Q. 8

What types of vascular disease affect the liver?

- Tumours, benign and malignant
- Congenital malformations, including Hereditary haemorrhagic telangiectasia
- Arterial disease – aneurysm, thrombosis, arteritis
- Peliosis hepatis
- Idiopathic portal hypertension
- Sinusoidal obstruction syndrome = veno-occlusive disease
- Portal vein thrombosis / splanchnic veins
- **Budd-Chiari Syndrome**

Definition of Budd-Chiari syndrome

- Hepatic venous outflow obstruction but excludes cardiac causes and SOS
- Independent of level, so could be from small hepatic venules to level of IVC into right atrium
- Independent of mechanism, so could be **Secondary**
  - Local invasion: HCC, RCC, ACC, EHE, ASa
  - Compression of wall: abscess, cysts, trauma, FNH, kinking

Primary causes Budd-Chiari Syndrome

-

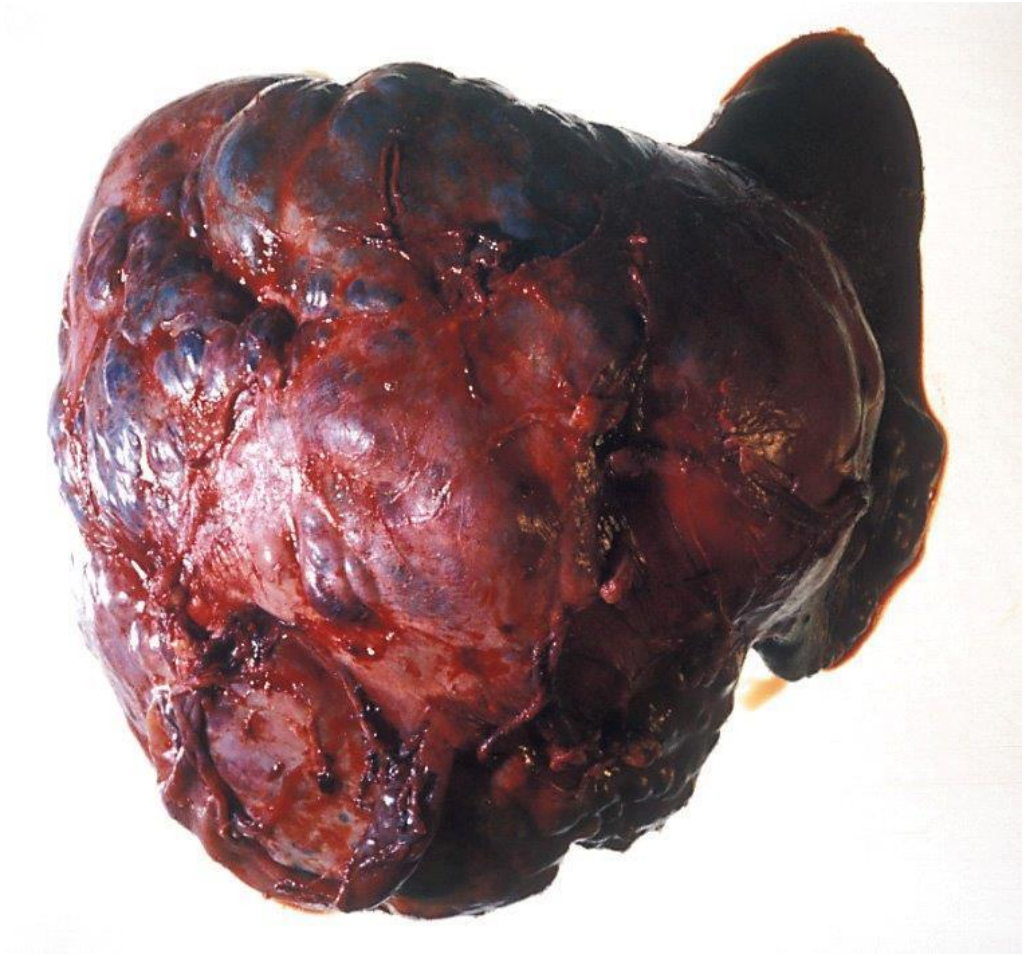
Risk factor	BCS Frequency (%)	PVT Frequency (%)
Thrombophilia		
Inherited	21	35
Acquired	44	19
Myeloproliferative neoplasm	49	21
JAK2 pos	29	16
Hormonal factors	38	44
Oral contraceptives	33	44
Pregnancy	6	0
PNH	19	0
Other systemic factors	23	n.d.
Local factors	0	21

EASL J Hepatol 2016, 64.

