CASE 1:
A Common Scenario in Medical Indoor

A 55-year-old man was admitted with-

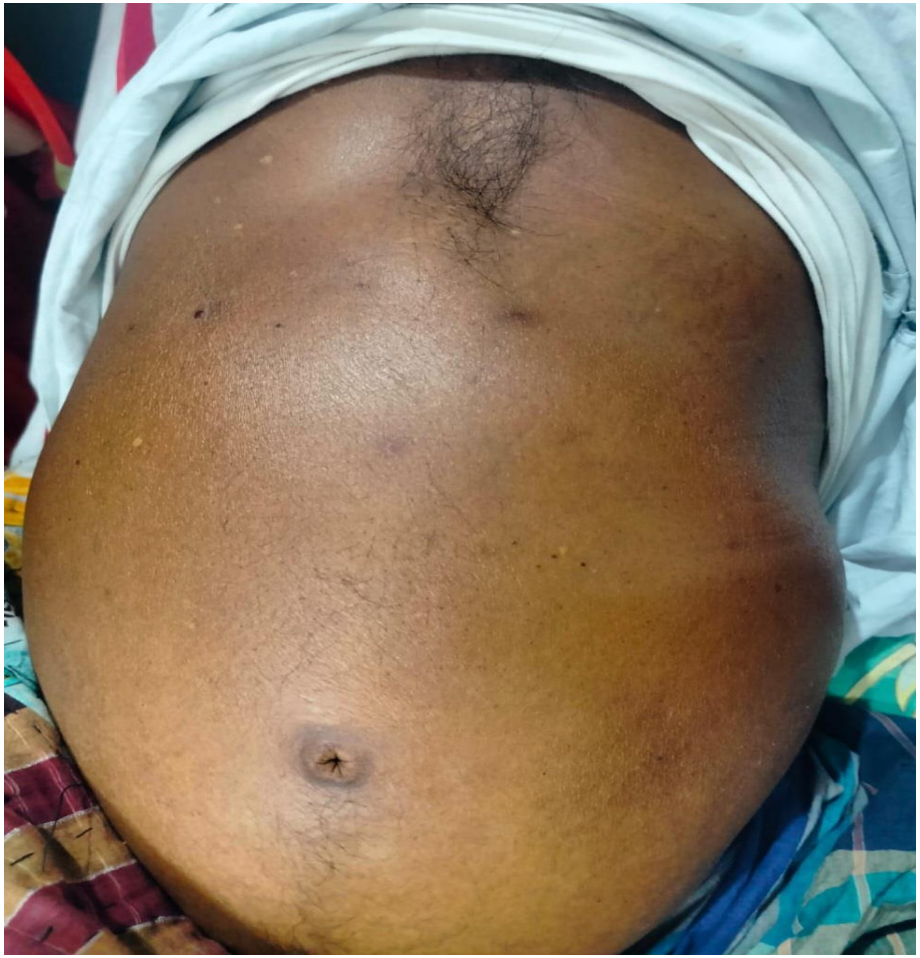
- Hematemesis and melena - 7 days
- Swelling of abdomen and legs - 3 months
- H/O Jaundice- 1 year back

On examination:

