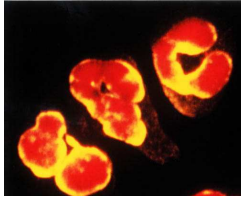


Sclerosing Cholangitis

- Primary
 - Immune-mediated (gut?)
- Secondary
 - Bacterial, recurrent pyogenic cholangitis
 - Ischemic cholangitis
 - IgG4-associated cholangitis
 - Posttraumatic, sepsis, burns
 - AIDS cholangiopathy
 - Portal biliopathy



Immunological Mechanisms

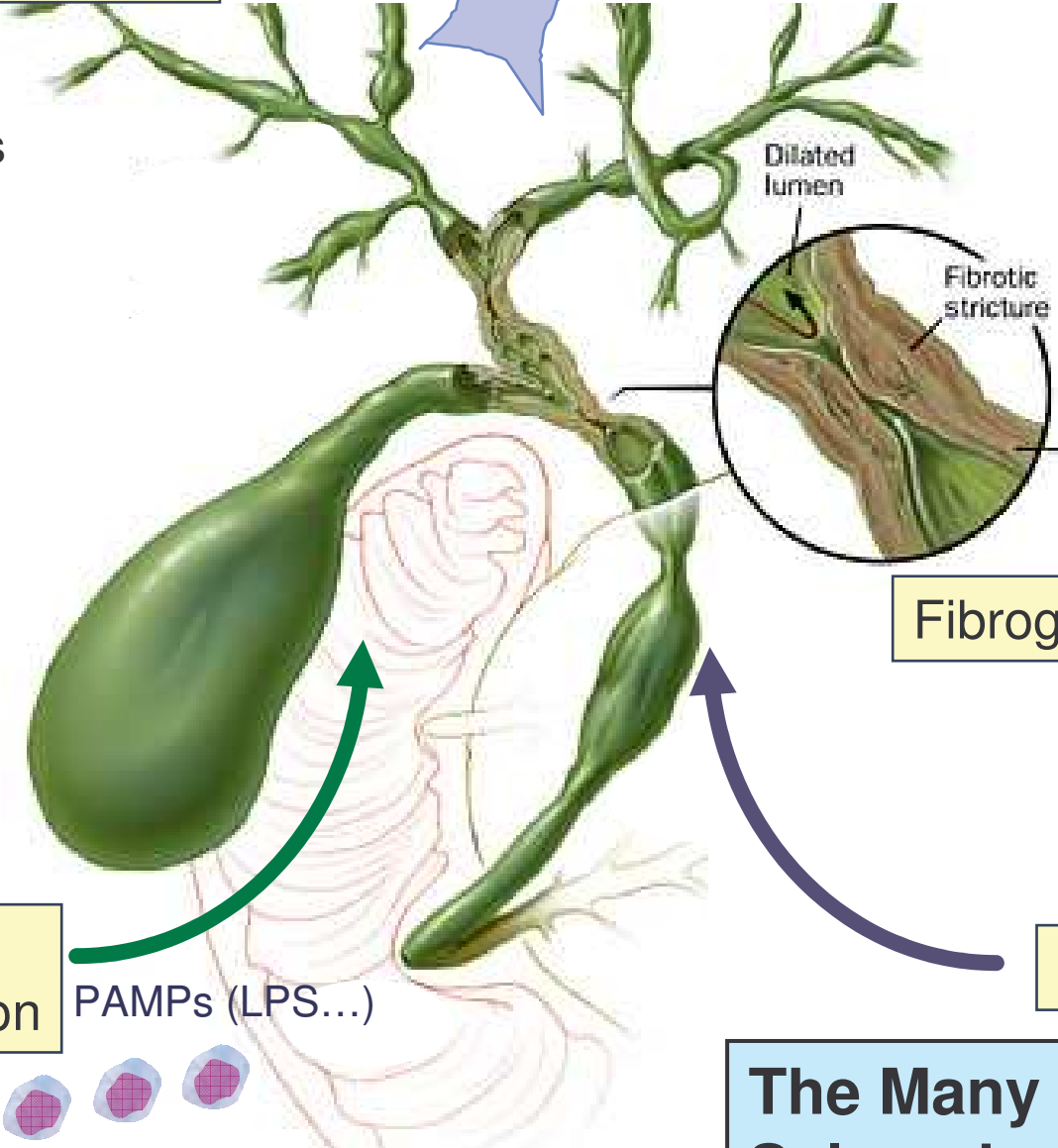
Atypical pANCA
CD4-pos. Infiltrates



Cytokines

Transport Defects

Toxic Bile



Fibrogenesis



Intestinal Translocation

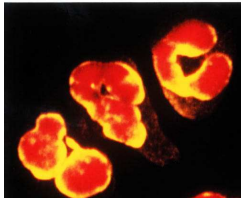
PAMPs (LPS...)

