

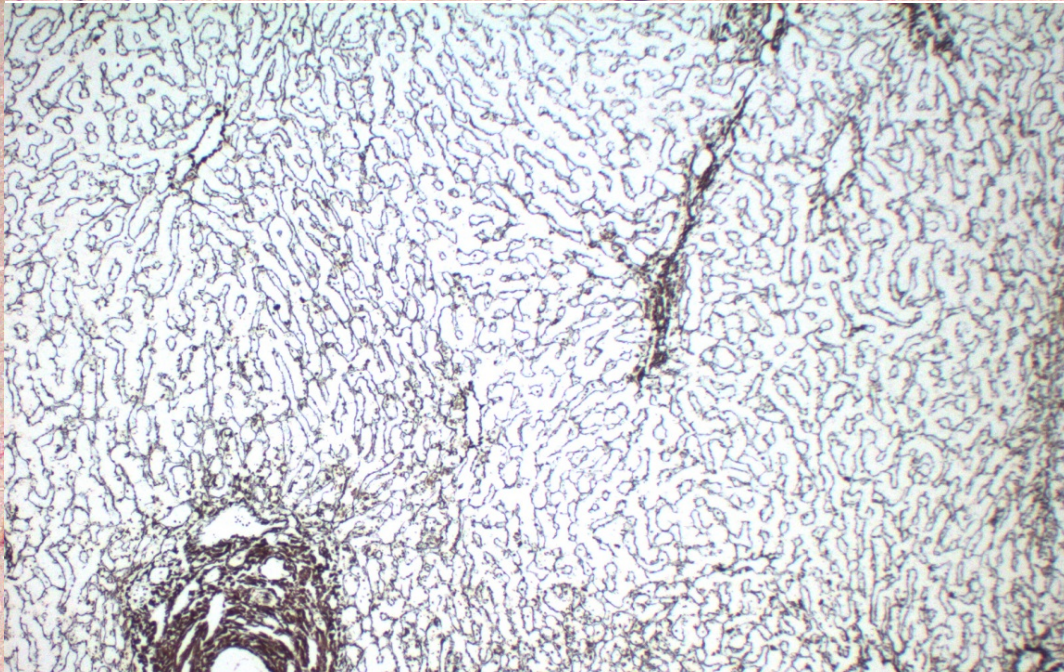
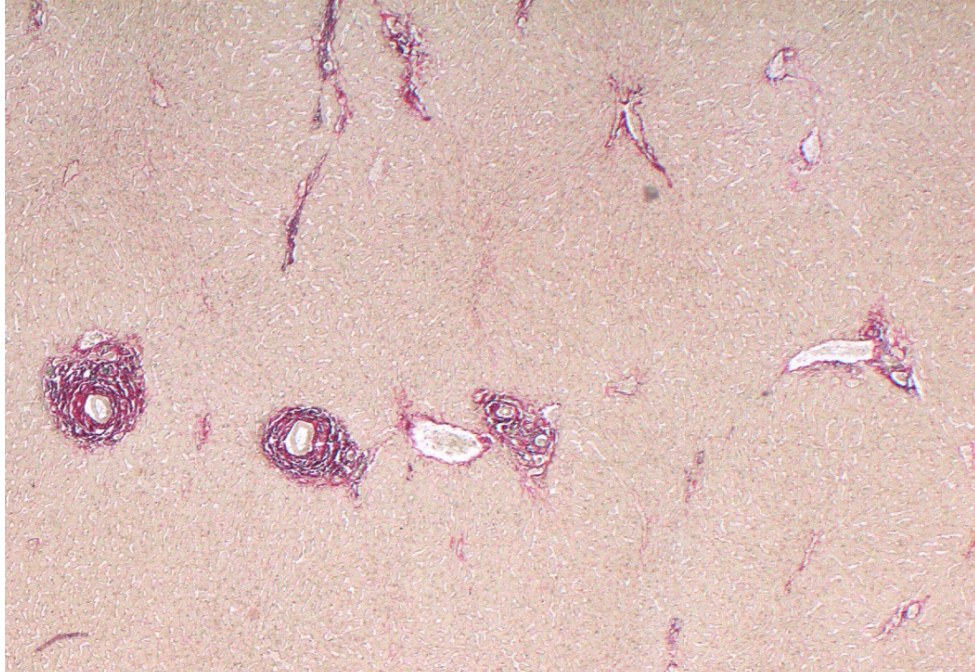
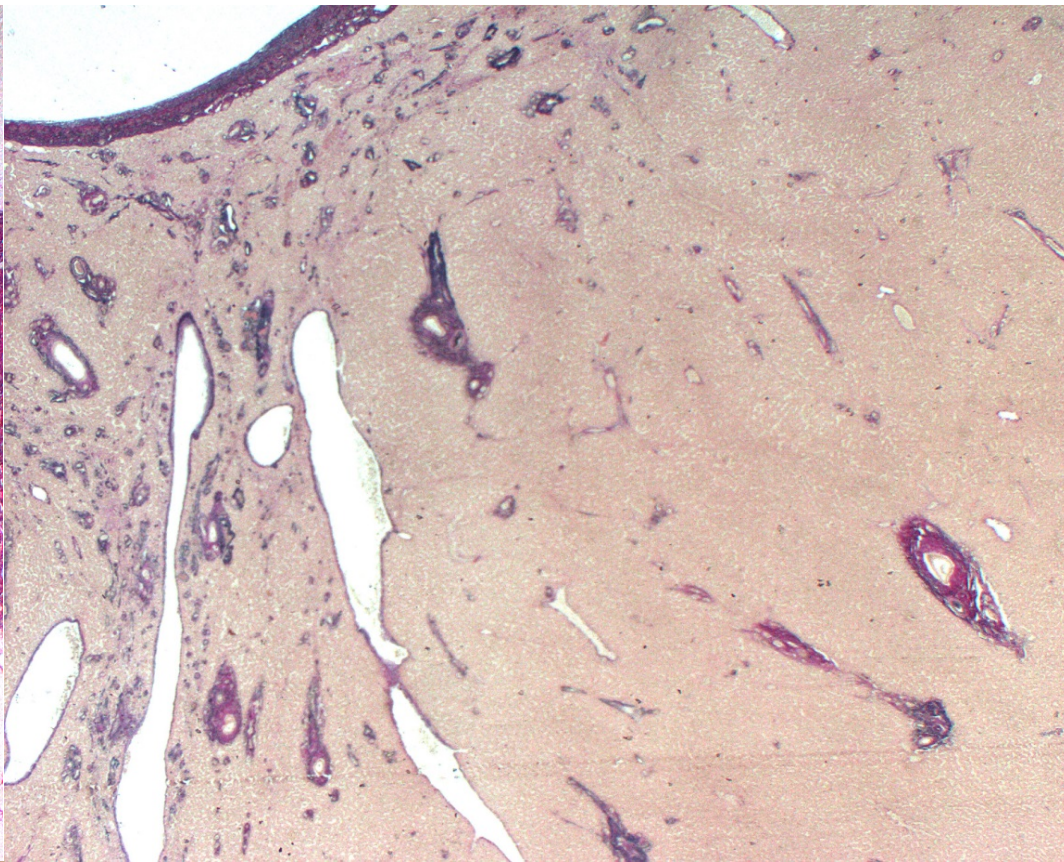
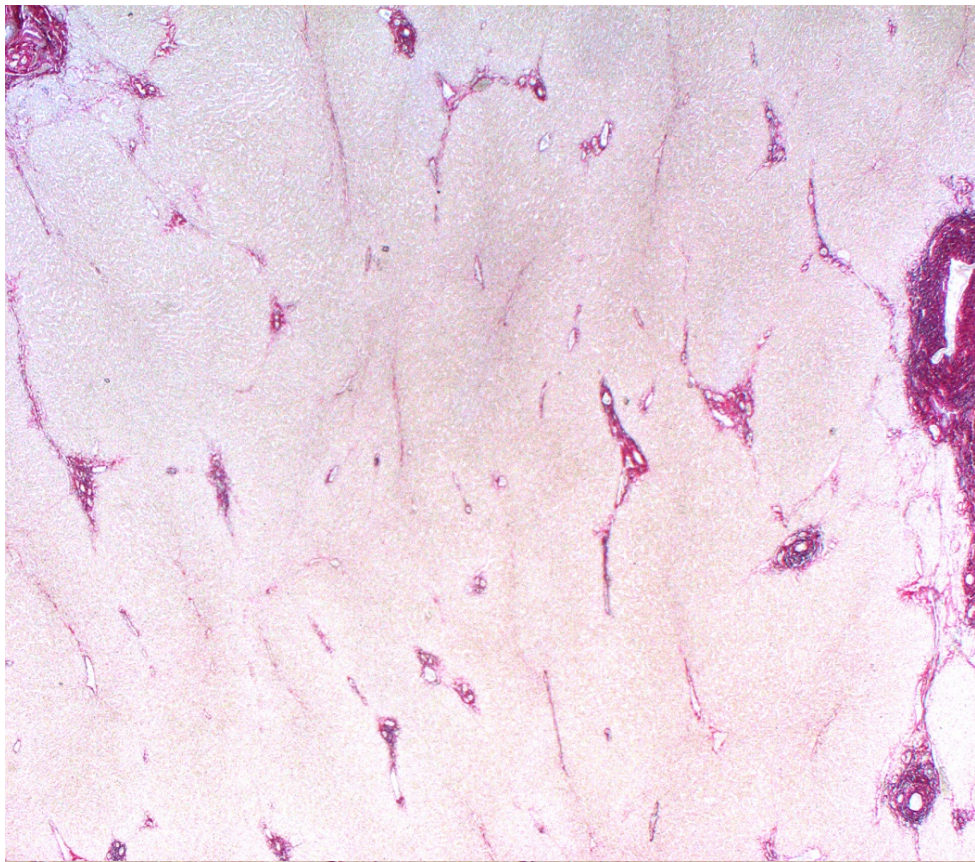
Non-cirrhotic Portal Hypertension