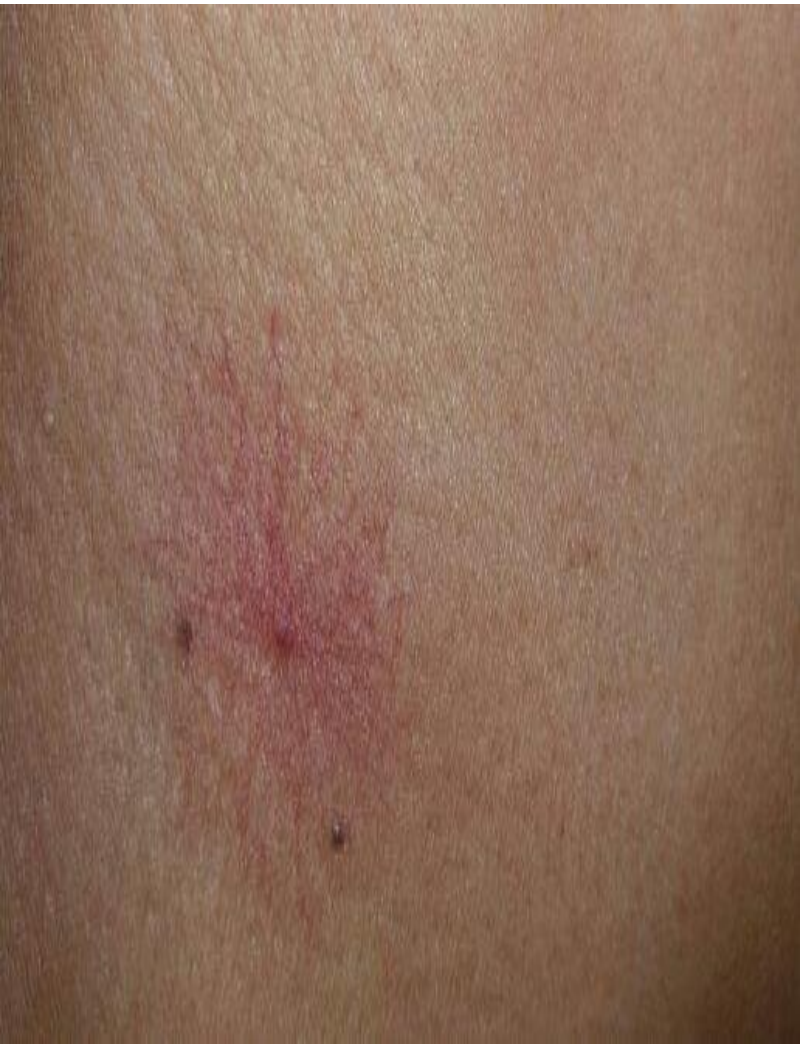
The patient was-

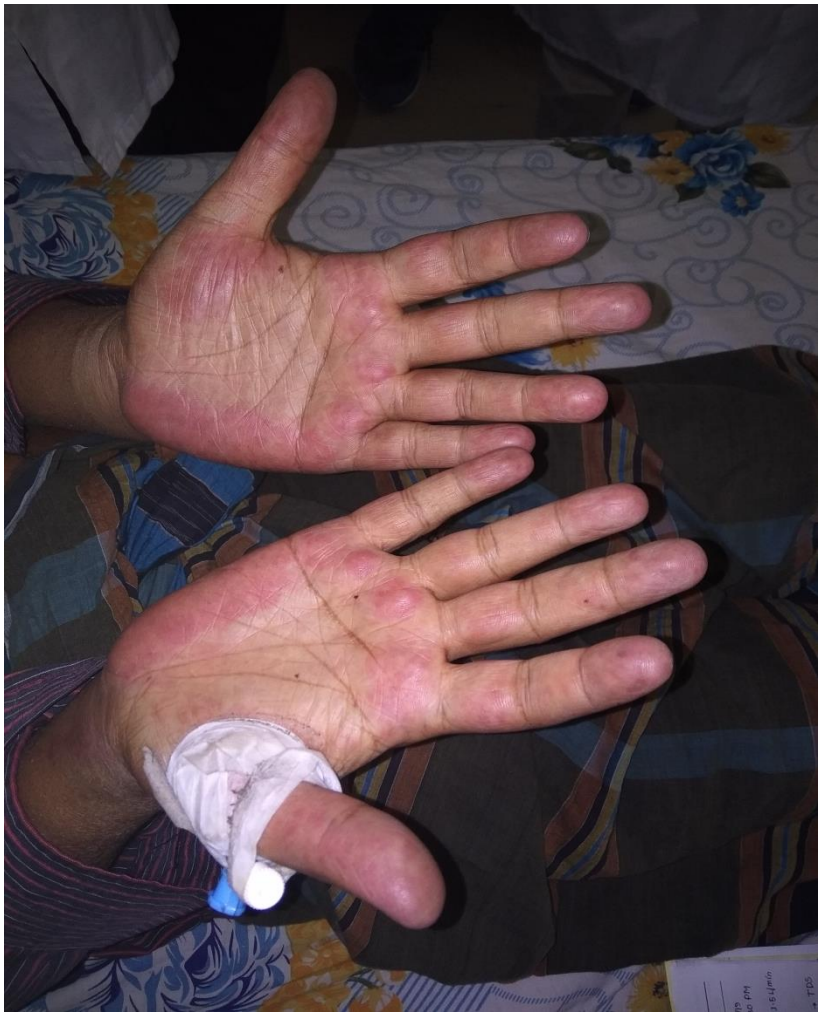
- Mildly icteric
- Moderately anaemic
- Ascites- positive shifting dullness
- Splenomegaly
- Testicular atrophy

On examination:



On examination:





Investigation reveals:

- HBs Ag (+)
- A: G Ratio= 0.5:1
- USG of Abdomen-
  - **coarse hepatic parenchyma**
  - gross ascites
  - splenomegaly
- Endoscopy Upper GI- Grade III Esophageal varices

Diagnosis:

Chronic Liver Disease with Portal Hypertension due to HBV infection

Cirrhosis of liver- if histopathology could be done

Some almost similar scenarios-

CASE -2

A 28-year-old pregnant (18 weeks) lady with-

- Rapid swelling of abdomen and legs - 10 days
- Abdominal pain- 10 days
- Irritability - last 2 days

CASE 2 contd…

- Ascites- evidenced by fluid thrill
- Tender hepatomegaly
- Flapping tremor

CASE 2 contd…

- SGPT- 240 U/L
- Prothrombin time- 25 seconds
- Doppler USG- **occlusion of hepatic veins**

CASE 3:

A 55-year-old man with history of **Primary Myelofibrosis** presented with worsening abdominal distension for last 2 months. The patient had a history of splenectomy 1 year back due to the progression of his primary disease.

