

פגישת מוכנים 1.3.2016



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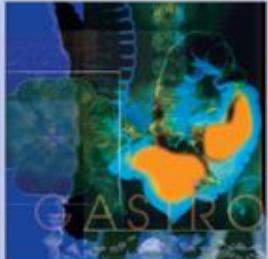


Sclerosing Cholangitis, Not only PSC

1.3.2016

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Case 1 – Z.G

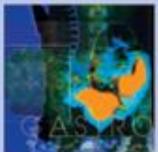
57 y/o male

Medical History- Wegener's Granulomatosis from 2014

ENT- Hemoptysis, sinusitis

RENAL- Hemodialysis

Treatment- steroids, cyclophosphamide, rituximab



Case 1 – Z.G

10/2015 ICU-pneumonia and severe septic shock:

- Mechanical ventilation
- Vasopressors
- Severe lactic acidosis
- CMV viremia



Lab results

- Bilirubin- 8 mg/dL
 - AST 450 U/L
 - ALT 580 U/L
 - ALKP 2900 U/L
 - GGT 4700 U/L
- HBsAb- pos
 - HBsAg, HbcAb- neg
 - HCV- neg
- ANA pos 1:160
 - AMA, ASMA, anti LKM- neg
 - IgG, IgM- normal limits



Differential Diagnosis ???

Ischemic hepatitis

DILI

amiodarone
antibiotics- vancomycin, ceftazidime
piperacillin/Tazobactam
meropenem, ceftriaxone
colliracin, resprim, azithromycin
gancyclovir
quetiapine

Infections

CMV, HBV, EBV
bacteria

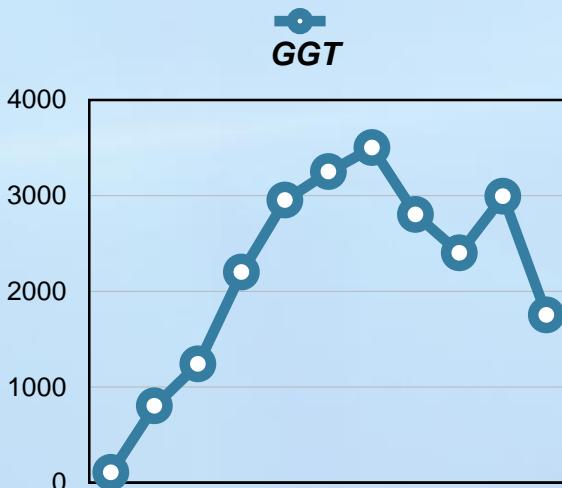
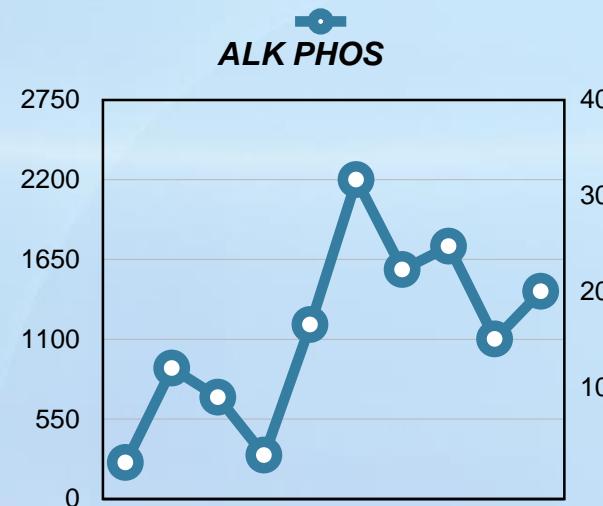
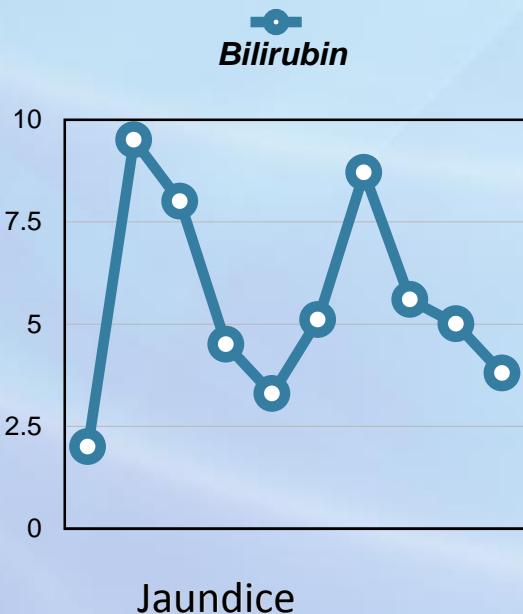
Granulomatous hepatitis

Autoimmune



3 months later...