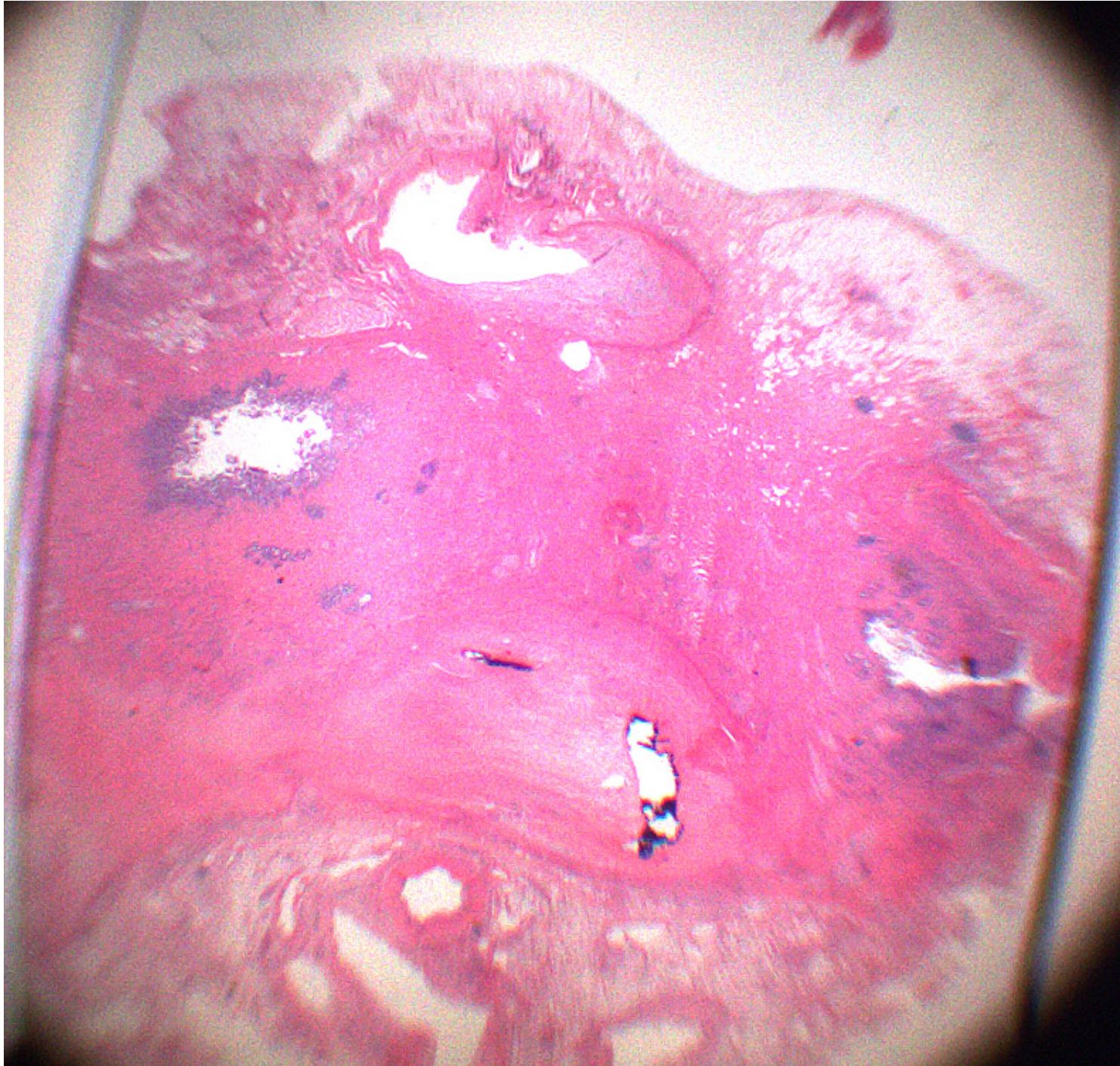
Liver EQA, Circulation LU, Case 7
Winter 2019