CASE 3 contd…

- The patient was dyspneic with huge ascites evidenced by fluid thrill
- Liver function tests - normal
- USG - **normal hepatic parenchyma with ascites with extensive portal vein thrombosis**

CASE 4:

- A 50-year-old man with HIV, on treatment for 8 years, presented with progressive abdominal distension and 2 episodes of hematemesis
- He was receiving Tenofovir, Didanosine and Nevirapine.
- Examination revealed ascites evidenced by shifting dullness

CASE 4 contd…

- Lab values - normal Hb and WBC count with low platelets (1, 10,000 IU/ml)
- Liver and renal functions - normal
- Doppler USG and Abdominal CT revealed only **ascites and splenomegaly**

CASE 4 contd…

- CLD work up was negative including HBV, HCV, Anti-HBc Ab, AMA, ASMA, ANA
- Stool examination - normal
- Endoscopy revealed **grade II esophageal varices**

CASE 4 contd…

A transjugular liver biopsy revealed a **hepatic venous pressure gradient (HVPG) of 14 mm Hg without any bridging fibrosis of liver.**

CASE 5:

- A 60-year-old lady was admitted for ascites and had a recent paracentesis. She was being treated as a case of SLE.
- 8 months back, she developed hematemesis. Endoscopy revealed esophageal varices for which EVL was done.

CASE 5 contd…

On examination-

- Her vitals were almost normal except being a bit dyspneic
- Ascites was present evidenced by shifting dullness

CASE 5 contd…

- Liver functions - normal
- Serological evaluations - negative for -
  - Viral
  - auto immune
  - Genetic causes
- Liver biopsy - consistent with **nodular regenerative hyperplasia of liver parenchyma but absence of fibrous septa**

What is the COMMON FEATURE in last four cases?

- **All of them had Portal HTN**

BUT

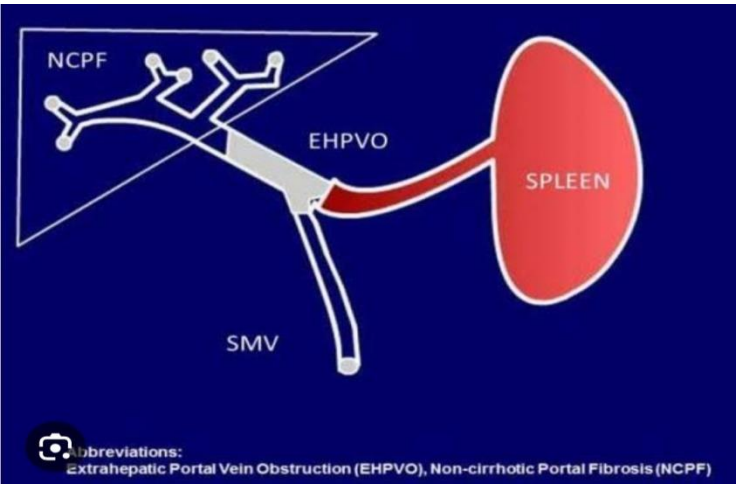
- **Hepatic Function- preserved**
- **Hepatic morphology- preserved**

NOT ALL PORTAL HYPERTENSIONS MEAN CIRRHOSIS

Portal Perspectives:

A Holistic Approach to

Non-Cirrhotic Portal Hypertension (NCPH)Management



Dr. Sarmistha Biswas

What is NCPH?

A rare disease characterized by

- Splenomegaly- moderate to massive
- Hypersplenism- +/-
- Liver function- preserved / mild alteration
- **No liver cirrhosis**

[Ref: S.K Sarin et al.Clin Liver Dis.2006]

What is NCPH?

Hepatic Venous Pressure Gradient (HVPG)

WHVP- FHVP >5 mm Hg

[Ref: Harrisons Principles of Internal Medicine. 20th edition.]

