Porto-sinusoidal vascular disorder: When to suspect, how to diagnose

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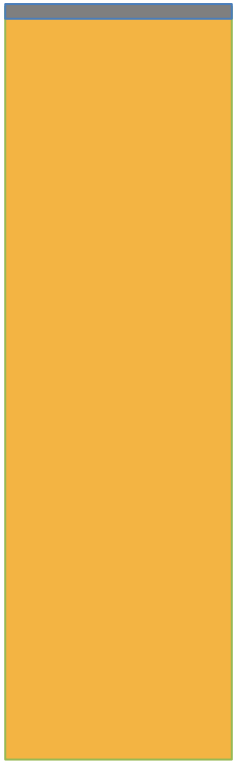
Porto-sinusoidal vascular disorder

- When to suspect?
- Definition

PSVD among cirrhosis

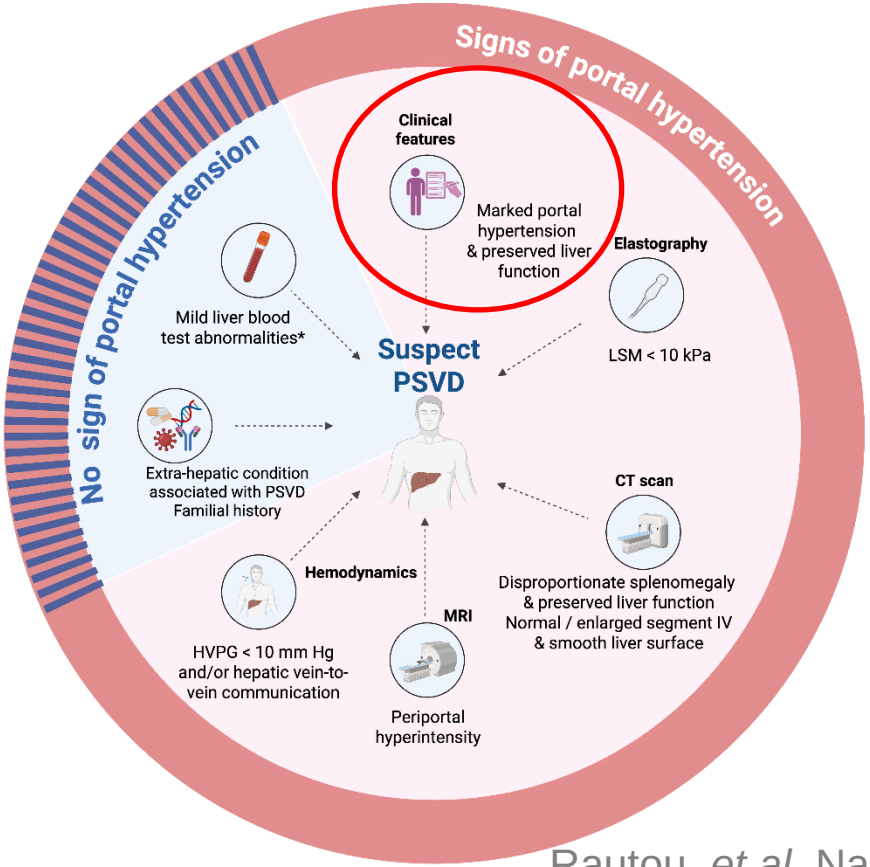


PSVD
0.4%

Cirrhosis
99.6%