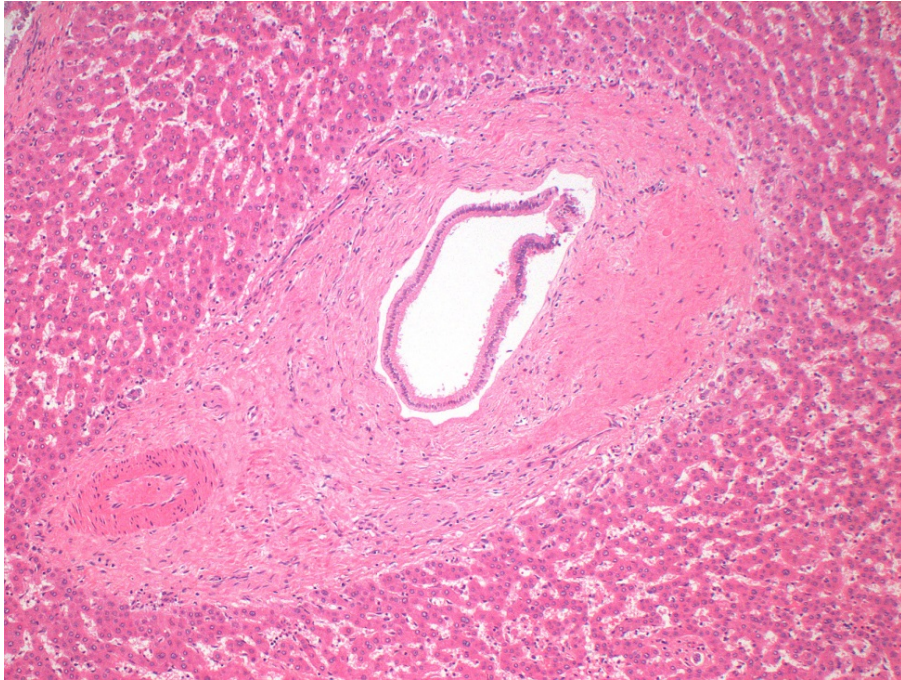
Case LU7 follow-up

- Transplanted 1 year later – also stomach and small bowel transplanted
- 1200g liver – coarsely lobulated
- Septa prominent, occasional areas of fibrosis
- Initial presentation age 4yr with PVT