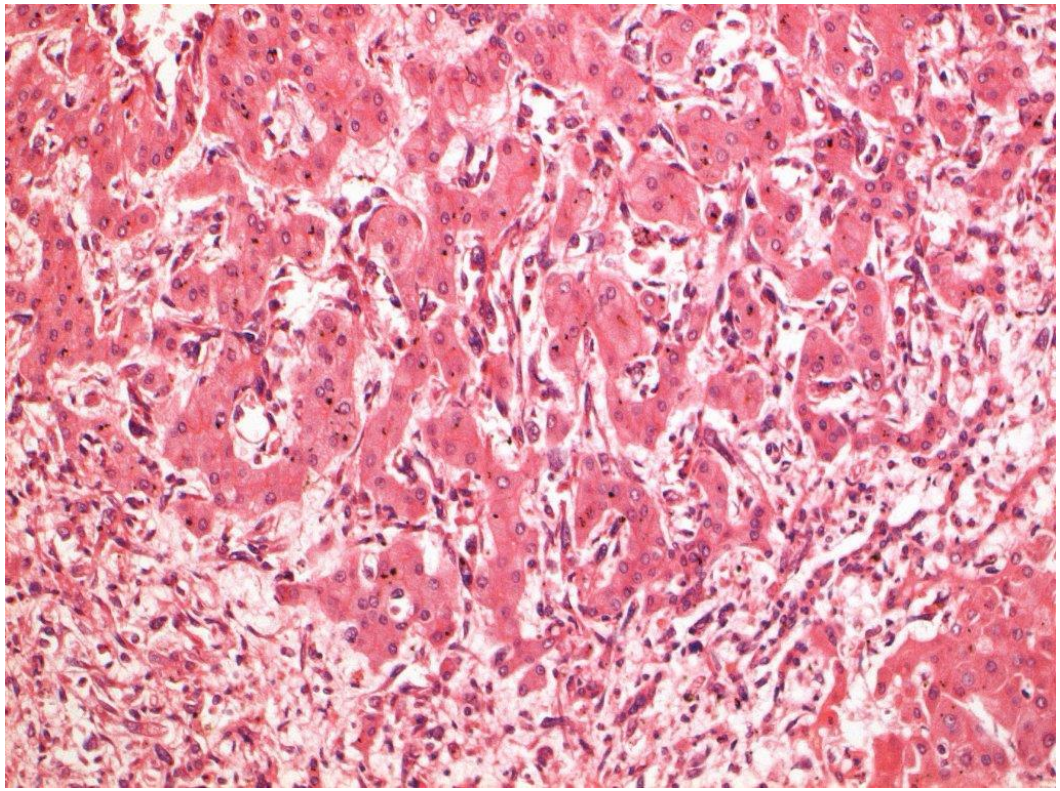
Presentation of B-C S

- Marked variability – asymptomatic to acute liver failure, but more often acute on chronic, with decompensation.
- Duration of symptoms not correlate well with age of thrombi
  - Ascites, hepatomegaly, pain, varices