No change in liver enzymes



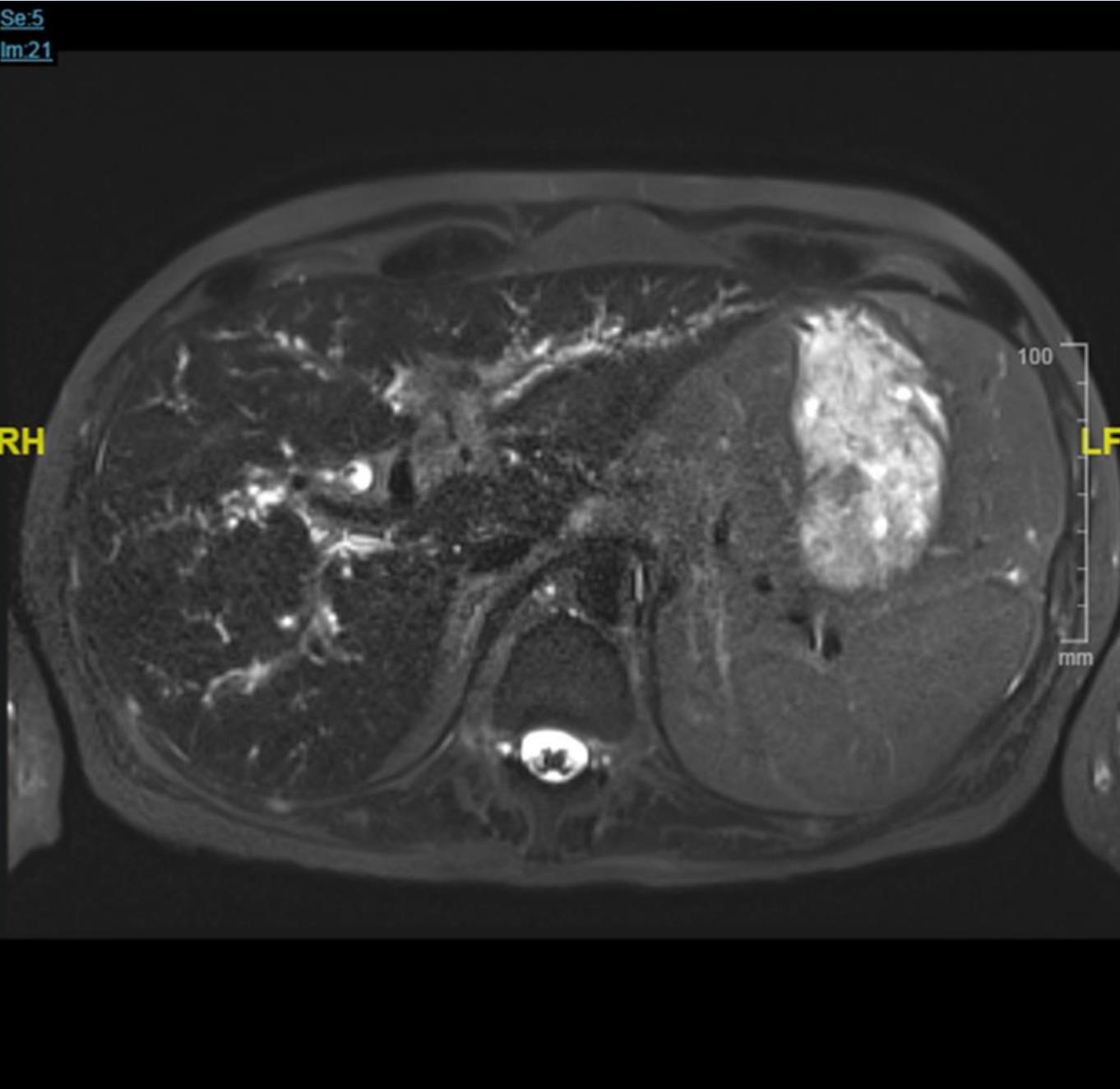
Jaundice

Encephalopathy, ammonia 280



MRCF

- Alteration in biliary flow
- Dilated bile duct
- Ductal obstruction
- CBD dilation
- Diffuse dilation of extrahepatic bile ducts
- Hepatolithiasis



