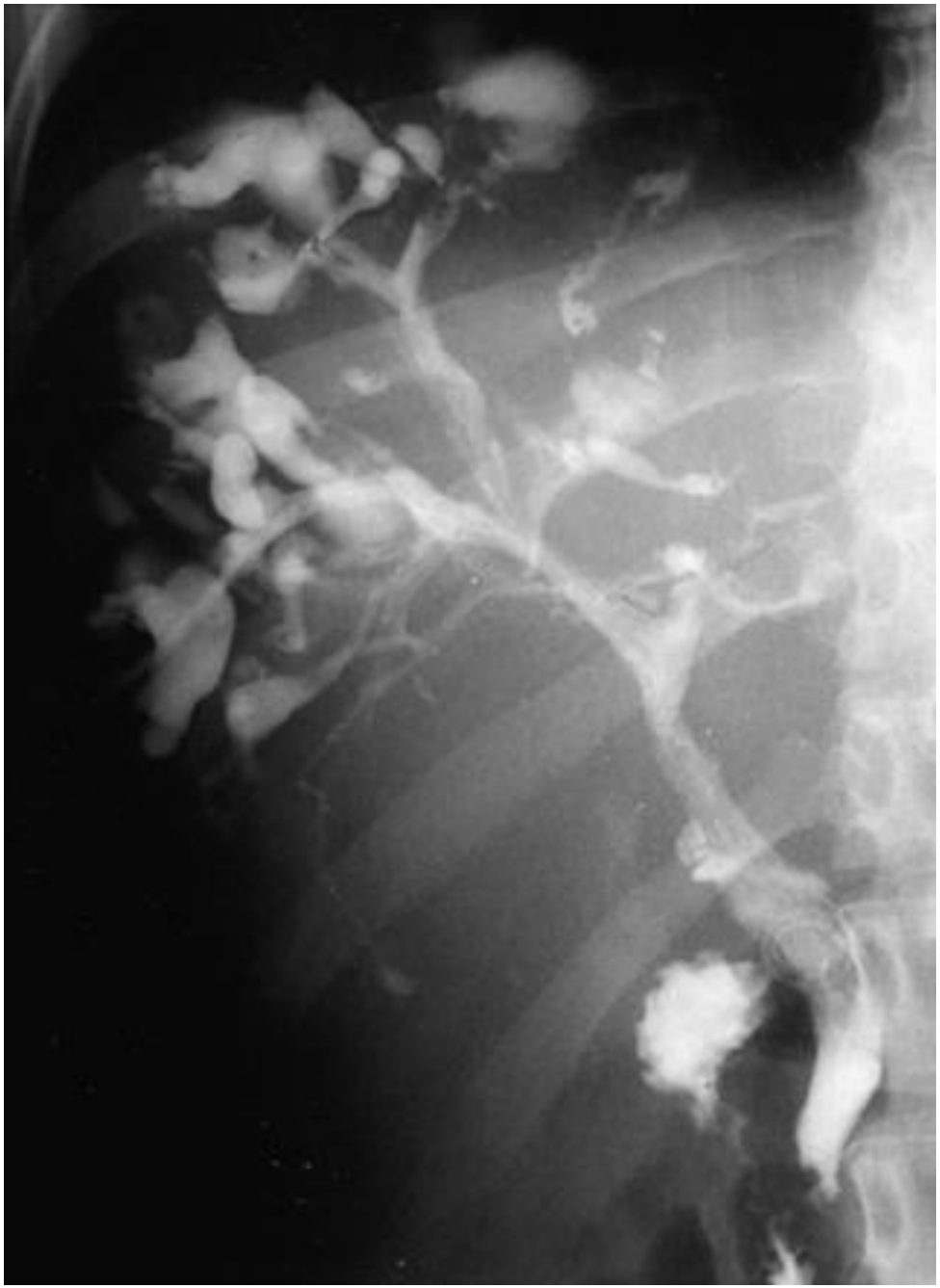
HIV Cholangiopathie



