Vascular disease of the liver

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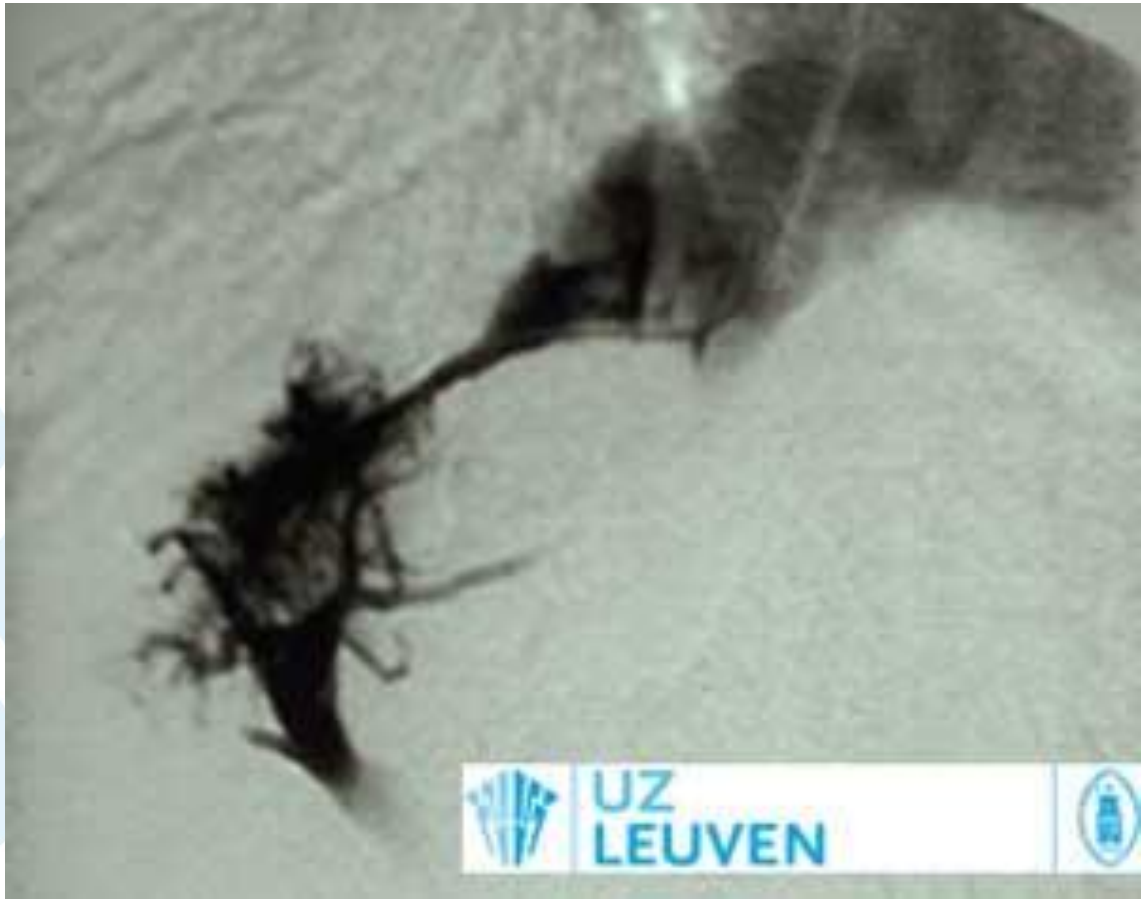
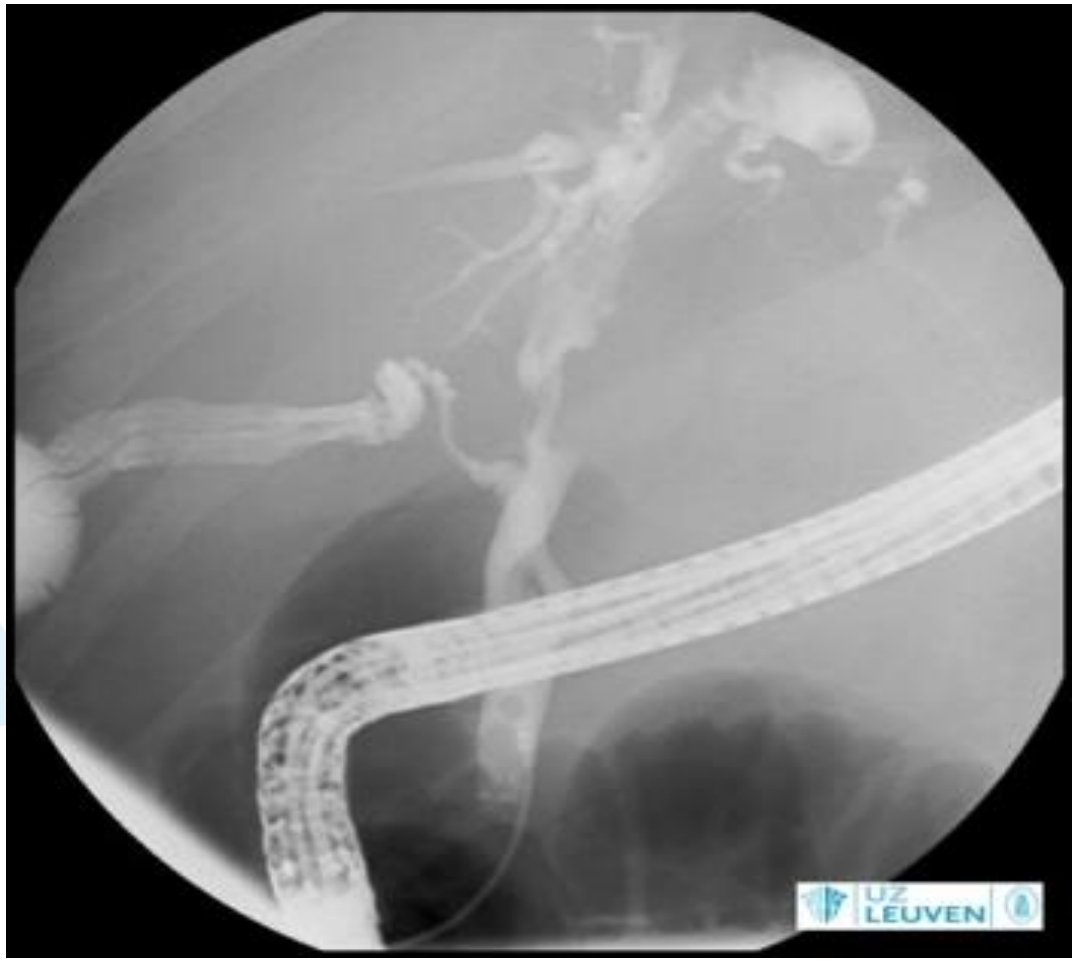