Ischemia



Gut-primed T cells

The Many Faces of Sclerosing Cholangitis



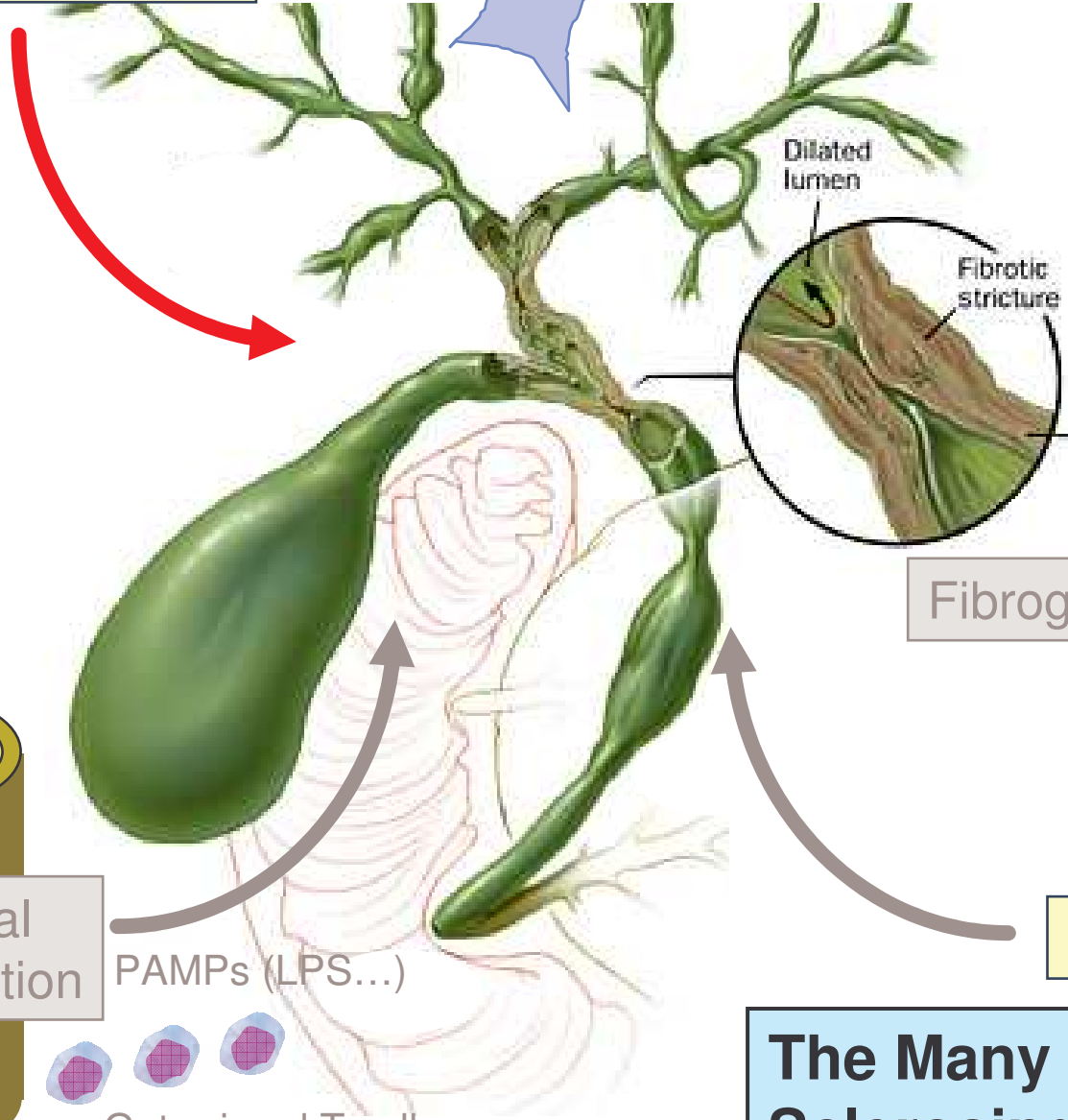
Immunological Mechanisms



Cytokines

Transport Defects

Toxic Bile



Intestinal Translocation

PAMPs (LPS...)