➔ Screening tools needed

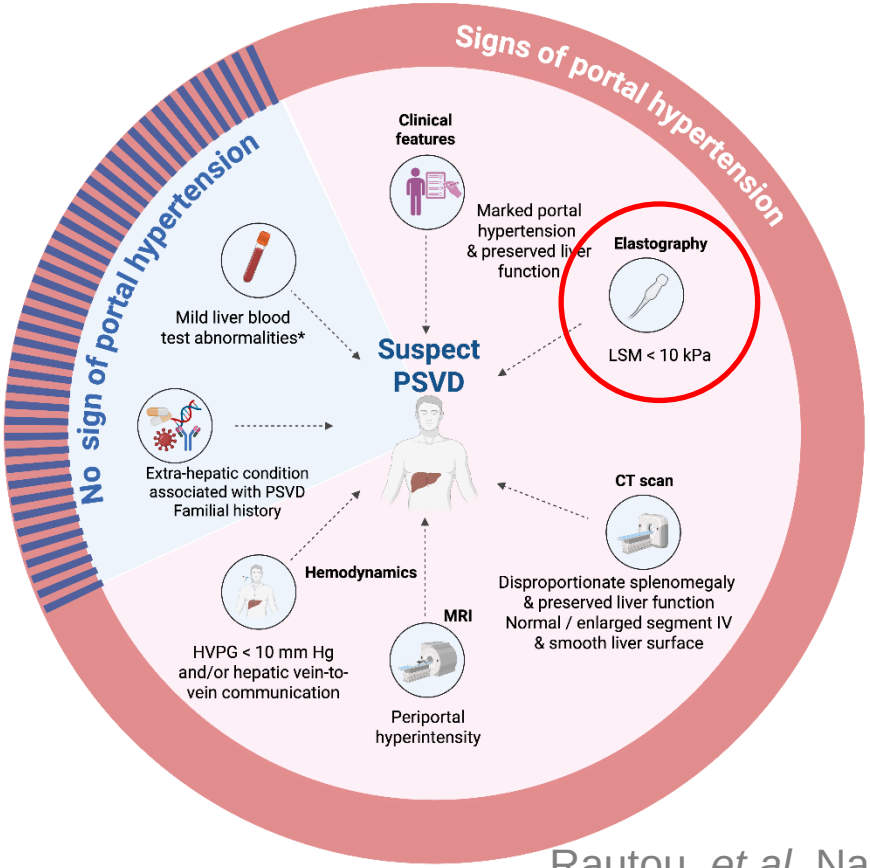
PSVD: when to suspect?



Paris PSVD meeting

Rautou, *et al.* Nat Rev Gastro Hepato 2026

PSVD: when to suspect?



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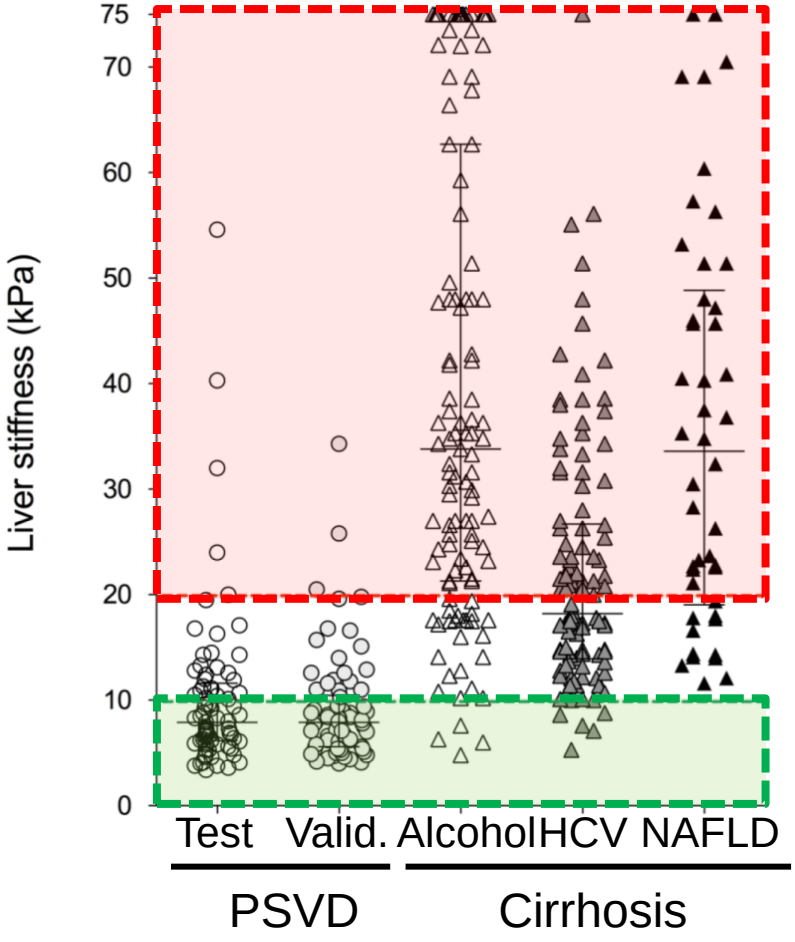
Rautou, *et al.* Nat Rev Gastro Hepato 2026