Wegener's Granulomatosis

Severe shock

Elevated cholestatic enzymes

Strictures and dilatations of bile ducts



Case 2- K.B

46 y/o male

Medical History- gout

Since 2013 elevated liver enzymes



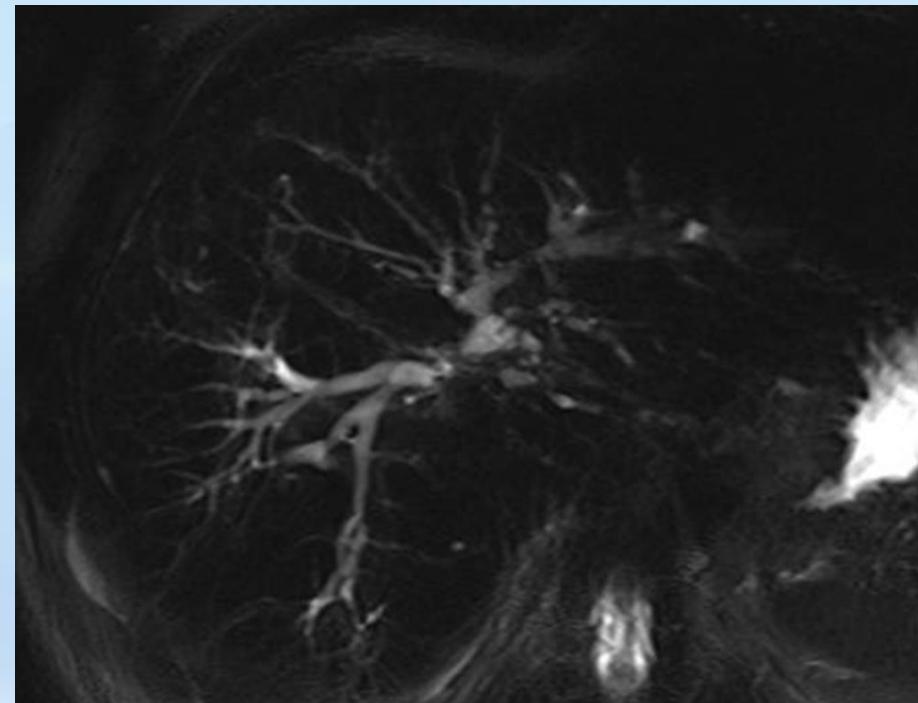
Lab results

- Bilirubin- normal
 - AST 50 U/L
 - ALT 45 U/L
 - ALKP 250 U/L
 - GGT 150 U/L
- HBsAb- neg
 - HBsAg, HbcAb- neg
 - HCV- neg
- ANA, AMA, ASMA, ANCA anti LKM- neg
 - IgG, IgM- normal limits
 - Celiac- neg



MRI/MRCP

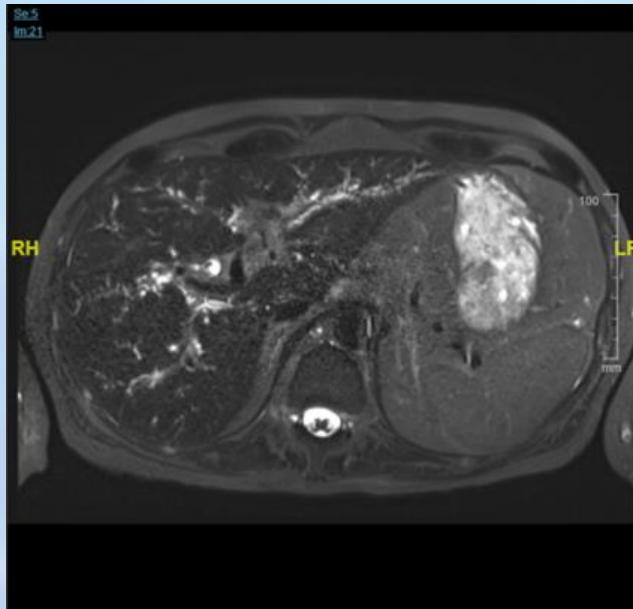
- Alternating strictures and dilatations of intra and extrahepatic bile ducts no dominant stricture
- Spleen 17 cm
- Varices in upper left quadrant
- Portal vein thrombosis with cavernous transformation