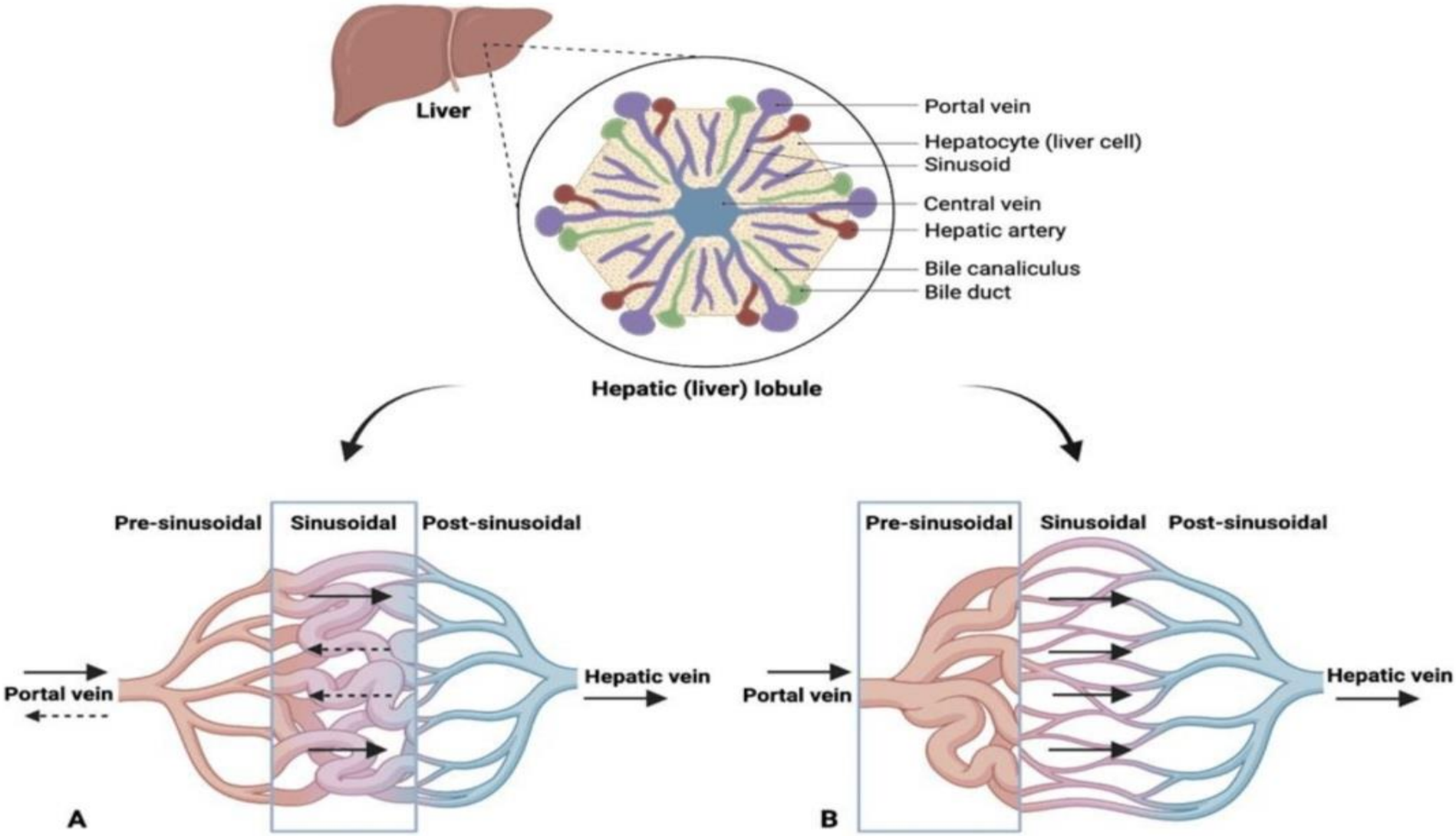
A. J. Sanyal et al. Gastroenterology 2008]

How common it is?