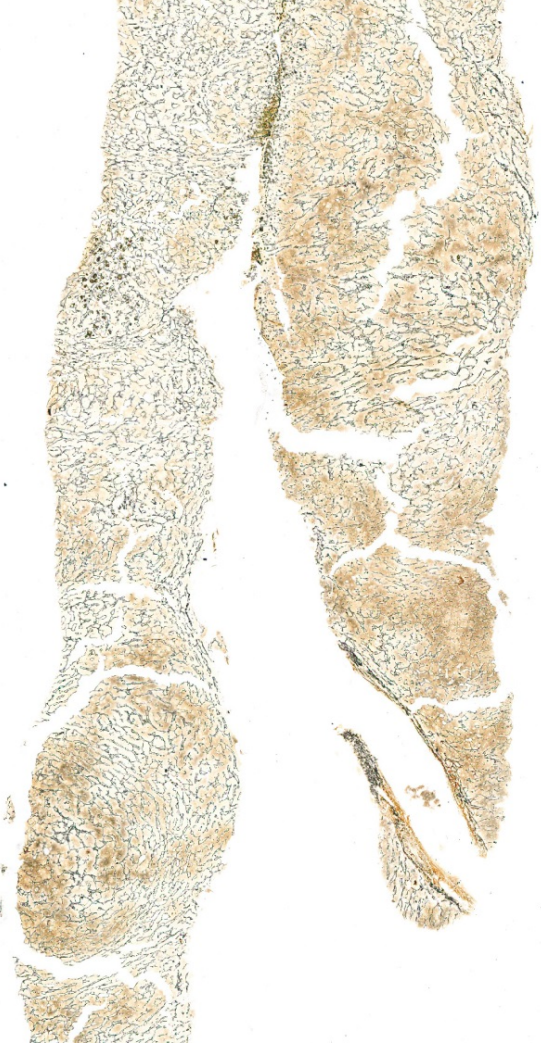
Non-Cirrhotic Portal Hypertension

- 2nd to cirrhosis in causing PH in West
- Heterogenous group of vascular disorders – self-evident features.
 - Exclude cirrhosis, malignant infiltrate, sarcoidosis, schistosomiasis, congenital hepatic fibrosis
- Nodular Regenerative Hyperplasia relatively common (>25%); due drugs, haematological, autoimmune, neoplastic or immunodeficiency; pre-sinusoidal or perisinusoidal portal hypertension (HVPG normal or mild increase).

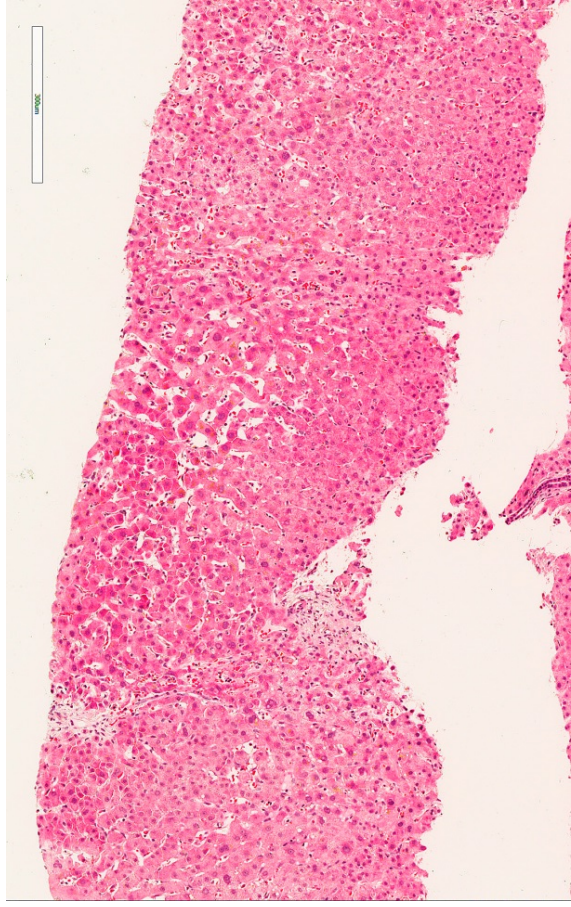
Idiopathic Non-Cirrhotic Portal Hypertension (INCPH)