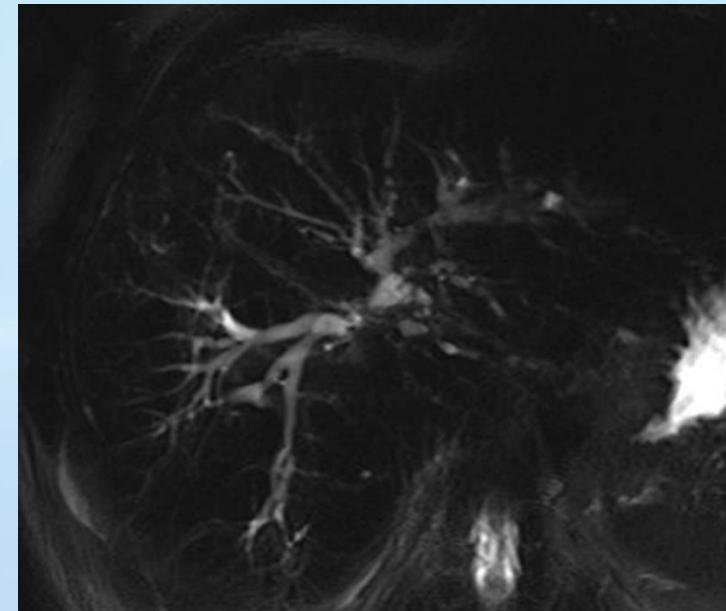
Abdominal US



Case 1



Case 2



SCLEROSING CHOLANGITIS

Primary sclerosing
cholangitis

Secondary sclerosing
cholangitis

same pathological and clinical manifestations

Different Etiology



Secondary sclerosing cholangitis

HIV negative

AIDS cholangiopathy

Cholangiocarcinoma

Choledocholithiasis

Diffused sediment in the bile ducts

Diffuse intrahepatic metastasis

Eosinophilic cholangitis

Hepatic inflammatory pseudotumor

Histiocytosis X

IgG4-associated cholangitis

IgG4 in normal range

Intra-arterial chemotherapy

Ischemic cholangitis

US doppler was normal, no evidence of hepatic artery stenosis

Mast cell cholangiopathy

Portal hypertensive biliopathy

Recurrent pancreatitis

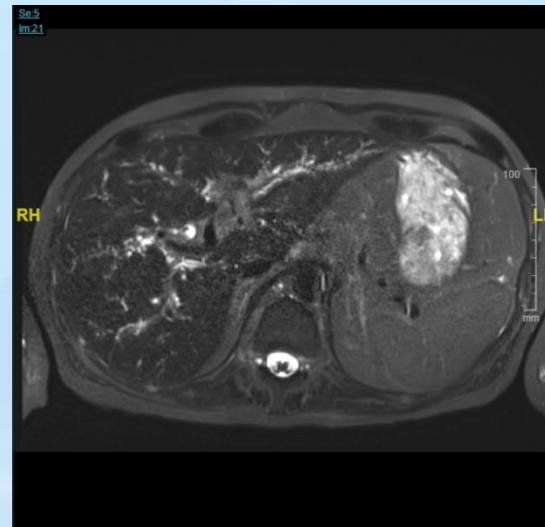
Recurrent pyogenic cholangitis

Surgical biliary trauma

Chapman R, Fevery J, Kalloo A, Nagorney DM, Boberg KM, Shneider B, Gores GJ; American Association for the Study of Liver Diseases. **Diagnosis and management of primary sclerosing cholangitis.** Hepatology. 2010



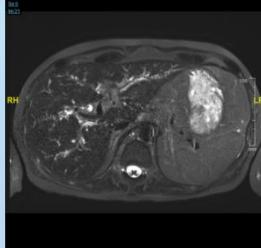
Case 1



What is the likely diagnosis ?