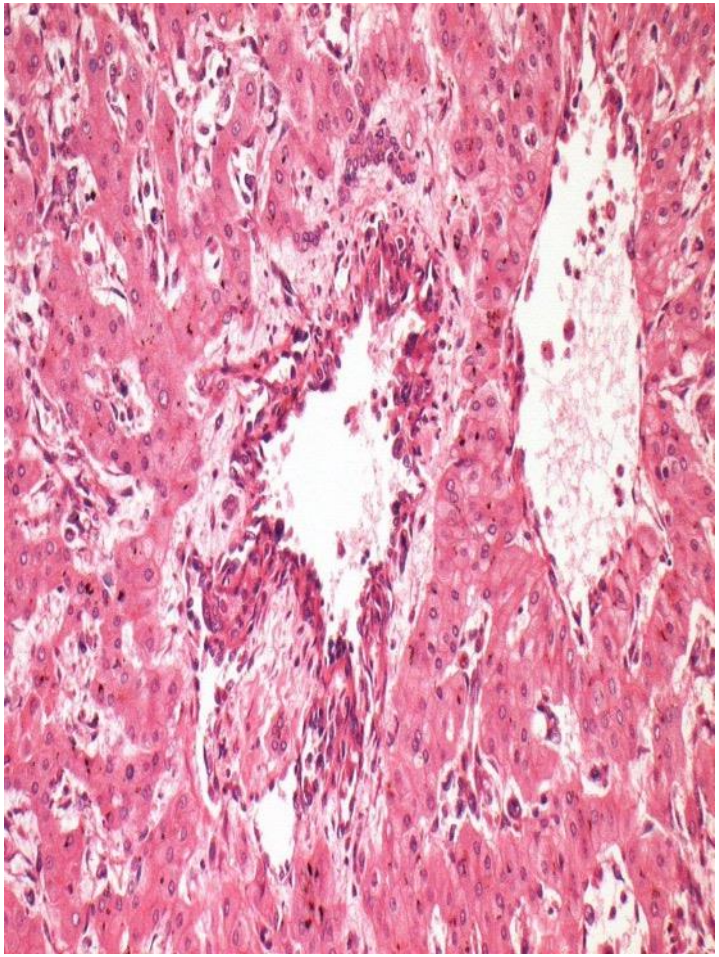
- Management is different
- Acute severe BCS – super-urgent listing for transplantation



48yr M acute RUQ and ascites
with hepatomegaly –
transplanted Liver 3kg



Atypical endothelial like cells in THV and within sinusoids , with dilatation
=Angiosarcoma

