

## Hepatoportal sclerosis

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## Terminology

- Hepatoportal sclerosis
- Non-cirrhotic portal fibrosis
- Idiopathic portal hypertension
- Obliterative portal venopathy of the liver
- Non-cirrhotic intrahepatic portal hypertension

## Clinical presentation

- Portal hypertension
  - EV bleeding
  - Splenomegaly
  - Ascites
- Hypersplenism

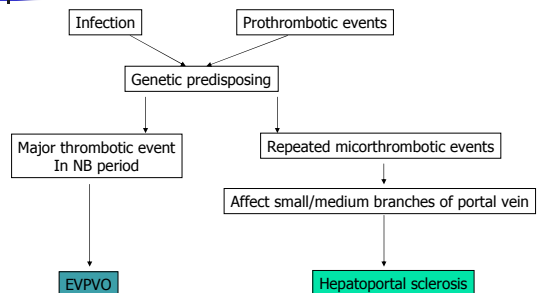
## Clinical presentation

- Liver size- usually normal
- LFTs – normal, but some evidence of liver impairment can occur in the later course of disease

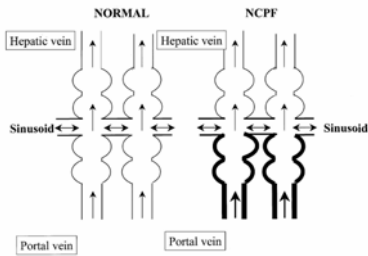
## Pathogenesis

- Infection
  - Umbilical sepsis, bacterial infection, diarrheal episode
- Trace metals/chemical exposure
  - Arsenic, vinyl chloride monomer, copper sulfate, chemotherapeutic agents e.g. MTX, 6-MP, AZT, steroids
- Immunologic process
  - Decreased T4/T8 lymphocyte
  - Decreased CMI status

## Pathogenesis

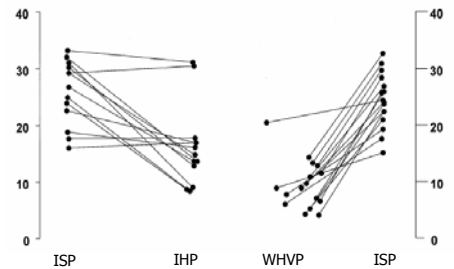


## Pathogenesis



Sarin SK, et al. J Gastroenterol Hepatol 2002;17:526

## Hemodynamics



Sarin SK, et al. Gut 1987;28:260.

## Diagnosis

- Doppler ultrasound – periportal hyperechogenicity
- Portal angiogram, MRV, multidetector spiral CT
  - Intrahepatic portal vein occlusion
  - Cavernous transformation at the portal hilar
  - Collateral vessels
- Liver biopsy

## Pathology

- Normal/subtle changes
- Preserved liver architecture
- No regenerative nodule
- Bile ducts and hepatic artery: normal

Mikkelsen WP, et al. Ann Surg 1965;162:602.

## Pathology

- Fibrous bands in the portal area; disruption of limiting plate may be seen
- Subendothelial/subintimal fibrous thickening of the vessel wall or deposition of perivascular collagen of portal vein branches, central vein, and hepatic vein
- No thrombosis
- Minimal/no chronic inflammation in the portal tract
- Dilated sinusoids

Mikkelsen WP, et al. Ann Surg 1965;162:602.

## Pathology

- Extrahepatic portal vein
  - Normal
- Intrahepatic portal vein
  - Normal
  - Partial occlusion
  - Complete occlusion
    - With collateral/cavernous transformation giving a “withered tree” appearance.

Mikkelsen WP, et al. Ann Surg 1965;162:602.  
Maksoud JG, et al. Am J Surg 1986;151:484.

## Prognosis and treatment

- Prognosis – Good
- Surgical shunt is initial treatment of choice

## Congenital hepatic fibrosis

Childhood type polycystic disease

- as isolated anomaly
- in Meckel's syndrome
- in Adult type polycystic disease
- with dysplastic kidneys

Congenital hepatic fibrosis

Choledochal cyst

Dilated intrahepatic ducts

Caroli's disease

Murray-Lyon IM, et al. Gastroenterology 1973;64:653

Thank you for your attention

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