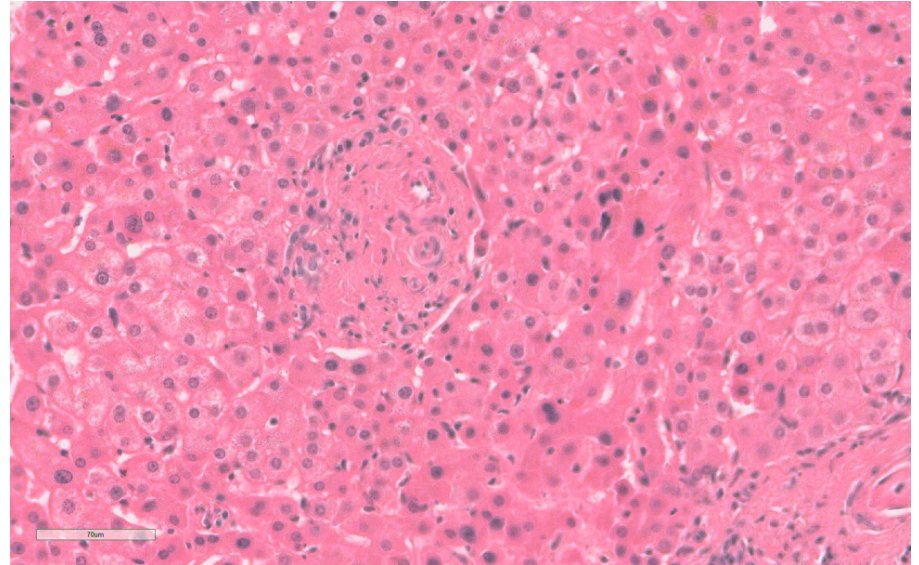
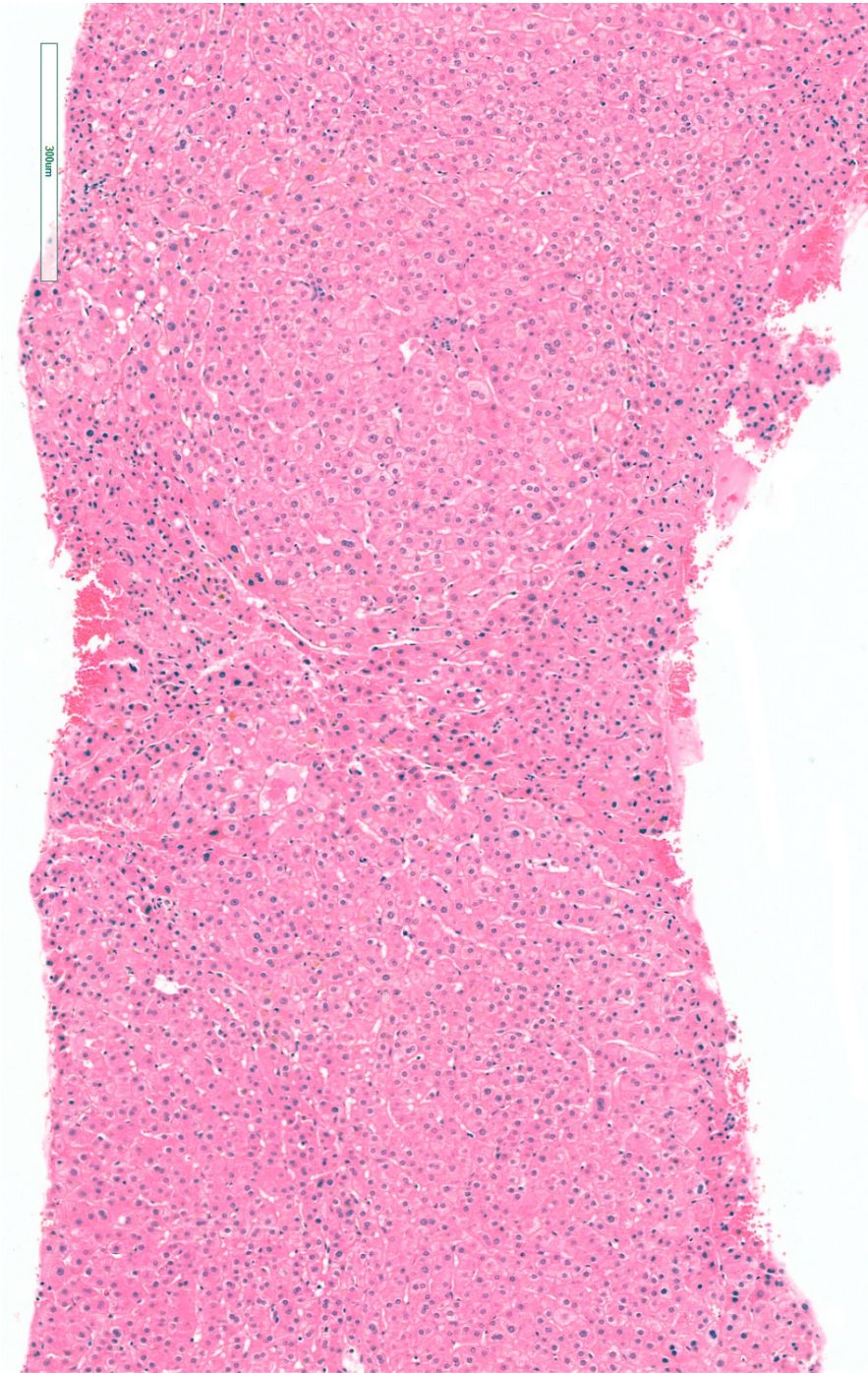
- Confusing and Confusion!
 - Multiple terms; *hepatoportal sclerosis, Banti syndrome, Incomplete septal cirrhosis, non-cirrhotic portal fibrosis, obliterative portal venopathy, nodular regenerative hyperplasia*
 - Not all features always present in any one biopsy
 - Pathophysiology poorly understood
 - Range of features, several **SUBTLE**
 - Could be thought to be cryptogenic cirrhosis
 - Incidence unknown, India & eastern common, related to infection; rarer in West, pro-thrombotic states
 - Children often extra-hepatic portal venous obstruction