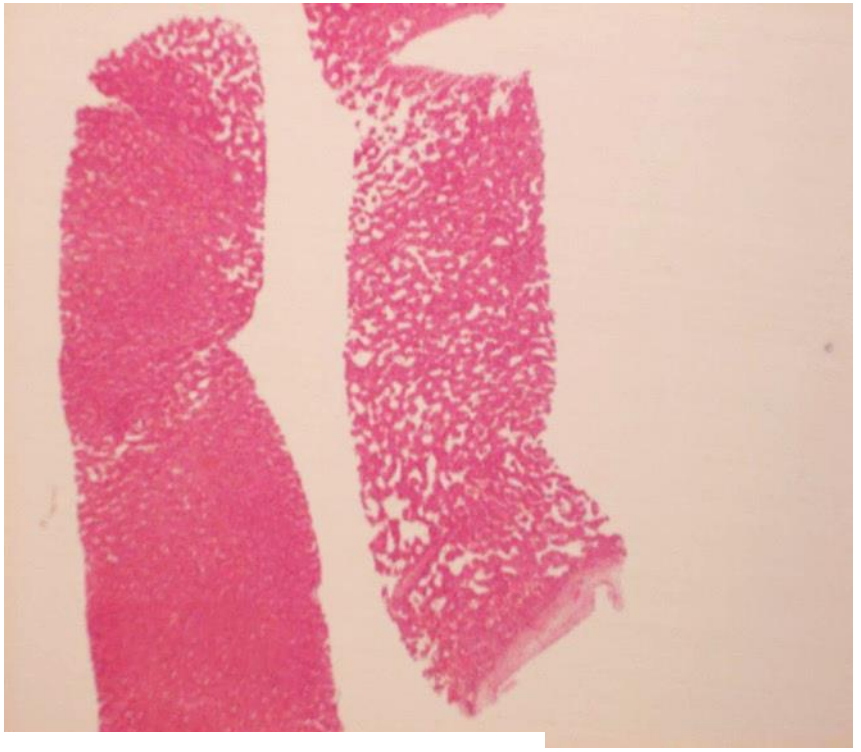


Now severe acute BCS assessed by TJ biopsy – and pressures

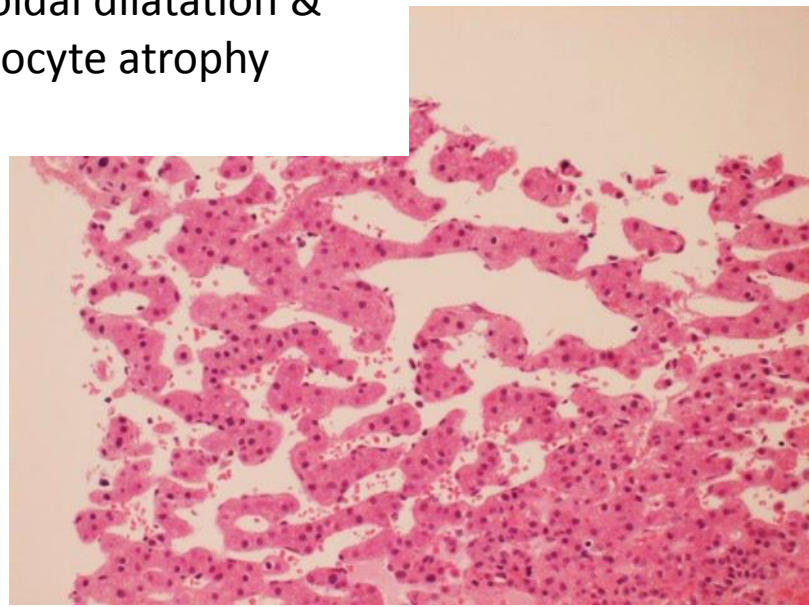
Histology on a biopsy

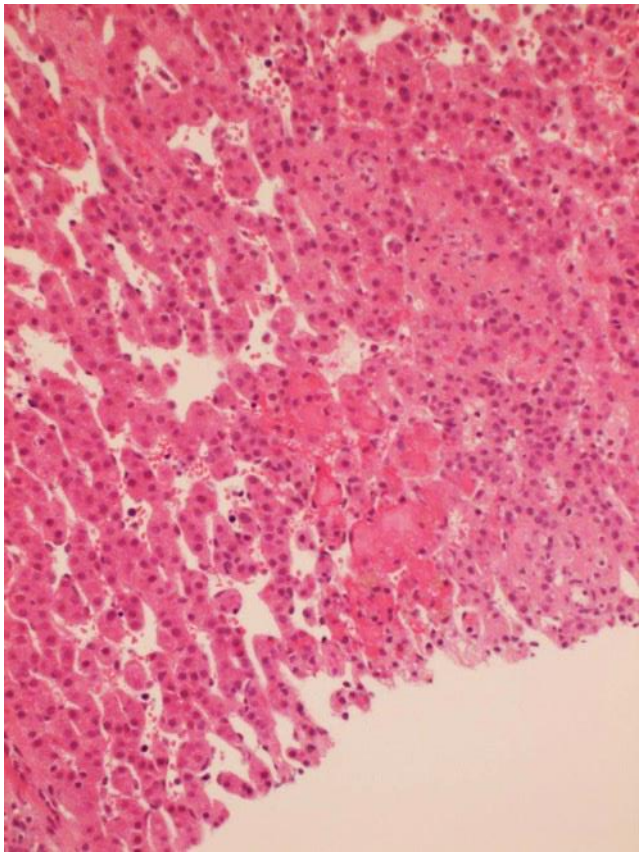
- Unusual to see thrombi
- Indirect evidence is congestion, dilatation, haemorrhage into plates with cell loss and may be fibrosis in perivenular areas.