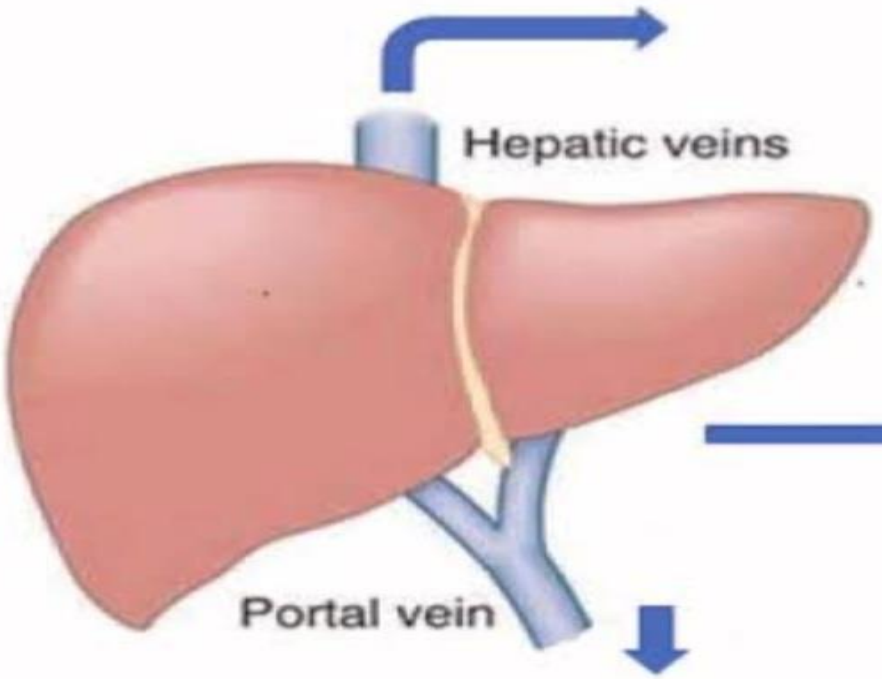
- The incidence varies globally
- **In many cases- exact cause is unknown**
- Only **10-15% cases** of PH- non-cirrhotic

[Ref: Davidson's principle and Practice of Medicine, 23rd Edition]

Cirrhotic and non-cirrhotic PH:



Etiologies:



Post-hepatic
Budd Chiari syndrome
IVC obstruction
Pericarditis
Right heart failure
Tricuspid regurgitation

Hepatic*
Schistosmiasis
Congenital hepatic fibrosis
Nodular regenerative hyperplasia
Idiopathic NCPH
Veno-occlusive disease

Pre-hepatic
Portal vein thrombosis
Extrahepatic portal vein obstruction
Splenic vein thrombosis
Massive splenomegaly

Etiologies:

5 basic categories-

- Prothombotic conditions
- Immunological disorders
- Infections
- Toxins and medications
- Idiopathic

[Ref: Michele Feordaliso et al. Diagnostics 2023]

Prothrombotic conditions:

- Myeloproliferative diseases
- PNH
- Thrombophilia- inherited/ Acquired
- Pregnancy
- OCP
- Neoplasms

[Ref: Harrison's Principles of Internal Medicine, 20th Edition]

Immune related Factors:

Evidences are in favor of-

- Anti phospholipid Ab syndrome
- MCTD
- SLE
- Systemic Sclerosis
- IBD etc.

[Ref: Inagaki H et al. Gastroenterol 2002. Rai T et al.Hepatol.2004 Eapen C et al.Dig. Dis.Sci.2011]

INFECTIONS:

Schistosomiasis

[Commonest infective etiology, but rare outside Egypt]

Data are also in favor of-

- **AIDS**
- **E. Coli**
- **CMV**

[Ref: Davidson's Principles and Practice of Medicine,23rd Edition.]

Amitrano L et al. Gastroenterol 2007.]

INFECTIONS:

- More in the **socio economically deprived population**
- Childhood infections-
  - **Frequent umbilical sepsis**
  - **Bacterial infections**
  - **Diarrhea**

[Ref: Rai T et al.Hepatol.2004. Austin A et al. Gut 2004]

TOXINS AND DRUGS:

Evidences are there for-

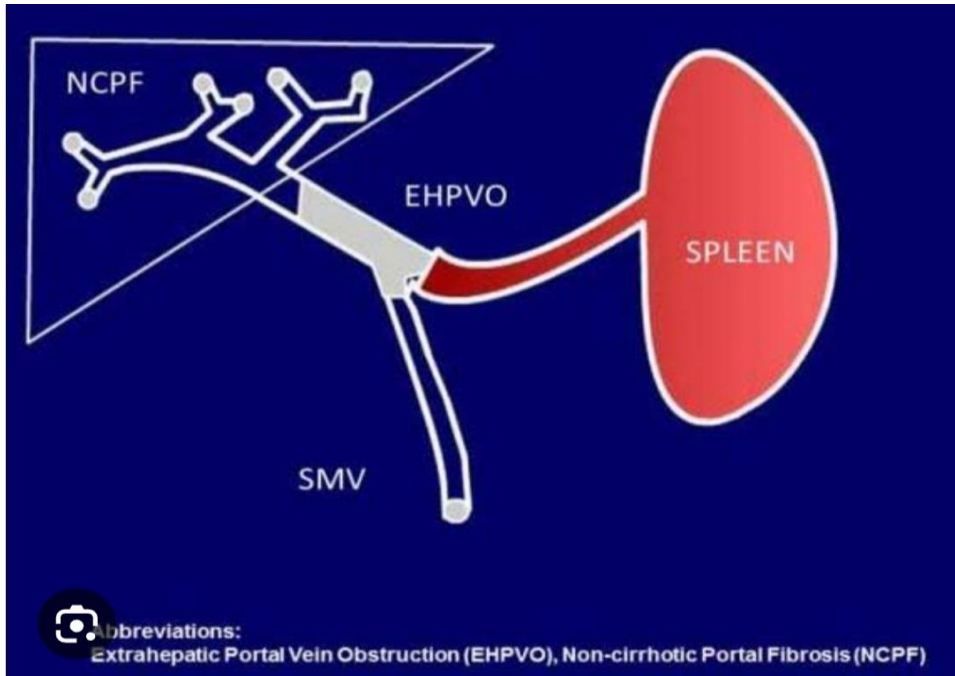
- Azathioprine
- HAART therapy
- Chemotherapy
- Radiotherapy
- Arsenic
- Vinyl Chloride etc