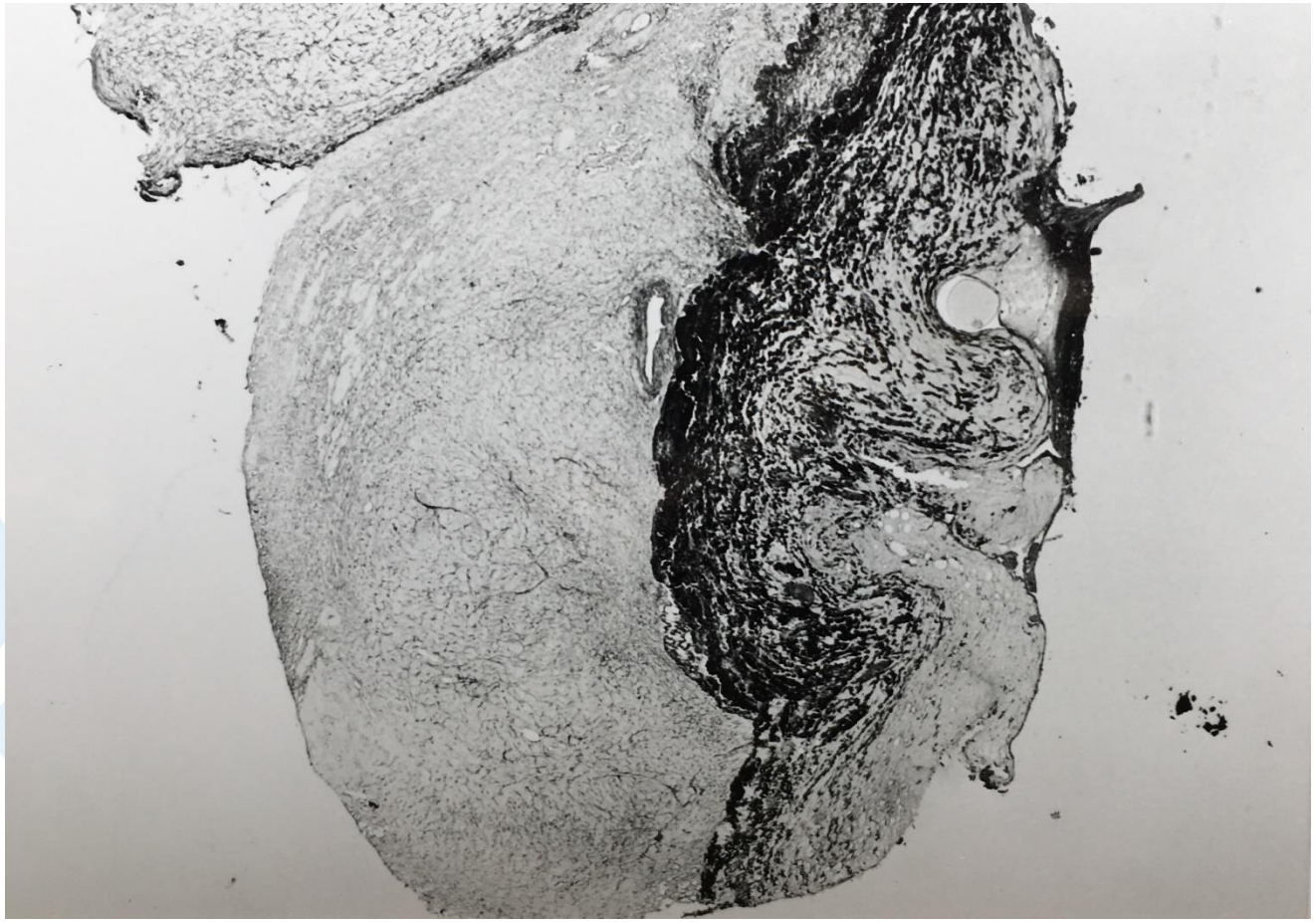
Young female patient with sudden tense ascites and hepatomegaly