Ischemia

Fibrogenesis



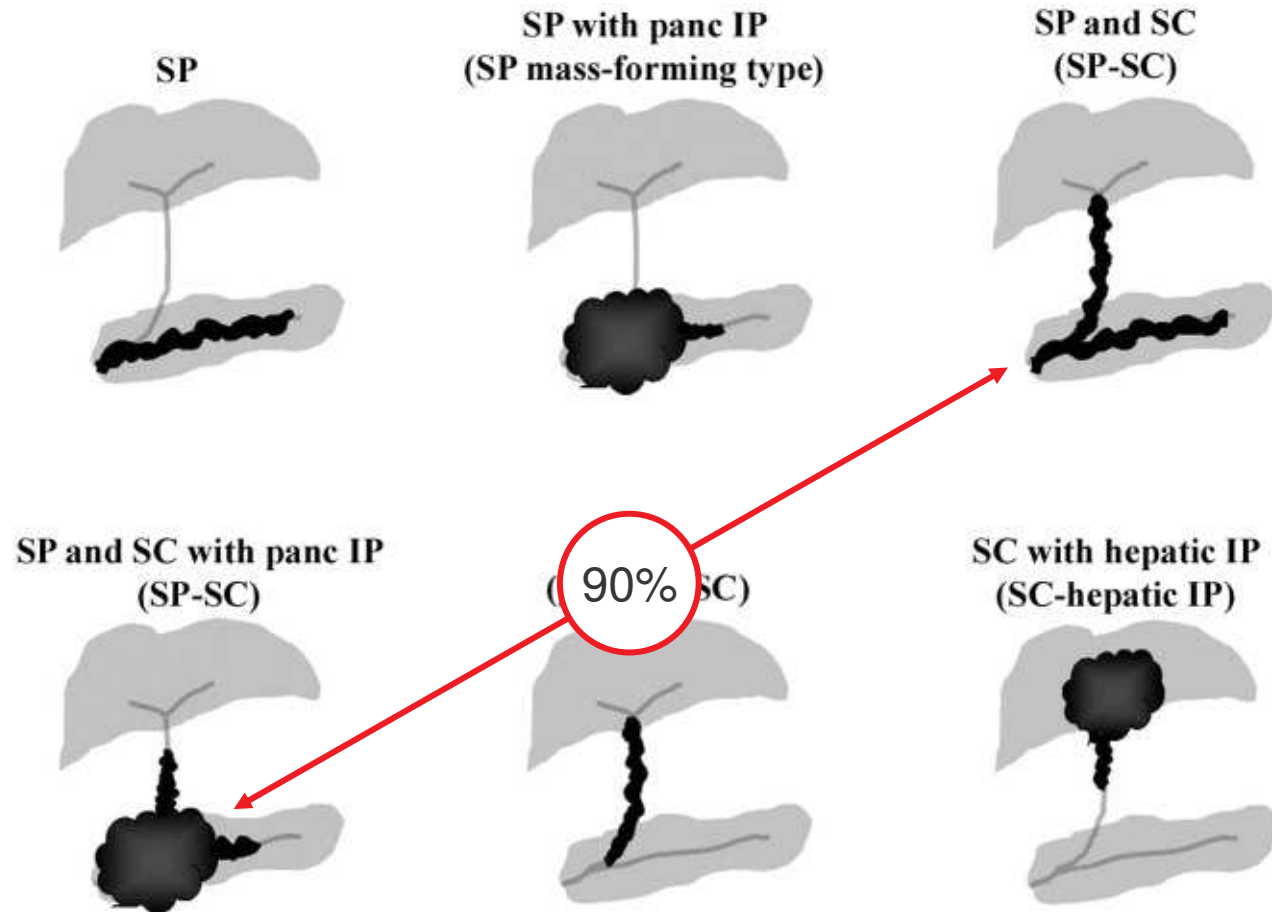
Gut-primed T cells

The Many Faces of Sclerosing Cholangitis

IgG4-associated Cholangitis

- Synonyms
 - Autoimmune pancreatitis-associated SC
 - Sclerosing pancreato-cholangitis
- Clinical presentation: 75% acute jaundice
- Diagnostic test
 - IgG4 > 140mg/dL, (plasma cells), imaging
 - DDx: 9% of PSC have elevated IgG4
- Therapy
 - Steroids, (azathioprine, MMF)