[Ref: Seo J W et al. Intest. Res 2021. Fuentes- Lacouture M C et al. J Med Cases 2021]

IDIOPATHIC (IPH)

Diagnosis is challenging

- No specific test to confirm
- Disease of exclusion
- Liver biopsy – needed for firm diagnosis



IPH: Diagnostic criteria of EASL

All of following 5 criteria must be fulfilled-

1. **Clinical Signs (any one)**:

- Splenomegaly
- Esophageal varices
- Ascites
- Increased HPG
- Portovenous collaterals

IPH: Diagnostic criteria of EASL

2. Exclusion of cirrhosis on liver biopsy

3. Exclusion of chronic liver disease

- HBV/HCV
- ASH/ NASH
- Autoimmune hepatitis
- Storage disease
- PBC

IPH: Diagnostic criteria of EASL

4. Exclusion of conditions causing non-cirrhotic PH

- Congenital hepatic fibrosis
- Sarcoidosis
- Schistosomiasis

5. **Doppler USG or CT- Patent portal and hepatic veins**

Now go back to our cases….

CASE 2: A pregnant lady with features of portal hypertension….

- **USG- obliteration of hepatic veins with reversed flow-**

DIAGNOSIS- BUDD- CHIARY SYNDROME

BUDD-CHIARY SYNDROME:

- Defined as the **obstruction of hepatic venous outflow** that can be located from small hepatic venules to the entrance of IVC into right atrium

- 2 types-
  - Primary
  - Secondary

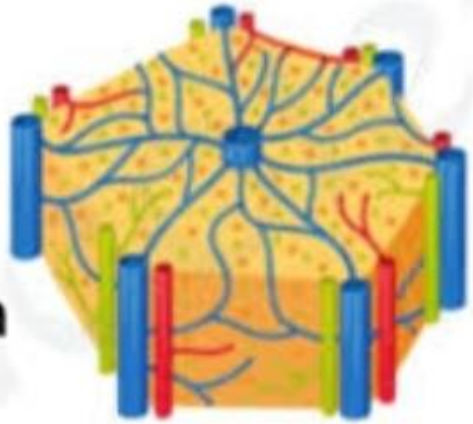
[Ref: EASL Clinical Practice Guideline 2016]

Budd Chiari Syndrome

- Hepatic venous outflow obstruction

Causes :- "5P"

- Polycythemia
- Pills(OCPS)
- PNH
- Protein C & S deficiency
- Pregnancy



Classic Triad

- Abdominal pain
- Ascites
- Hepatomegaly

CASE 3:

A 55-year-old man with Myelofibrosis and radiological evidence of **portal vein obstruction**

DIAGNOSIS: PORTAL VEIN THROMBOSIS

CASE 4:

A patient getting HAART therapy with features of Portal HTN-

Possible culprit- Long term Didanosine therapy

[Ref: Helen Covary et al. Clinical Infectious Diseases.2009.Plessier A et el. Hepatol 2012.]

CASE 5:

An elderly lady with SLE with features of portal hypertension

DIAGNOSIS: Nodular Regenerative Hyperplasia of liver

Commonest cause of non-cirrhotic PH in the developing world.

[Ref: Davidson's Principles and Practice of Medicine,23rd Edition.]

MANAGEMENT :

Vast etiological spectrum, so diagnosis is challenging

- **High quality liver biopsy**
- **Detailed clinical information**
- **Expert pathologist opinion**
- **Radiological expertise**

necessary for diagnosis

MANAGEMENT:

- **Considered as a rare entity**

- **Paucity** of-

  -well-designed,

  -large scale

  -prospective studies on NCPH

poses a great obstacle to formulate evidence-based guideline

GUIDELINES??

EASL Clinical Practice Guidelines: Vascular diseases of the liver

European Association for the Study of the Liver *

Published: October 27, 2015 •

DOI: https://doi.org/10.1016/j.jhep.2015.07.040 •

GUIDELINES??