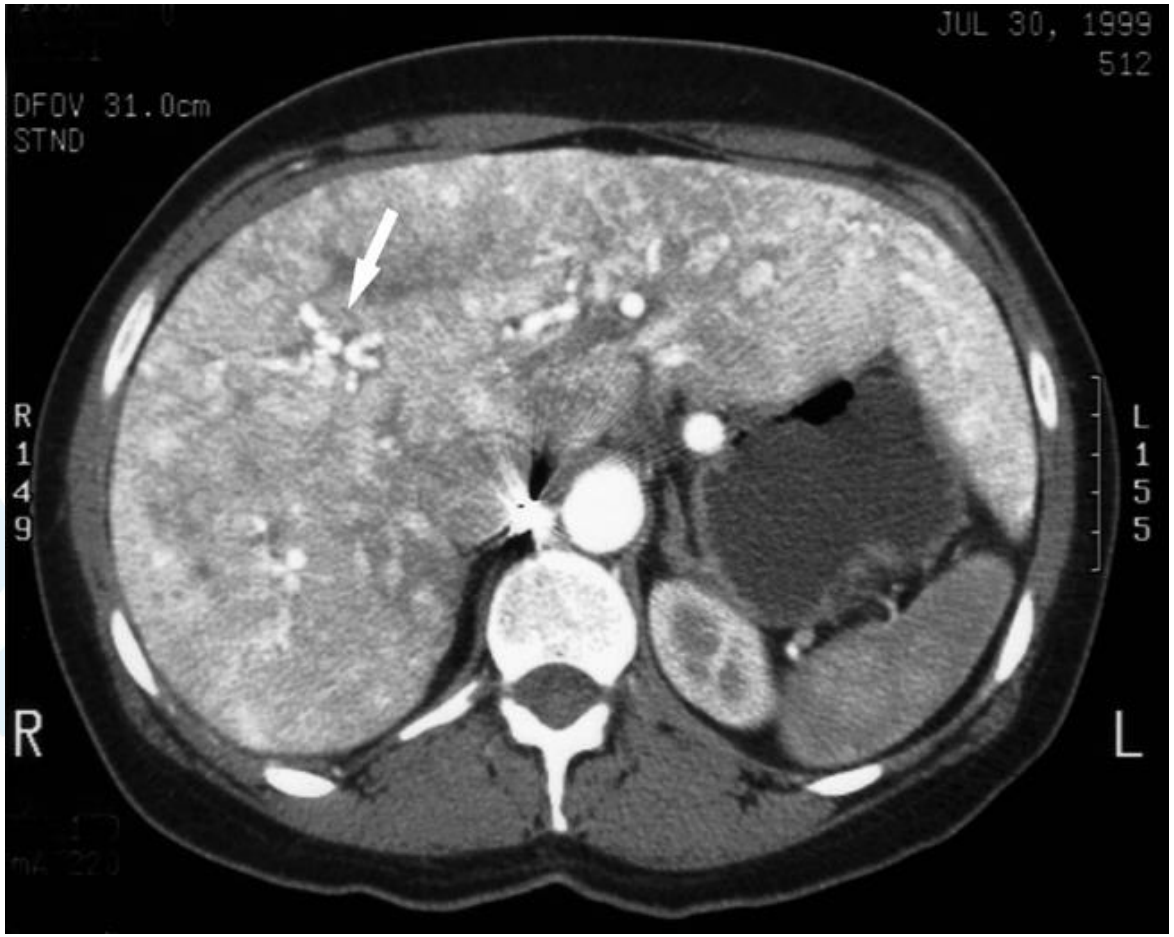
Patient with portal vein trombosis with jaundice and recurrent cholangitis