***Sclerosing Cholangitis
in critically ill patients***



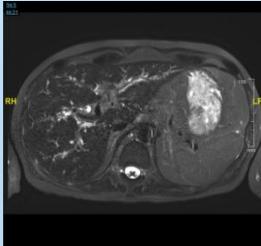


Sclerosing Cholangitis in critically ill patients

- Rare cholestatic liver disease
- Develops in patients with:
 - sepsis
 - acute respiratory distress syndrome
 - during a long-term intensive care unit (ICU) treatment
- Rapid progression to liver cirrhosis and hepatic failure.

Kirchner GI, Rümmele P. Update on Sclerosing Cholangitis in Critically Ill Patients. Viszeralmedizin. 2015 Jun;31





Diagnosis SC-CIP

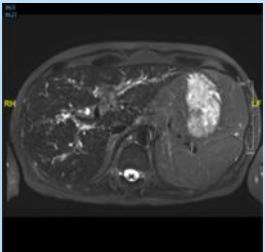
Labs

increased bilirubin
and cholestatic
liver enzymes
mainly GGT

Imaging

- biliary casts
- progressive destruction of intrahepatic bile ducts
- pruned tree





Pathomechanism

Arterial hypoperfusion

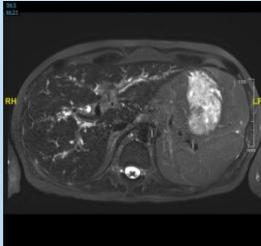
Ischemic injury to the bile duct epithelium