No thrombi – near normal

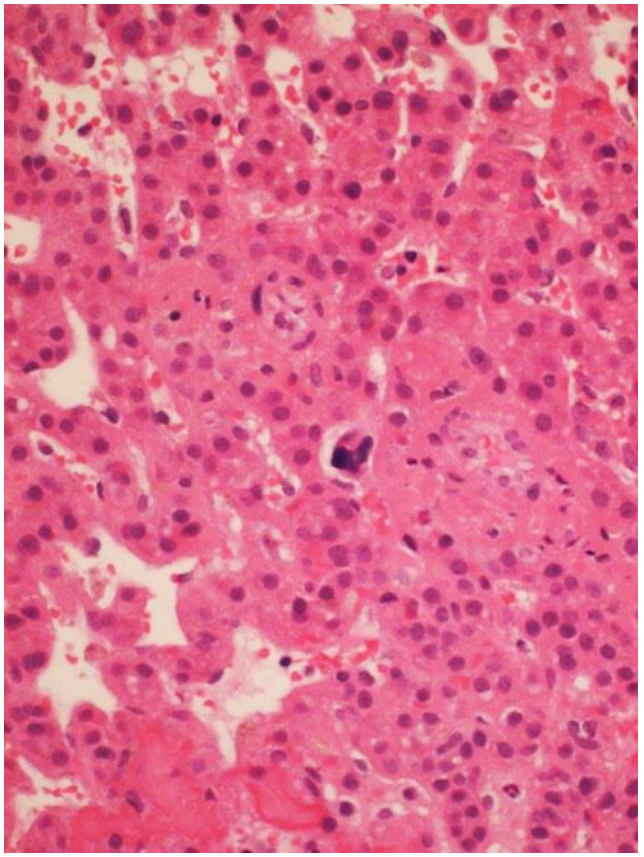


Focal prominent sinusoidal dilatation & hepatocyte atrophy

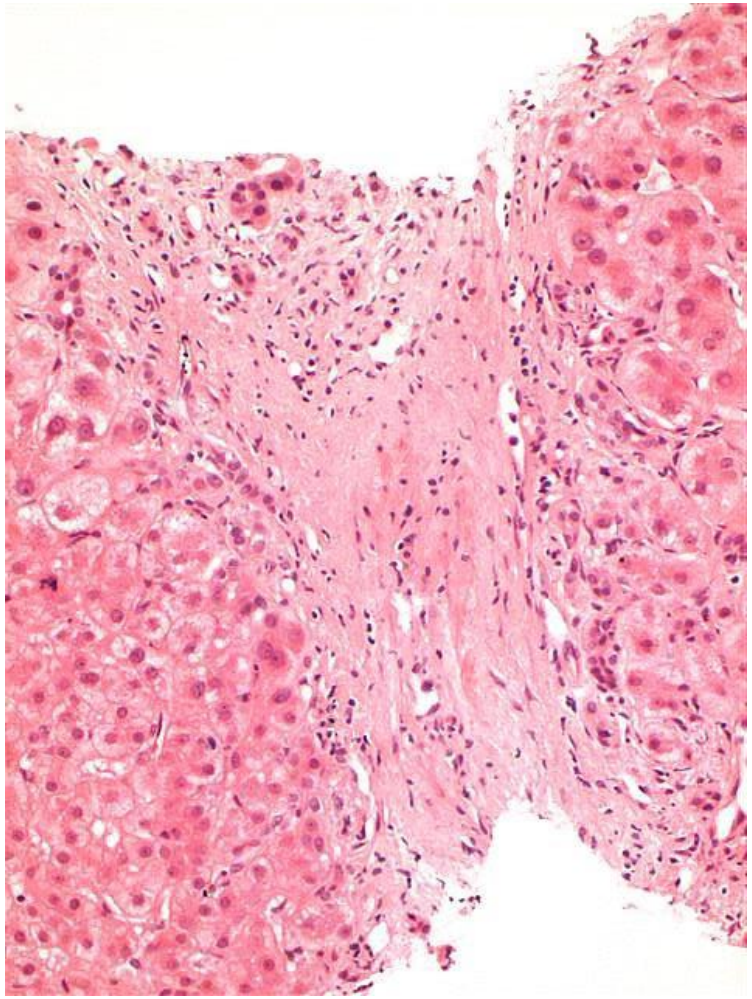




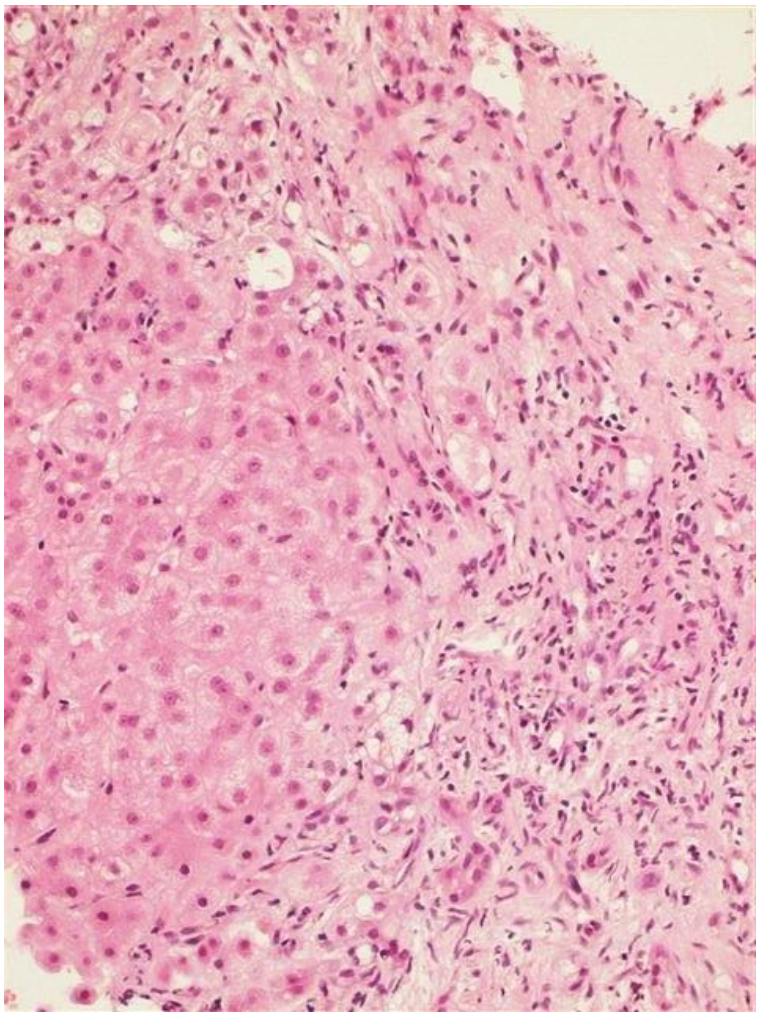
Haemorrhage into plates



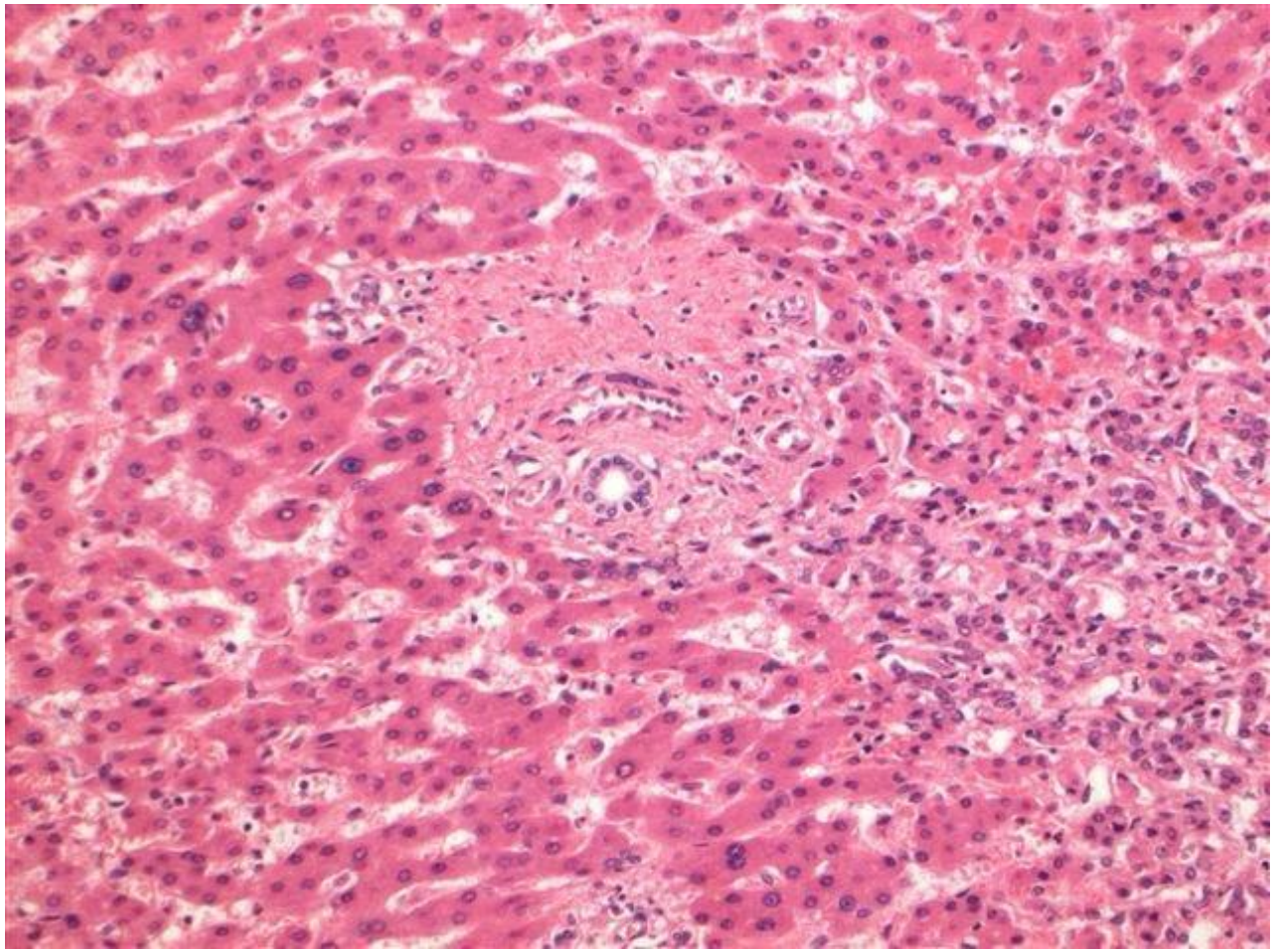