Patient with variceal bleeding without cirrhosis



Patient with hemoglobine of 4 g/dl and enlarged liver & numerous dilated vessels



Vascular diseases of the liver

- 1.Primary Budd-Chiari syndrome
- 2. Portal vein and splanchic vein trombosis (non-cirrhotic)
- 3. Idiopathic non-cirrhotic portal hypertension
- 4. Hereditary haemorrhagic telangiectasia
- 5. Sinusoidal obstruction syndrome (SOS)

General characteristics

- Main complications
  = non-cirrhotic portal hypertension
  ≠ liver impairment
- Rare disorders : 5/10.000
- Young

Budd Chiari and Portal vein trombosis : Aetiology of trombosis (1)

1. Systemic risk factors (inherented and acquired trombophilia factors)
    - Budd Chiari : 84% (combined factors : 46%)
    - PV trombosis : 42% (combined factors : 10%)

2. Local risk factors (abdominal surgery, infections/inflammation and malignancies)

Budd Chiari and PV trombosis occur simultaneously in 15%

Darwish MS, Ann intern Med 2009
Plessier A, Hepatology 2010

Budd-Chiari and portal vein trombosis : Aetiology of trombosis (2)

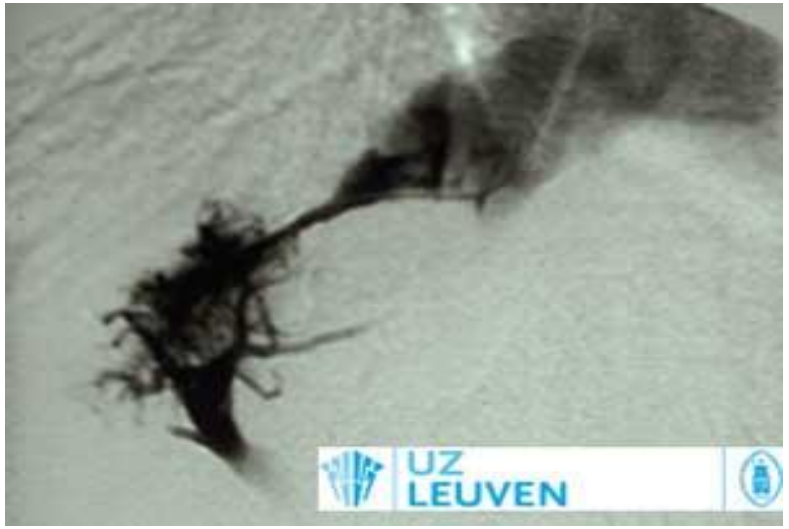
Aetiological factors in Budd-Chiari syndrome and portal vein thrombosis

	BCS	PVT
Risk factor	Frequency (%)	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

BCS, Budd-Chiari syndrome; PVT, portal vein thrombosis; PNH, paroxysmal nocturnal haemoglobinuria; n.d, no date.

EASL Clinical Practic Guidelines 2016

1.Primary Budd-Chiari



- Clinical presentation (Acute liver failure – “cirrhosis”)
  - Ascites : 83%
  - Hepatomegaly : 67%
  - (Abdominal pain : 61%)

Darwish MS, Ann Intern Med 2009

Treatment

- Anticoagulant therapy as soon as possible for an indefinite time *
  - even in case of oesophageal varices
  - even after LTx

Martens P and Nevens F, United European Gastroenterol J 2015

Pitfalls

- Differential diagnosis with pericardial disease

- Hepatic nodules : 60-80%
  (due to perfussion disturbance) [1]
  dd HCC (cumulative incidence of 4% at 5 years)

- LTx is indicated even if in some of the patients its is a premalignant condition !

(1) Moucari R, GUT 2008

2.Portal vein trombosis (non-cirrhotic)

- Clinical presentation :
  Acute abdominal pain (90%) and systemic inflammatory response

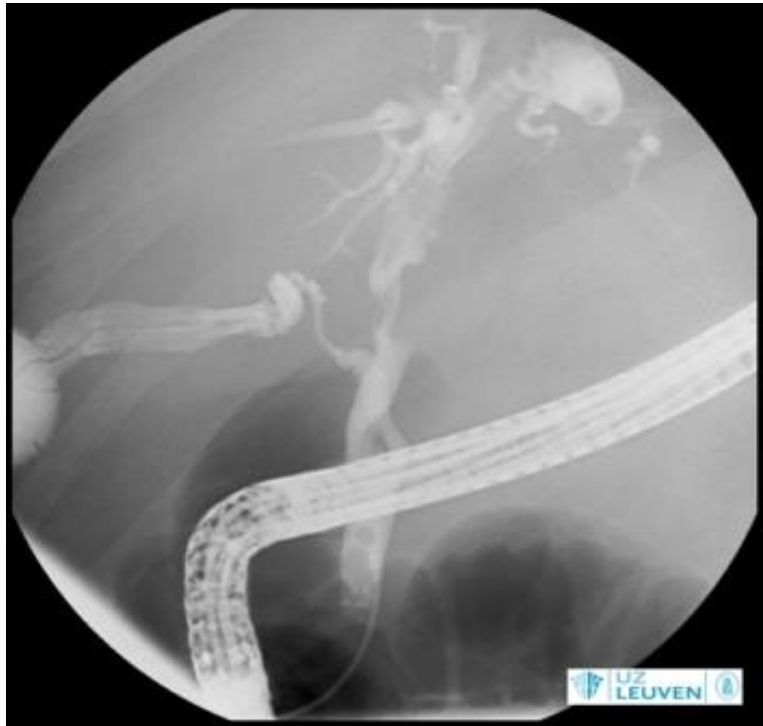
- Complication (short term):
  Intestinal infarction in case of porto-mesenteric vein trombosis (mortality of 60%)

Complications (long-term)

= Cavernomatous transformation

- Non-cirrhotic PH features
- Abdominal angor (diffuse splanchnic trombosis)
  can be an indication for small bowel transplantation !