Inflammation of bile ducts

Cast formation

stenosis of biliary ducts

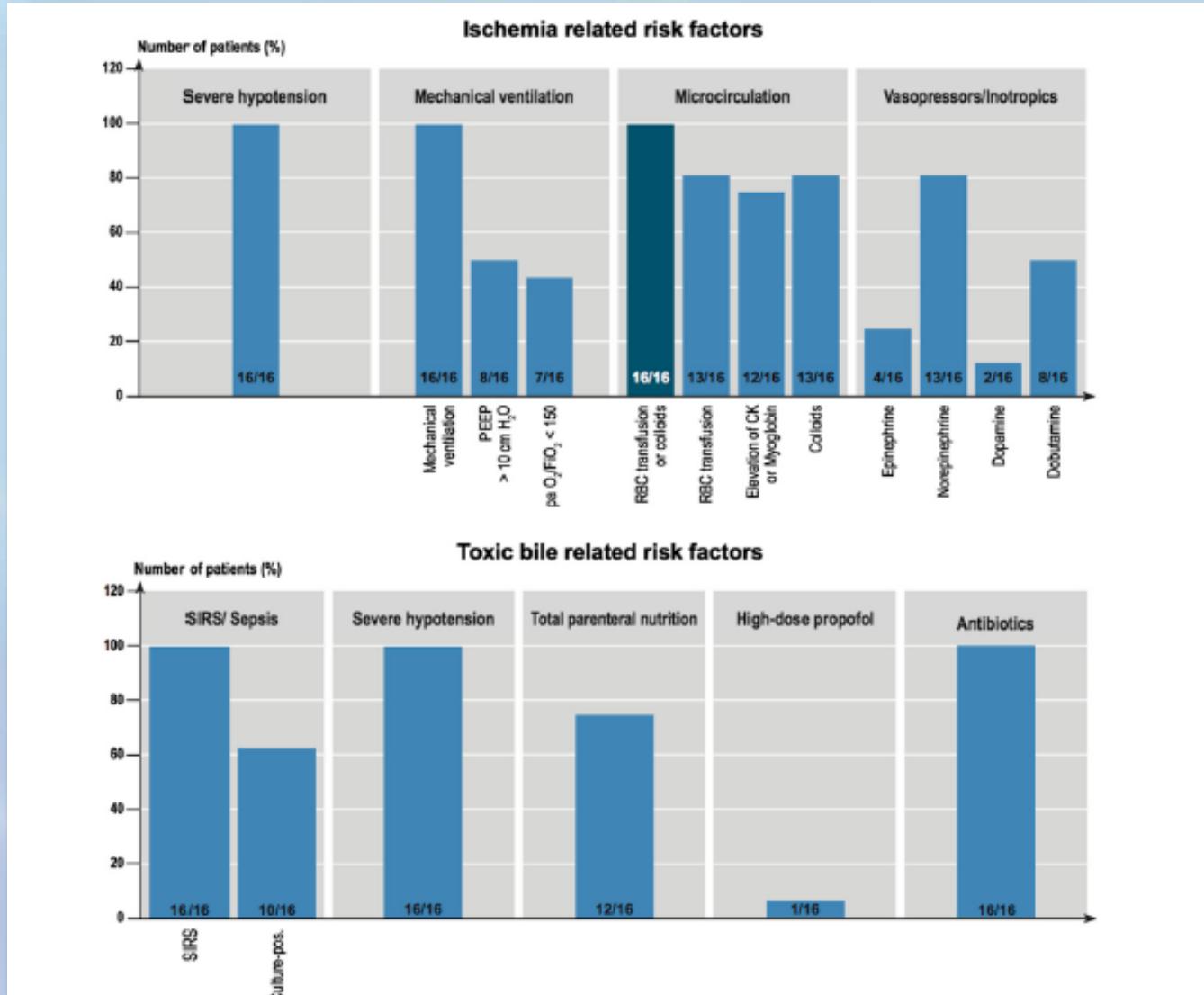




Risk factors SS-CIP

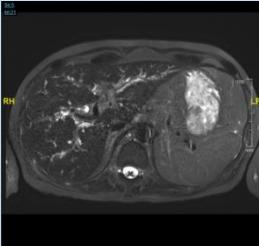
- Mechanical ventilation with high positive end-expiratory pressure
- High dose of vasoconstrictors
 \Rightarrow ***Disturbance to the hepatosplanchnic hemodynamics***





Leonhardt S, Veltzke-Schlieker W, Adler A, Schott E, Hetzer R, Schaffartzik W, Tryba M, Neuhaus P, Seehofer D. Trigger mechanisms of secondary sclerosing cholangitis in critically ill patients. *Crit Care*. 2015 Mar 31;19:131.





Treatment and Prognosis

- Poor prognosis
- Liver transplantation -the only curative treatment option
- Median survival - 13 months without liver transplantation

Gelbmann CM, Rümmele P, Wimmer M, Hofstädter F, Göhlmann B, Endlicher E, Kullmann F, Langgärtner J, Schölmerich J. **Ischemic-like cholangiopathy with secondary sclerosing cholangitis in critically ill patients.** Am J Gastroenterol. 2007 Jun;102(6):1221-9



Case 2



What is the likely diagnosis ?

Portal hypertensive biliopathy



Portal Biliopathy

Portal vein thrombosis

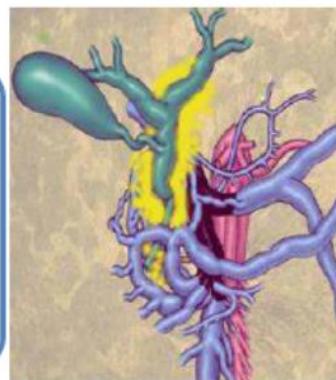
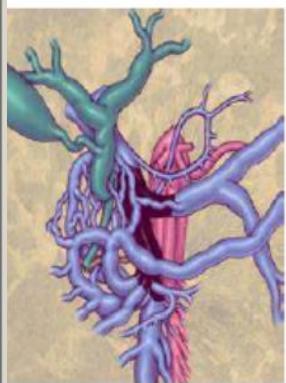
Obstruction of biliary tract

portal
cavernoma

Portal hypertension

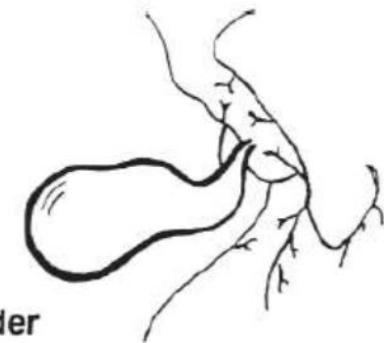
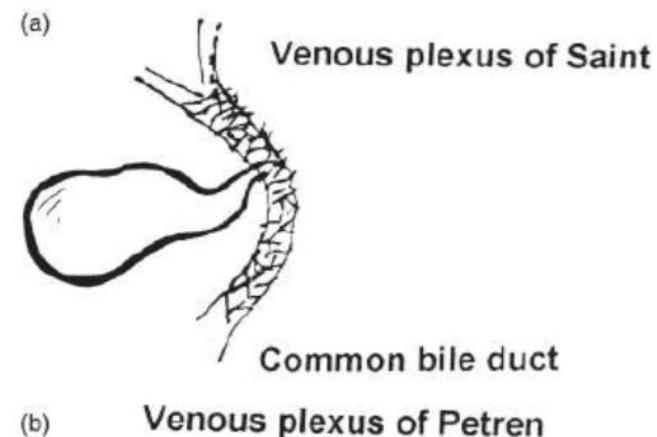