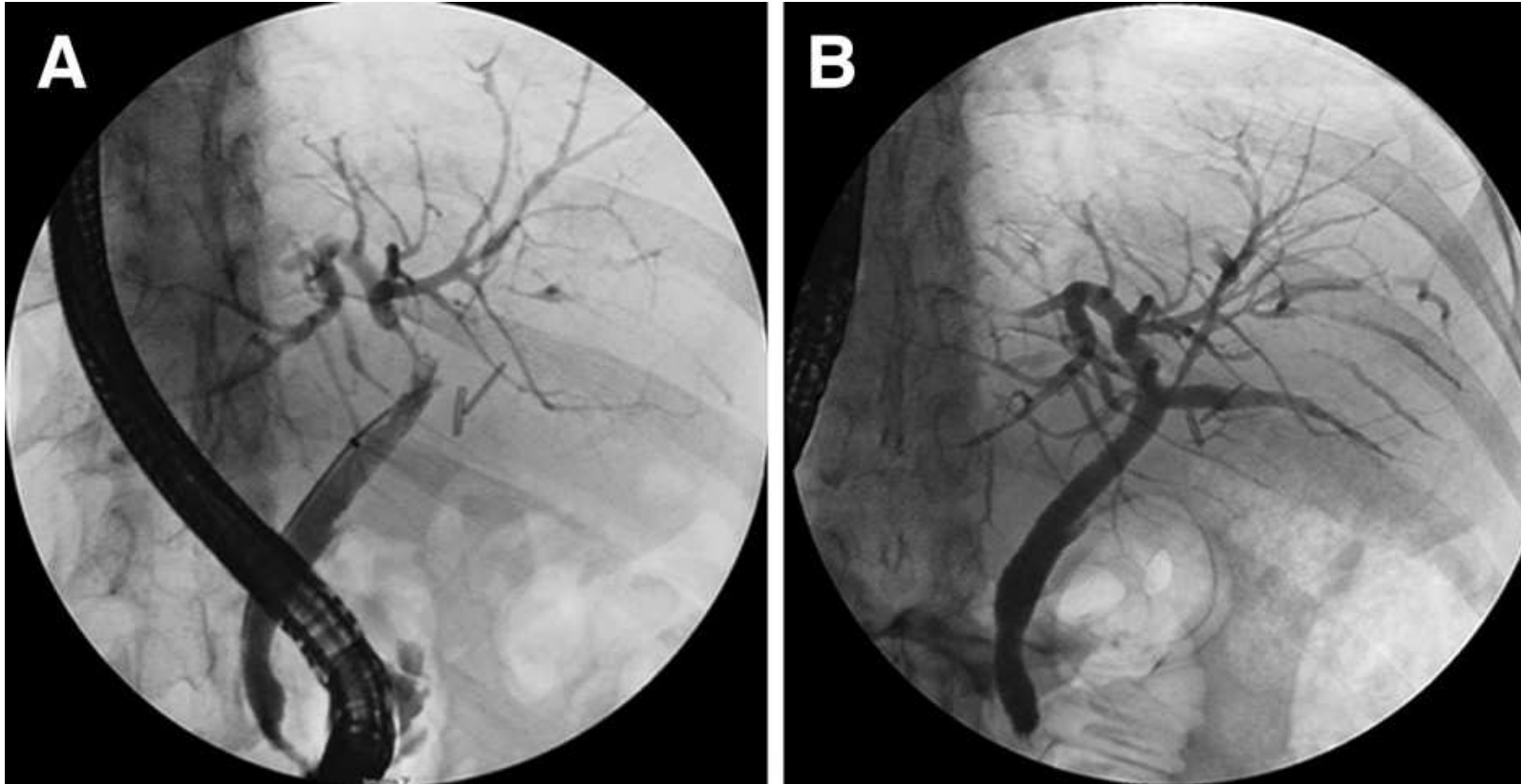
IgG4-associated Cholangitis



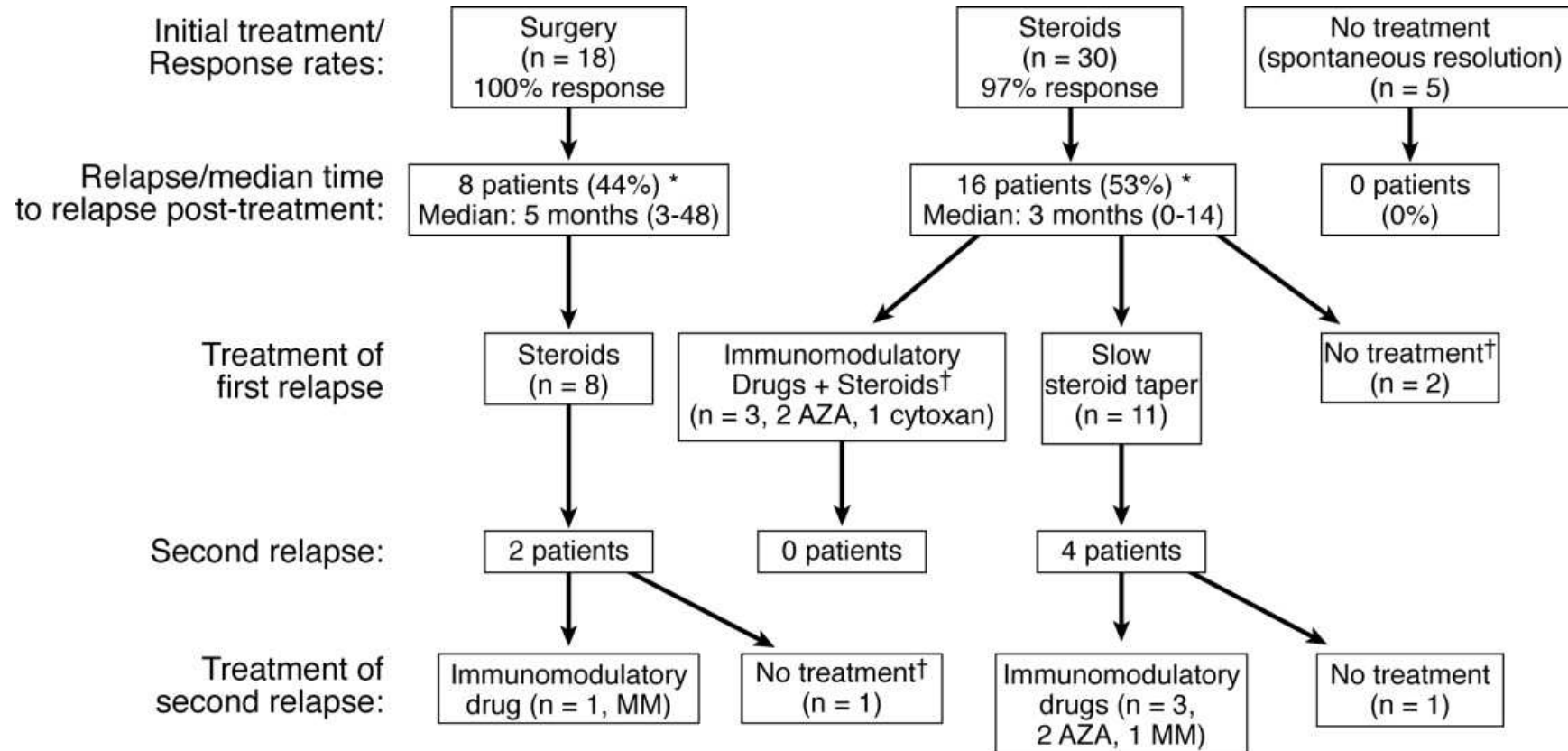
IgG4-associated Cholangitis

Before

After Steroids (3m)



IgG4-associated Cholangitis



IgG4-associated Cholangitis

Table 2. Pretreatment Laboratory and Serologic Data in 53 IAC Patients