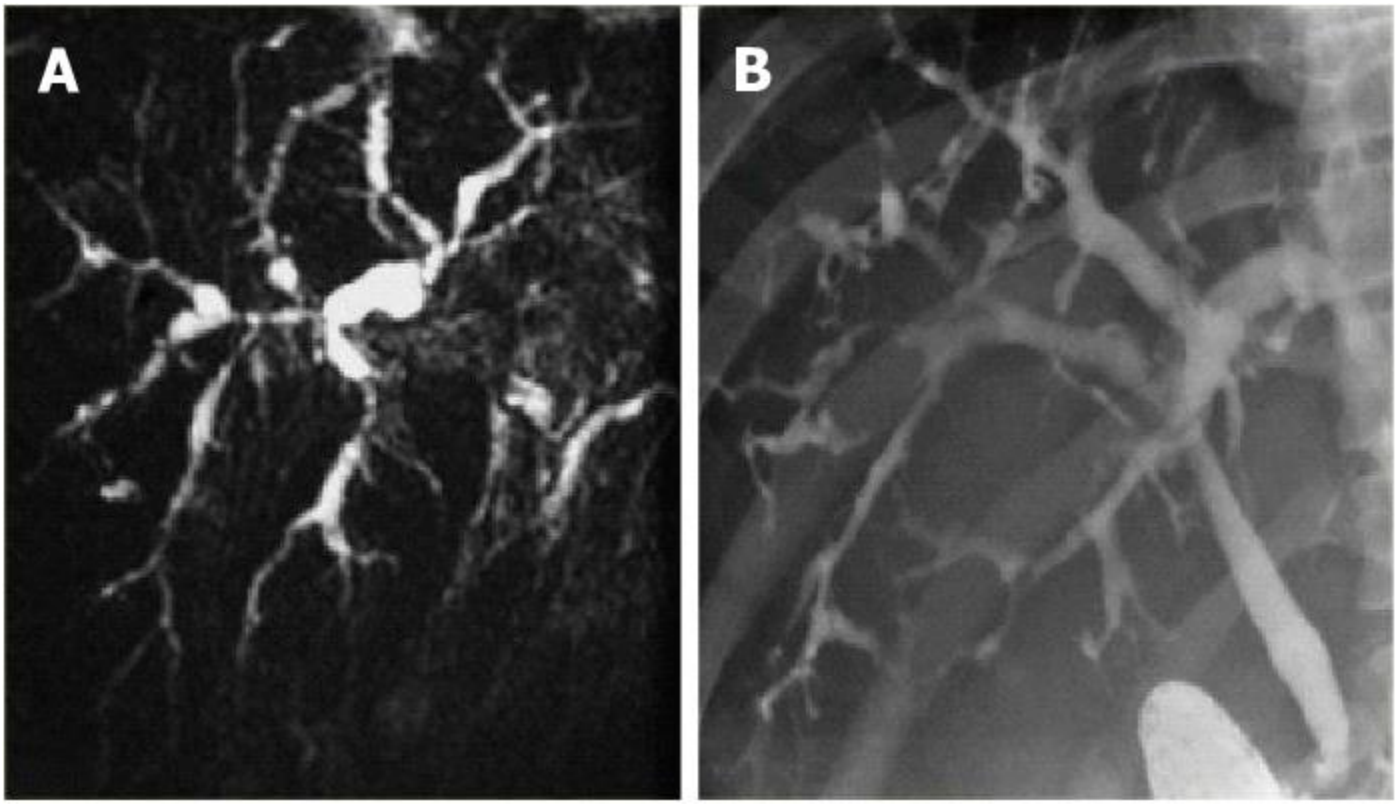
Typ IVa

PSC