Pathogenesis



External pressure of portal cavernoma

Ischemia secondary to extension of the thrombotic process to small venules of the bile ducts



Two venous plexuses of bile ducts and gallbladder:

- Epicholedochal venous plexus of Saint
- Paracholedochal veins of Petren



Clinical manifestations PB

Most (60-70%) asymptomatic

Symptomatic-

- obstructive jaundice
 - cholangitis
 - biliary pain due to bile duct stones
 - variceal bleeding
 - ascites

Bile stasis

Portal hypertension

Varma V, Behera A, Kaman L, Chattopadhyay S, Nundy S. **Surgical management of portal cavernoma cholangiopathy**. J Clin Exp Hepatol. 2014 Feb;4



Diagnosis PB

Labs

increased
cholestatic liver
enzymes, mainly
ALK PHOS

Imaging

Abdominal US/CT

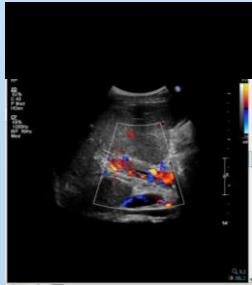
- extrahepatic portal vein obstruction
- portal cavernoma
- signs of portal hypertension

Imaging

MRCP

- strictures and dilatations
- filling defects



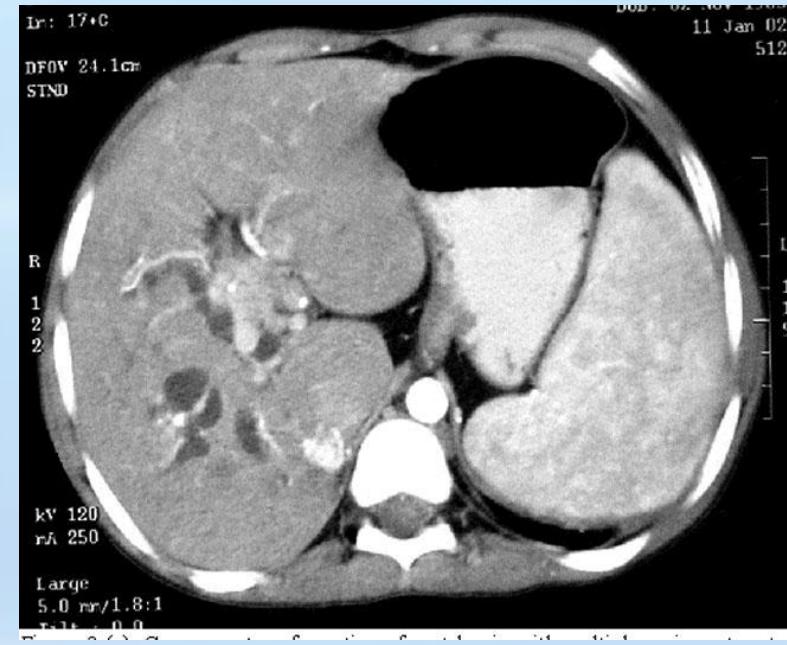


Diagnosis PB

Cavernous transformation



**Cavernous transformation and
intrahepatic biliary duct
dilatation**





Management PB

Endoscopic- ERCP

Surgical

sphincterotomy

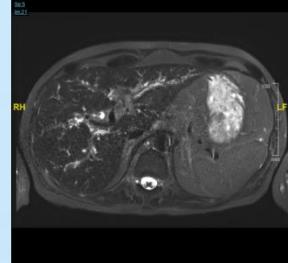
stenting

biliary bypass

liver
transplantation

portosystemic shunt
surgery





Conclusion

- Rare etiologies for secondary sclerosing cholangitis
- High index of suspicion is required for the diagnosis
- Importance of diagnosis for appropriate treatment