- Portal cholangiopathy (bile stones)



Treatment

- Anticoagulation for an indefinitive time
  for ≥ 6 months (no trombophilia)

  -to prevent mesenterial venous infarction
  (reduction to 2-20%)

  -to achieve portal vein recanalisation

Pitfalls

- Recognising venous mesenteric infarction is difficult
  = persisting several abdominal pain, rectal bleeding
  and organ failure
- Acute trombosis may superimpose on a cavernoma

- Avoid trombolysis
  (major procedure-related bleeds : 50%)

Llop E, Gut 2011

3.Idiopathic non-cirrhotic portal hypertension

Nomenclature !!

- Hepatoportal sclerosis
- Non-cirrhotic portal fibrosis
- Incomplete septal cirrhosis(1)
- Nodular regenerative hyperplasie

Is an exclusion diagnosis (sarcoidosis; congenital liver fibrosis,…)

(1) Nevens F, Gastroenterology1994

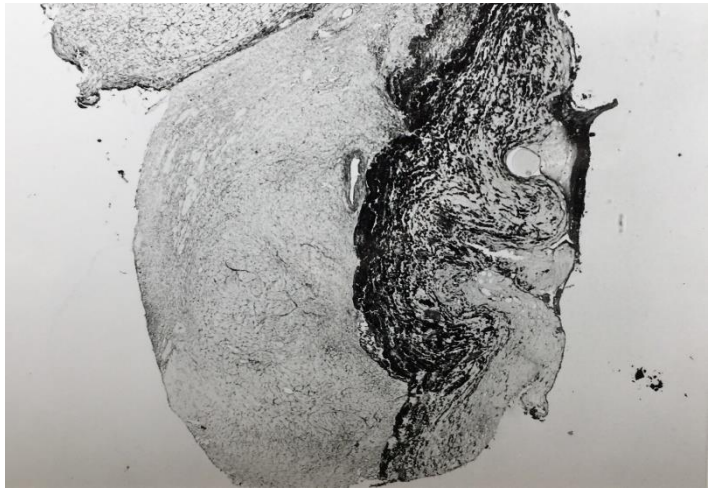
- Associated disorders:
  - Immunological/systemic disorder
  - Medication (azathioprine, didanosine)
  - Trombophilia disorders (40%)
  - HIV
- Clinical characteristics
  - Large spleen[1]

(1) Schouten JN, Aliment Pharmacol Ther 2012

- Diagnosis :

  Biopsy necessary (large specimen)

  - Phlebosclerosis (primary lesion)
  - Nodular regeneration
  - Sinusoidal dilatation
  - Paraportal shunt vessels
  - Perisinusoidal fibrosis



Disturbed intrahepatic circulation

- Prognosis:
  - Higher incidence for PV trombosis (indication for screening)
  - Chronic liver failure can develop with need of LT (presence of ascites is feature of poor prognosis)