50µm



50µm

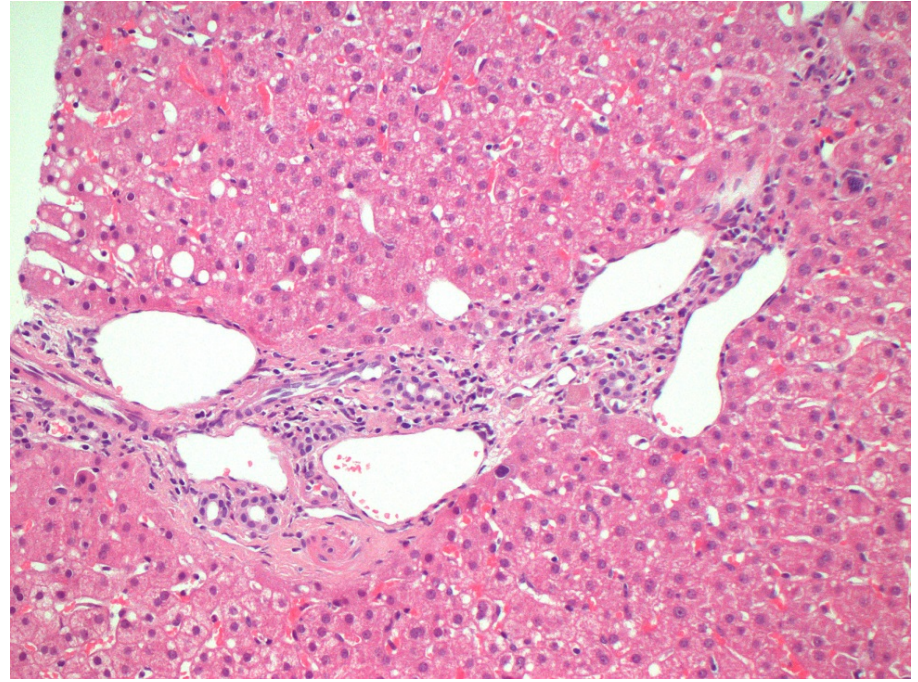
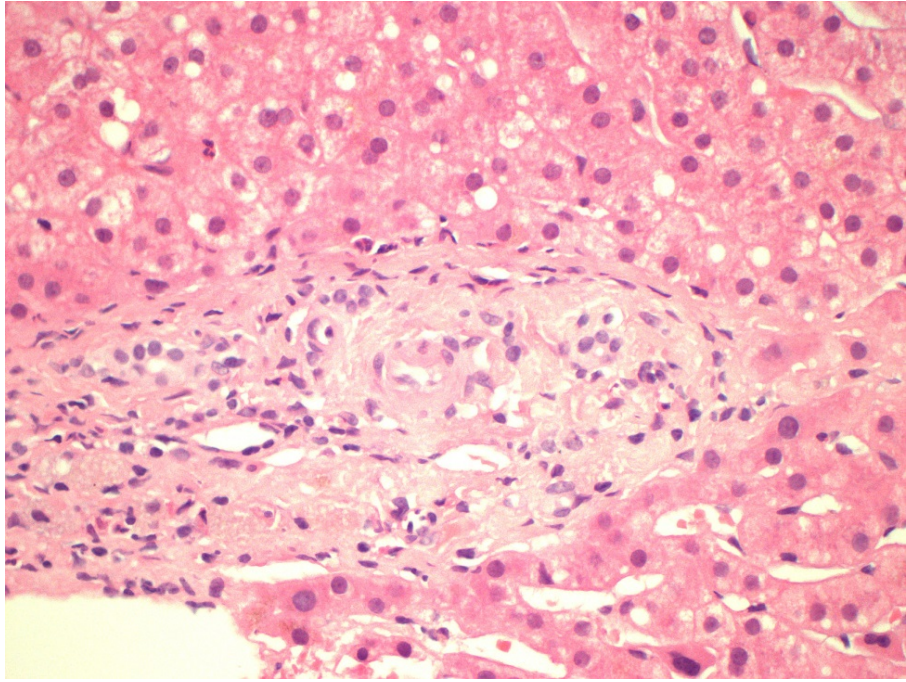