Hepatology International > Article

Asia–Pacific association for study of liver guidelines on management of ascites in liver disease

Guidelines | Published: 26 May 2023

Volume 17, pages 792–826, (2023)

GUIDELINES???

PubMed®

Advanced User Guide

Noncirrhotic portal fibrosis/idiopathic portal hypertension: APASL recommendations for diagnosis and treatment

Shiv Kumar Sarin et al. Hepatol Int. 2007 Sep.

Latest recommendations: for BCS and PVT

Investigate patients for local and systemic prothrombotic factors -

- **Systemic factors-**

  -Thrombophilia

  - MPDs

  - PNH

  - Autoimmune diseases

[Ref: EASL Clinical Practice Guideline: Vascular diseases of the liver.2016]

Latest recommendations: for BCS and PVT

- **Local factors-**

  - Intra-abdominal **inflammatory conditions**

  - Intra-abdominal **malignancy**

[Ref: EASL Clinical Practice Guideline: Vascular diseases of the liver.2016]

Latest recommendations: for BCS

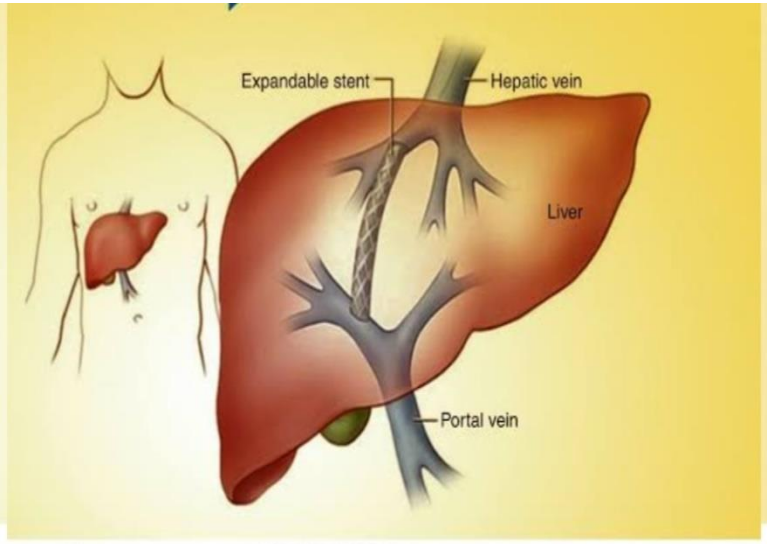
- **Doppler USG** -1st line investigation
- **MRI and CT** can be done for confirmation
- Refer BCS cases to expert centers
- **Treatment of Portal HTN -as recommended for cirrhosis**

[Ref: EASL Clinical Practice Guideline: Vascular diseases of the liver.2016]

Recommended stepwise therapeutic algorithm for BCS:

- **Medical Treatment**
- **Angioplasty/ Stenting/ Thrombolysis**
- **TIPSS**
- **Liver Transplantation**

[Ref: EASL Clinical Practice Guideline: Vascular diseases of the liver.2016]