Verheij J, Histopathology 2013
Schouten JN, Aliment Pharmacol Ther 2013

Pitfalls

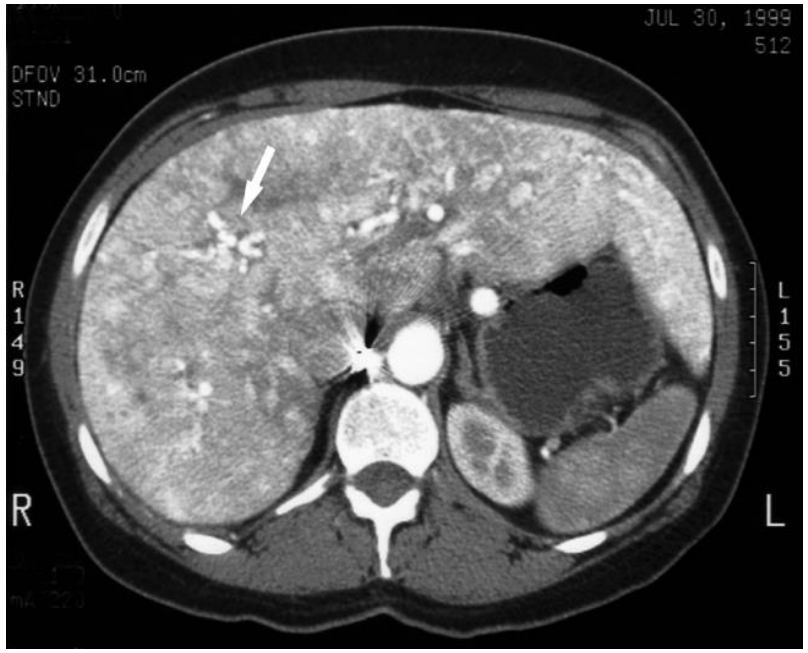
- Misclassified as cirrhosis
  - On ultrasonography : -liver surface nodularity
    -thickening of portal vein wall
    -signs of portal hypertension
  - But elastography < 12 kPa

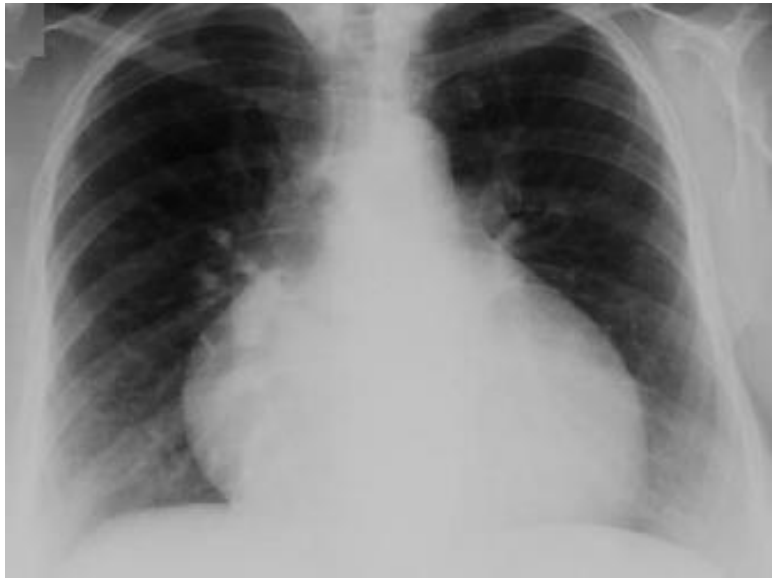
4. Hereditary haemorrhagic telangiectasia (Rendu-Osler-Weber)

- Genetic disorder (autosomal dominant) 1-2/10.000
- Clinical characteristics :
  - Epistaxis
  - Telangiectoses: lip, nose, fingers
  - Middle age woman

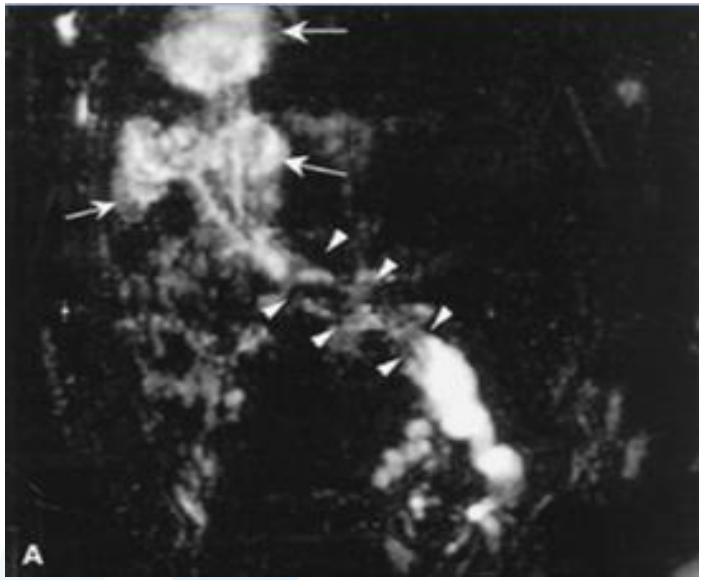
- Pathogenesis:
  Intrahepatic arteriovenous shunting
  - High-output cardiac failure
  - Portal hypertension
  - Biliary and mesenteric ischemia
    (blood flow steal)