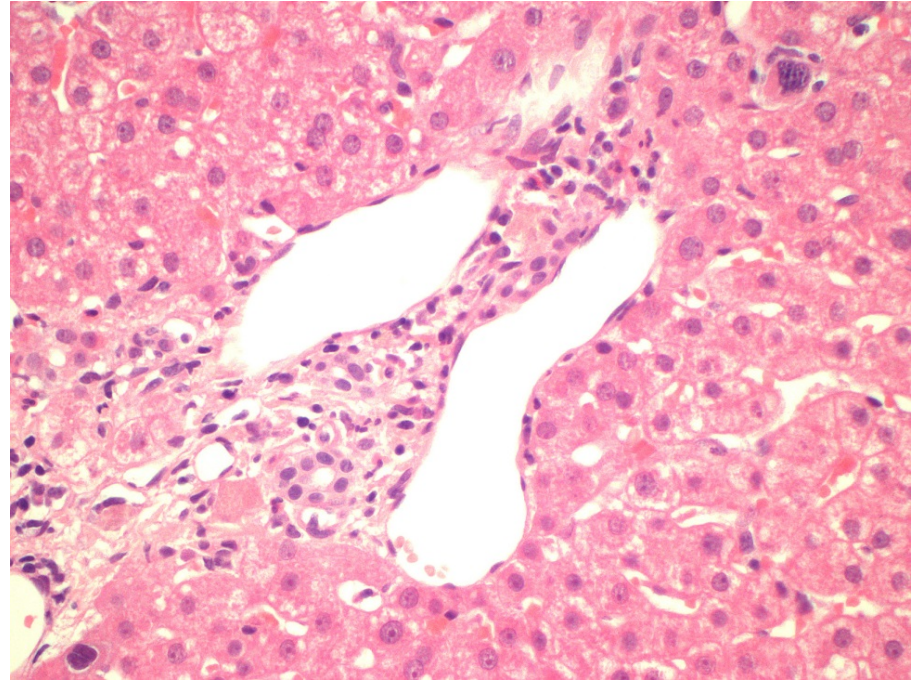
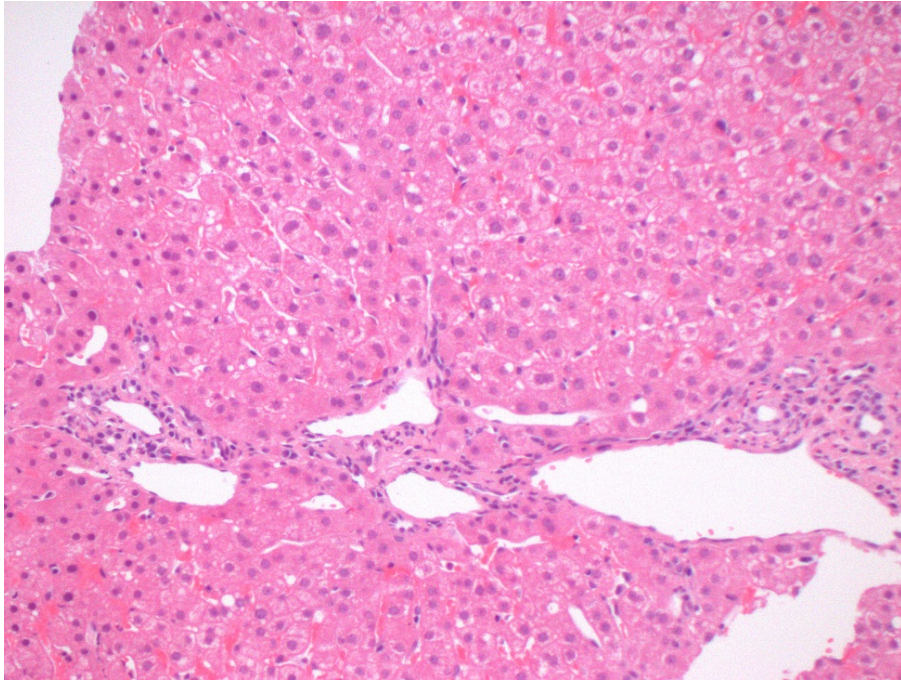


Portal Vein changes in INCPH

- Portal vein stenosis – complete or incomplete obliteration +/- thickening of the wall
- Herniated portal vein – vein from PT directly abutting periportal parenchyma
- Hypervascularised PT – multiple thin –walled vascular spaces
- Periportal abnormal vessels – single/multiple, different calibre close to PT

Guido M, Histopathology 2018

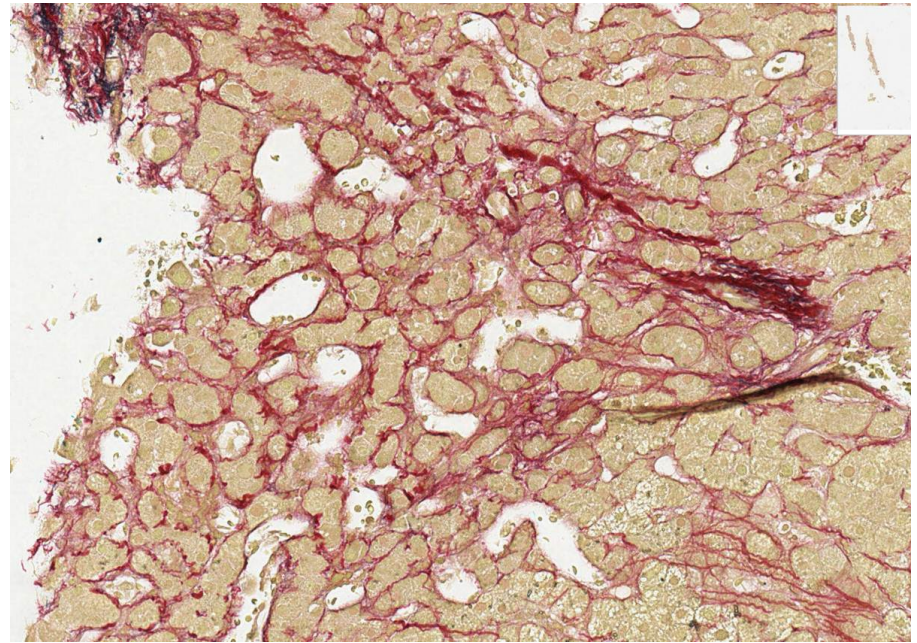
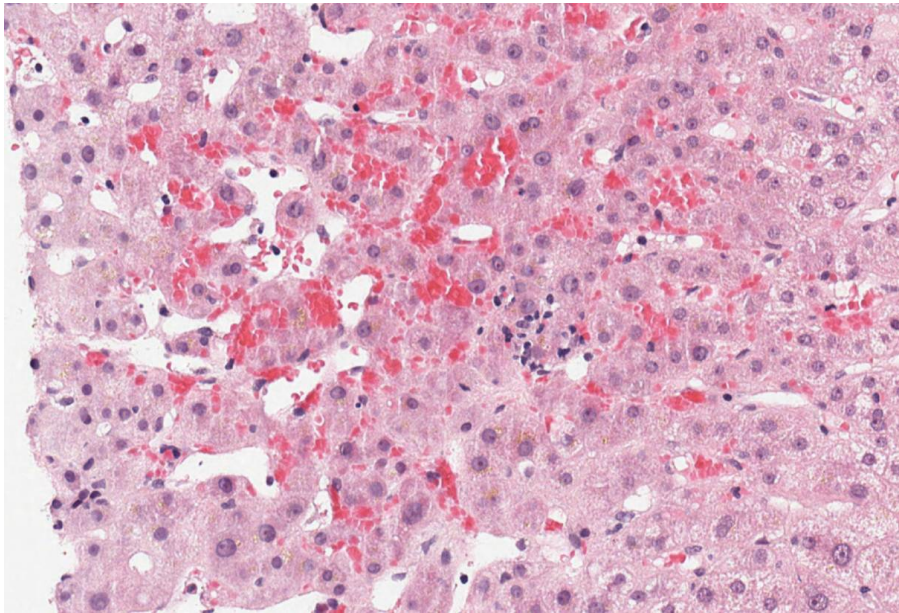