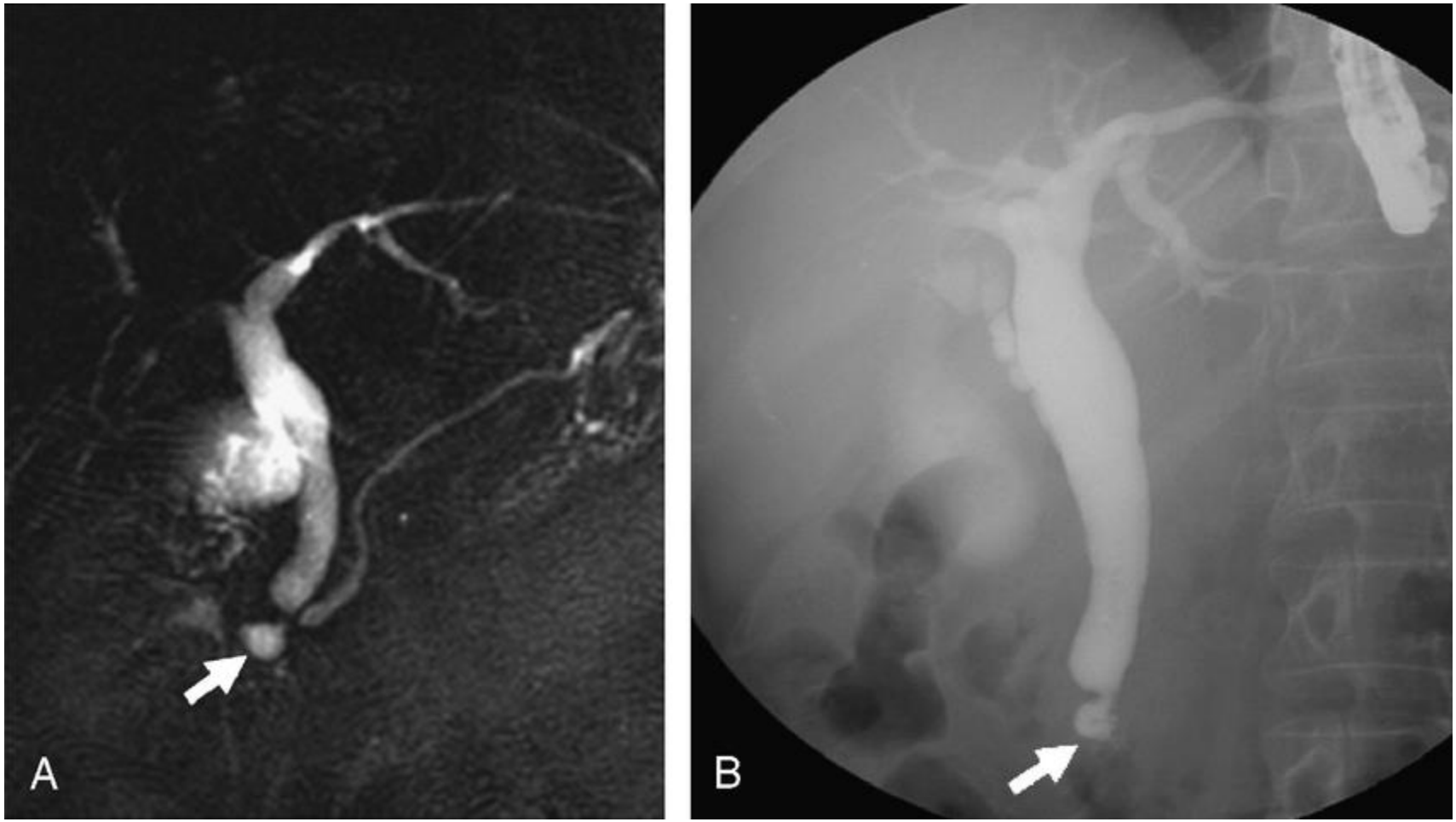




Todani V



PSC



Todany Typ III