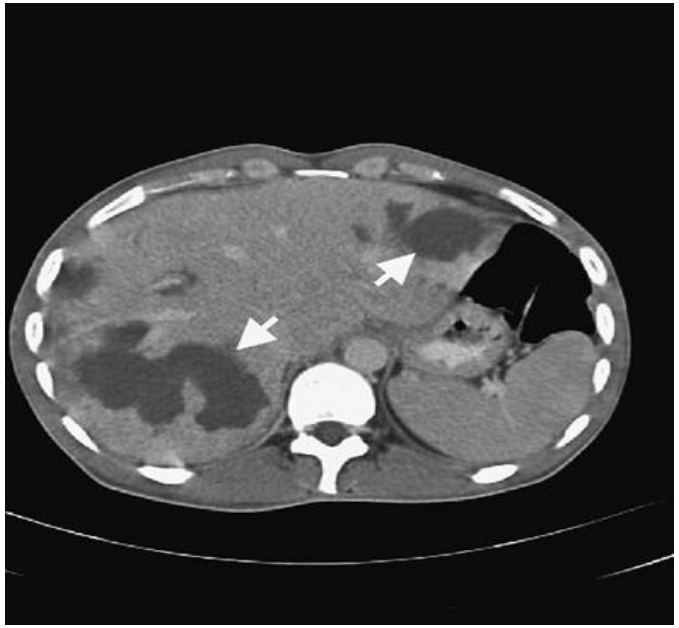
100 fold greater prevalence of FNH

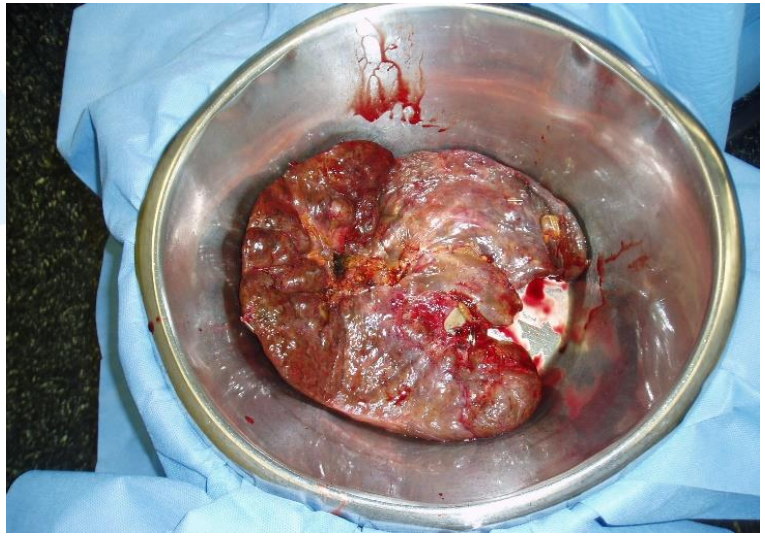




Biliary ischemia







- Treatment: LT

  In case of

  -intractable high-output cardiac failure

  -ischemic bile duct necrosis

But exclude severe pulmonary hypertension !

5. Sinusoidal obstruction syndrome (veno-occlusive disease)

= Sinusoidal congestive obstruction due to injury of sinusoidal endothelial cells resulting in outflow block

- Clinical characteristics
  - Jaundice
  - Tender hepatomegaly
  - Fluid retention

- In patients with -haemotopoietic stem cell transplantation
  -chemotherapy
  -immunosuppression in organ transplantation and inflammatory bowel

- Diagnosis :
  Transjugular liver biopsy (+ pressure measurements)

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