INCPH

- May be subtle
- Sinusoidal orientation, vague nodularity and dilatation are clue to a vascular aetiology

Outflow Obstruction



Causes of NCPH

Khanna,
J Hepatol 2014

Pre-hepatic		
FHVP normal, RAP normal, WHVP normal, HVPG normal, PVP high, ISP high		
Extrahepatic portal vein obstruction (EHPVO)		
Portal vein thrombosis		
Splenic vein thrombosis		
Splanchnic arteriovenous fistula		
Massive splenomegaly		
Infiltrative diseases-Lymphoma, myeloproliferative disorders		
Storage diseases-Gaucher's disease		
Hepatic		
FHVP normal, RAP normal, WHVP high, HVPG normal or high, PVP high, ISP high*		
Pre-sinusoidal	Sinusoidal	Post-sinusoidal
Developmental abnormalities	Sinusoidal fibrosis	Venoocclusive disease
Adult polycystic disease	Alcoholic hepatitis	Hepatic irradiation
Hereditary hemorrhagic disease	Drugs (methotrexate, amiodarone)	Toxins-Pyrrolizidine alkaloids
Arteriovenous fistulas	Toxins (vinyl chloride, copper)	Drugs-Gemtuzumab, ozogamicin, actinomycin D, dacarbazine, cytosine arabinoside, mithramycin, 6-thioguanine, azathioprine, busulfan plus cyclophosphamide
Congenital hepatic fibrosis	Metabolic (NASH, Gaucher's disease)	
Biliary diseases	Inflammatory (viral hepatitis, Q fever, healed cytomegalovirus, secondary syphilis)	
Primary biliary cirrhosis		
Sclerosing cholangitis		
Autoimmune cholangiopathy	Sinusoidal collapse	Phleboscclerosis of hepatic veins
Toxic-Vinyl chloride	Acute necro-inflammatory diseases	Alcoholic liver disease
Neoplastic occlusion of portal vein	Sinusoidal defenestration	Chronic radiation injury
Lymphoma	Alcoholic liver disease (early phase)	Hypervitaminosis A
Epithelioid hemangioendothelioma	Sinusoidal infiltration	E-ferol injury
Epithelial malignancies	Mastocytosis	Primary vascular malignancies
Chronic lymphocytic leukemia	Agnogenic myeloid metaplasia	Epithelioid hemangioendothelioma
Granulomatous lesions	Gaucher's disease	Angiosarcoma
Schistosomiasis	Amyloidosis	Granulomatous phlebitis
Mineral oil granuloma	Sinusoidal compression	Sarcoidosis
Sarcoidosis	By enlarged Kupffer cells (Gaucher's disease, visceral Leishmaniasis)	Mycobacterium species
Hepatoportal sclerosis	By enlarged fat-laden hepatocytes (Alcoholic hepatitis, AFLP)	Lipogranulomas
Peliosis hepatitis		Mineral oil granuloma
Partial nodular transformation		Hepatic vein outflow tract obstruction (HVOTO, Budd-Chiari syndrome)-Idiopathic, prothrombotic states
Noncirrhotic portal fibrosis (NCPF)/		
Idiopathic portal hypertension (IPH)		
Post-hepatic		
FHVP high, RAP normal or high, WHVP high, HVPG normal or high, PVP high, ISP high**		
Inferior vena cava obstruction-web, thrombosis, tumour, enlarged caudate lobe		
Constrictive pericarditis		
Tricuspid regurgitation		
Severe right-sided heart failure		
Restrictive cardiomyopathy		