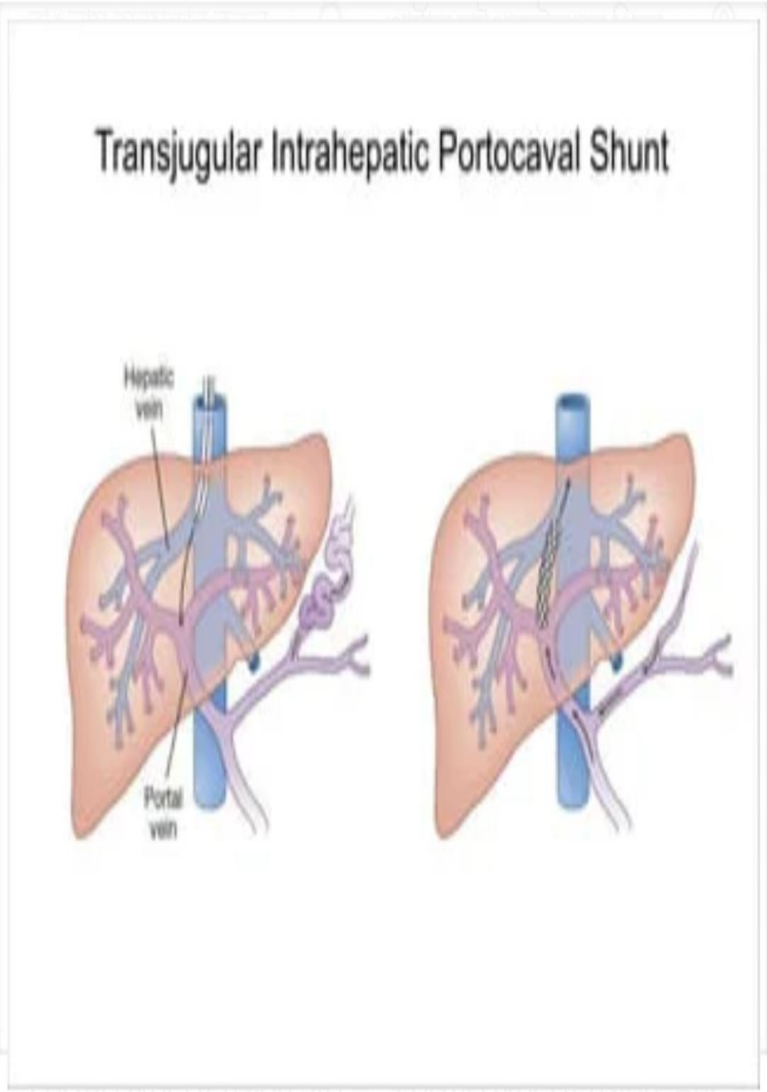
Recommendations for BCS

- **Treat underlying disease**.
- Treat all the patients with **anticoagulants,** in absence of major contraindications.
- Consider **angioplasty/ stenting as 1st line decompressive therapy**.

[Ref: EASL Clinical Practice Guideline: Vascular diseases of the liver.2016]

Recommendations for BCS

- If these fail- **TIPSS**
- **Propose liver transplantation as salvage treatment**



[Ref: EASL Clinical Practice Guideline: Vascular diseases of the liver.2016]

Algorithm for management of acute PVT

Abdominal pain+ Systemic inflammation +/ or thrombophilia

Confirm PVT by **unenhanced/ contrast CT scan**.

Screen for **general and local cause**

[Ref: EASL Clinical Practice Guideline: Vascular diseases of the liver.2016]

Algorithm for management of acute PVT

Low Molecular Weight Heparin

Antibiotics- if **sepsis**

1. **Persisting abdominal pain despite adequate anticoagulation**
2. **Organ failure**
3. **Rectal bleeding**

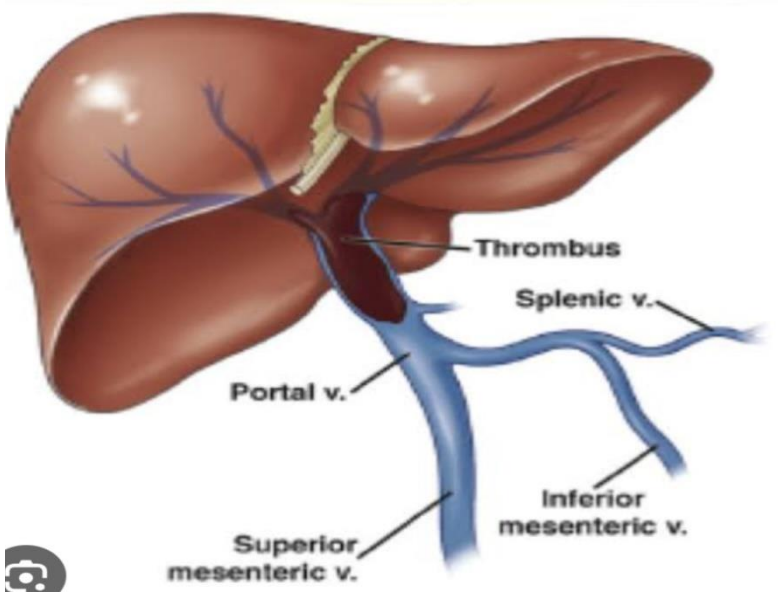
Algorithm for management of acute PVT

- If Yes - Urgent laparotomy

- If No -

  - Close monitoring

  - Anticoagulation for 6 months at least



Recommendations for INCPH:

- **Consider INCPH in any patient with PH if there is no other liver disease**

- **Exclusion of cirrhosis**

  and

  other causes of non-cirrhotic Portal Hypertension

- **Perform liver biopsy**

Recommendations for INCPH:

Manage PH **according to guideline for cirrhosis.**

Liver transplantation-

- **patient develops liver failure**

OR

- **unmanageable PH related complications**

[Ref: EASL Clinical Practice Guideline: Vascular diseases of the liver.2016]

NCPH :

- **Underestimated condition**
- Often difficult to diagnose

  due to-

  - **varied clinical presentation**
  - **unusual imaging finding**
  - **lack of consensus**
  - **Knowledge gap**

HOLISTIC APPROACH –needed.

THANK YOU