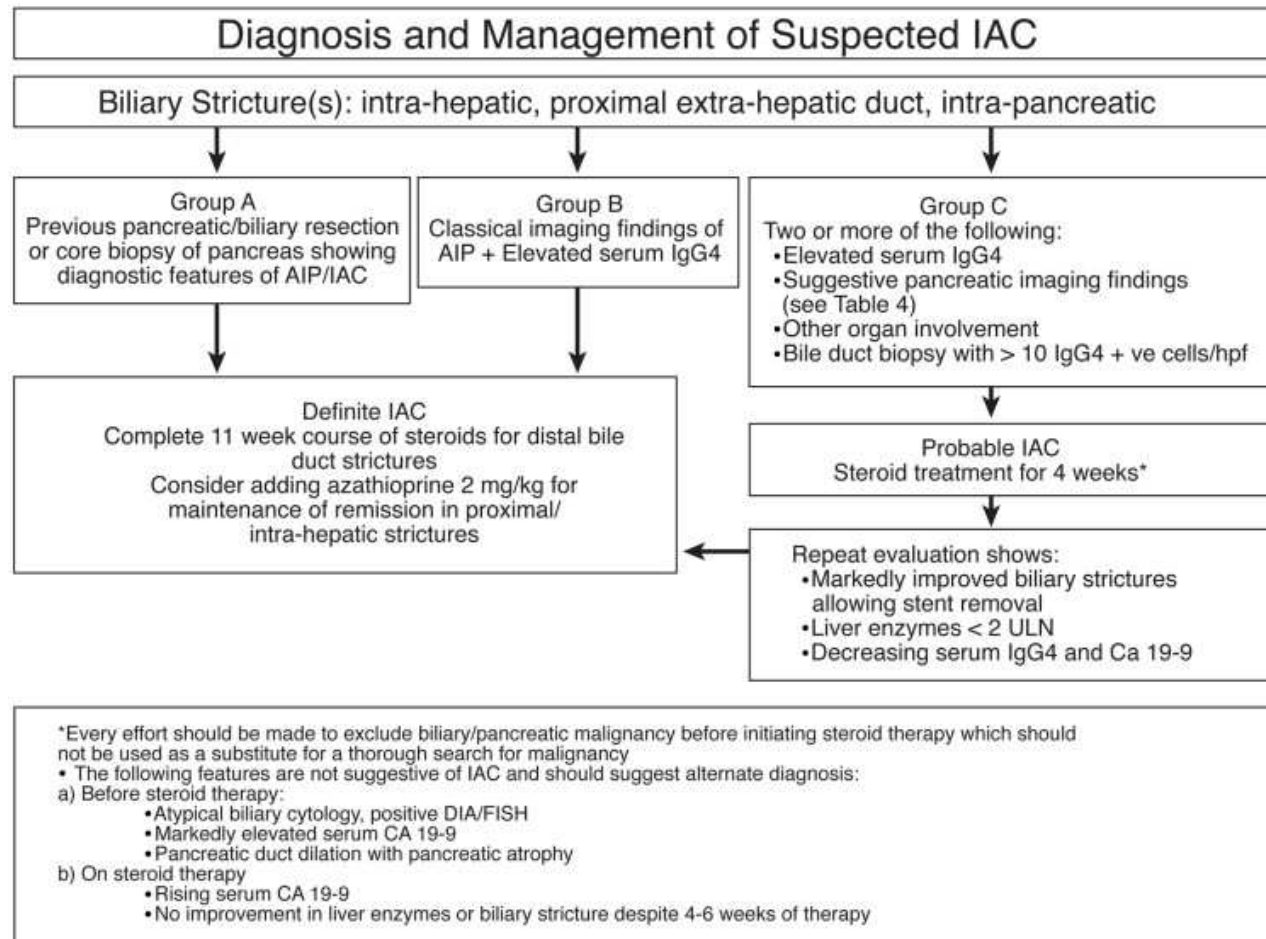
Laboratory tests	Mean ± SEM (range)	Normal range	Distribution
Serum IgG4 level, mg/dL	516 ± 98 (6–2490)	8–140	>140 mg/dL: 74% >280 mg/dL: 50%
CA 19-9 level, IU/mL	91 ± 30 (1–1005)	0–37	>37 IU/mL: 48% >100 IU/mL: 18%
Bilirubin level, mg/dL	7.5 ± 1 (0.3–19.1)	0.1–1.0	>5 mg/dL: 65%
Alkaline phosphatase level, U/L	512 ± 64 (77–1556)	45–115	>115 U/L: 84% >500 U/L: 34%
Alanine aminotransferase level, U/L	190 ± 64 (18–2445)	9–29	>100 U/L: 62%
Aspartate aminotransferase level, U/L	98 ± 17 (16–745)	12–31	>100 U/L: 32%