And features of EMH



Progressive perivenular fibrosis, bridging and nodule formation



Ductular reaction is common with progression



About 20% also develop portal venulear thrombosis and fibrosis

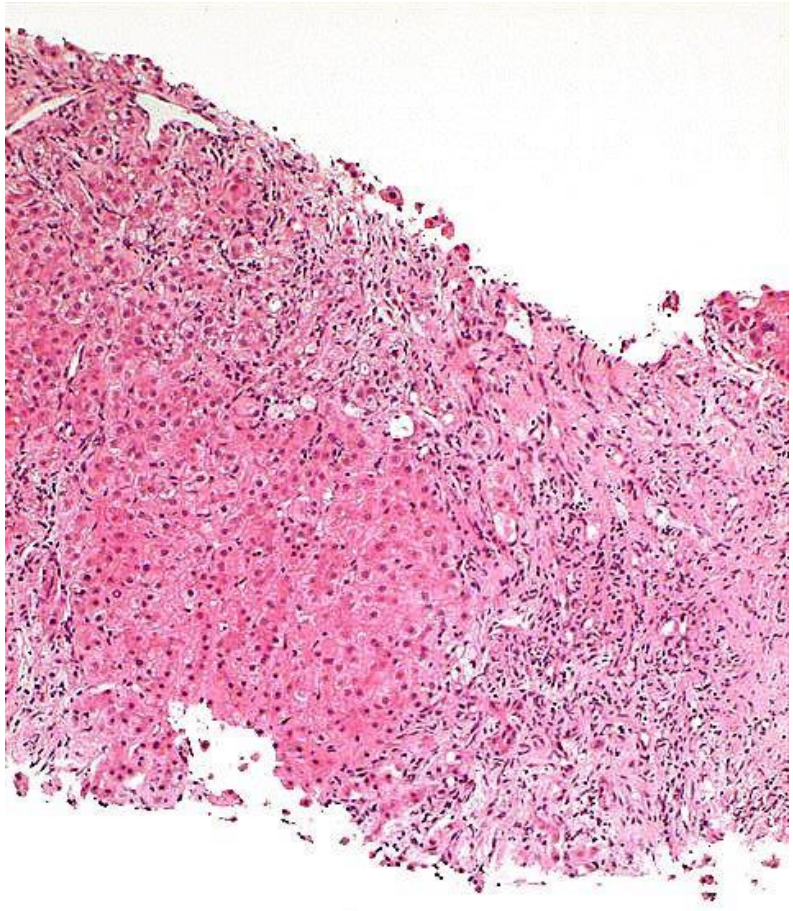
Treatment

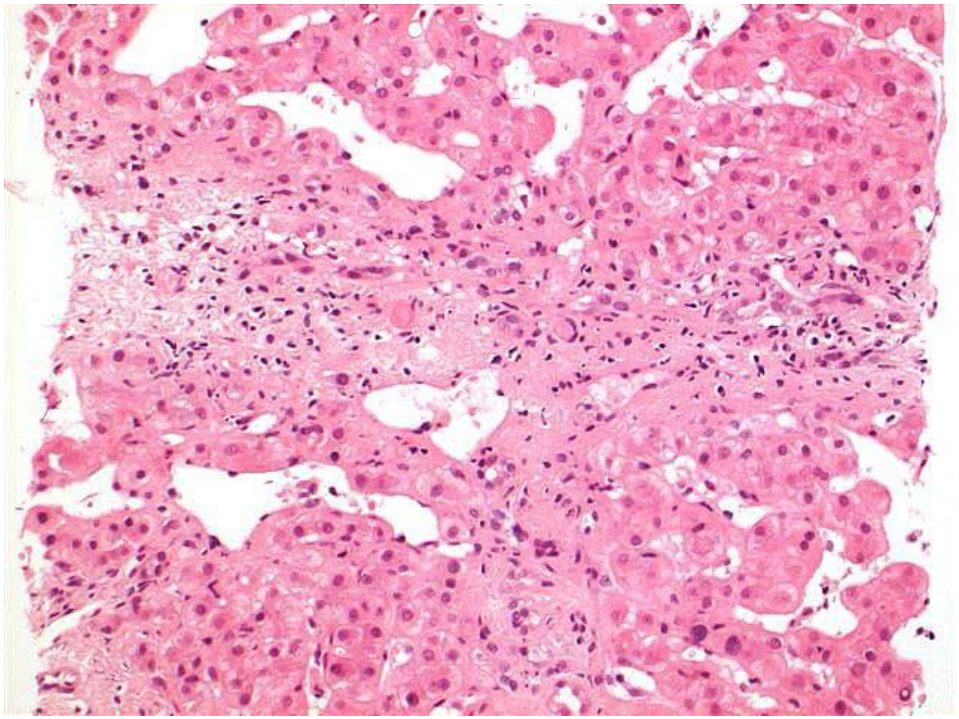
- Medical – anticoagulant; and underlying proliferative disorder
- Thrombolysis, stenting, angioplasty
- TIPS
- Liver transplantation (can also correct the congenital clotting abnormalities).

- Q from clinician – any suggestion of outflow – triple phase CT indicated, as veins not clear on other imaging
- Anything acute to go for
- Cause of cirrhosis – still use anti- thrombotic agents.

Severe refractory ascites –
liver transplant

Cirrhosis ? Aetiology (rather than ? BCS)





Little inflammation, no fat, ductular reaction but normal bile ducts

DO an EVG