Table 3. Summary of Largest Published Series of Biliary Involvement in Patients With AIP

Study	Number of patients	Diagnostic criteria for AIP	Diagnostic biliary histology?	Mean age, y	Male (%)	Obstructive jaundice	% Increased serum IgG4 levels	Treatment response	IBD (%)	Location of strictures
Zen et al, ¹³ 2004 (Japan)	14	Histology	Yes	67	79%	NA	NA	No treatment (surgical specimens)	0	Common BD: 71% Extrahepatic BD: 50% Intrahepatic BD: 43%
Nakazawa et al, ¹⁴ 2004 (Japan)	26	JPS criteria	NA	62	62%	NA	NA	NA	0	Extrahepatic BD (including CBD): 100% Intrahepatic BD: 9/26; 35%
Takikawa et al, ¹⁵ 2004 (Japan)	28	JPS criteria	NA	NA	64%	41%	NA	Steroids: 100% response	0	Extrahepatic BD: 23/28; 82% Intrahepatic BD: 23/28; 82%
Nishino et al, ¹⁶ 2005 (Japan)	16	JPS criteria, histology in 7 patients	Yes, 4 patients	63	75%	50%	NA	Steroids: 100% response	NA	Intrapancreatic CBD: 13/16; 81% Extrahepatic BD: 5/16; 31% Intrahepatic BD: 2/16; 13%
Van Buuren et al, ¹⁷ 2006 (The Netherlands)	10	JPS criteria, histology in 3 patients	NA	52	100%	80%	NA	Steroids (5 patients): 100% response	0	Both intrahepatic and extrahepatic BD strictures present in all patients
Kamisawa et al, ¹⁸ 2006 (Japan)	23	JPS criteria, histology in 10 patients	NA	68	81%	87%	82% (data from scatterplot)	NA	NA	Intrapancreatic bile duct: 20/23; 87% Extrahepatic bile duct: 1/23; 4% Intrahepatic bile ducts: 2/23; 9%

BD, bile duct; JPS, Japanese Pancreas Society; NA, information not available from article.

IgG4-associated Cholangitis



Overlap-Syndromes