PSVD: liver stiffness measurement (TE)



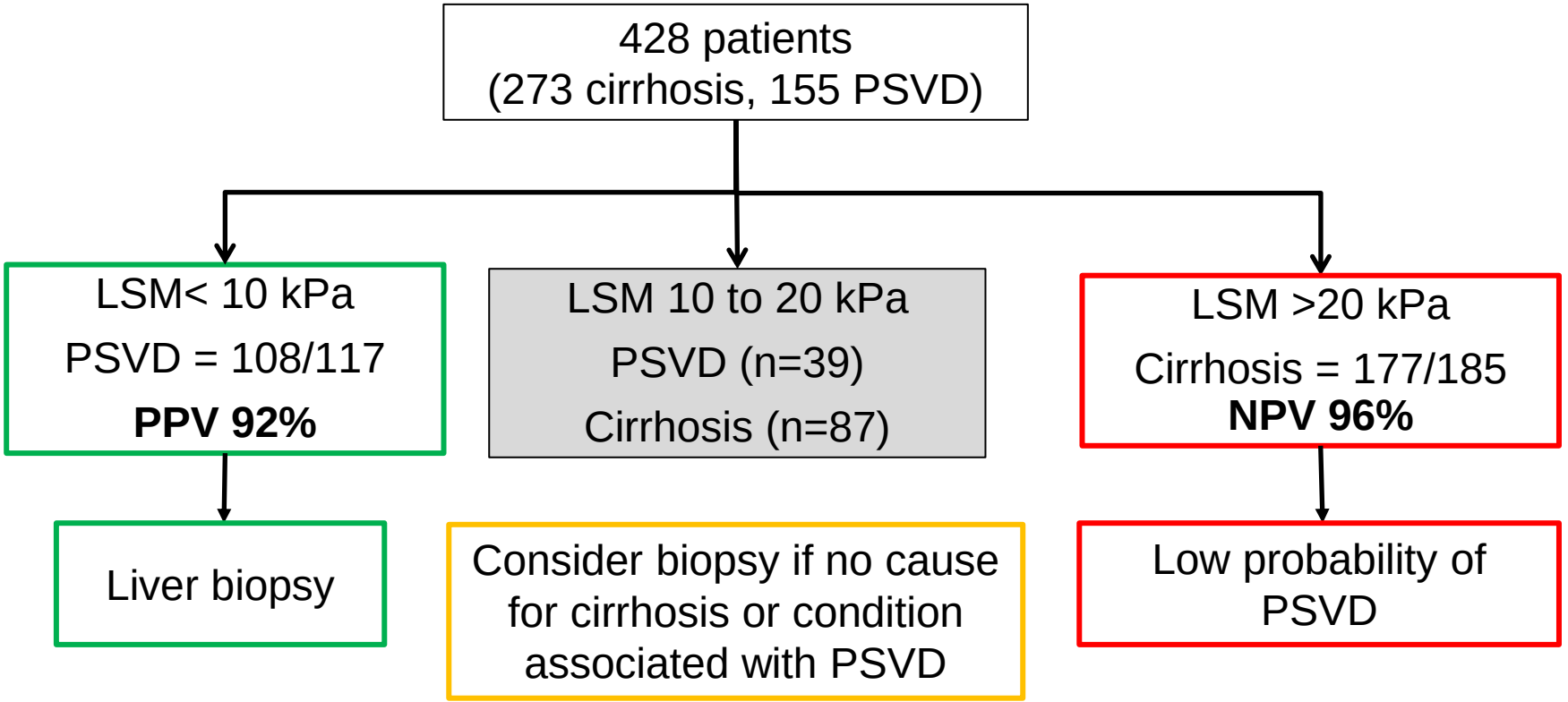
Patients with signs of portal hypertension
155 PSVD, 273 cirrhosis

Elkrief, Lazareth, *et al.*, Hepatology 2021

Similar results in:

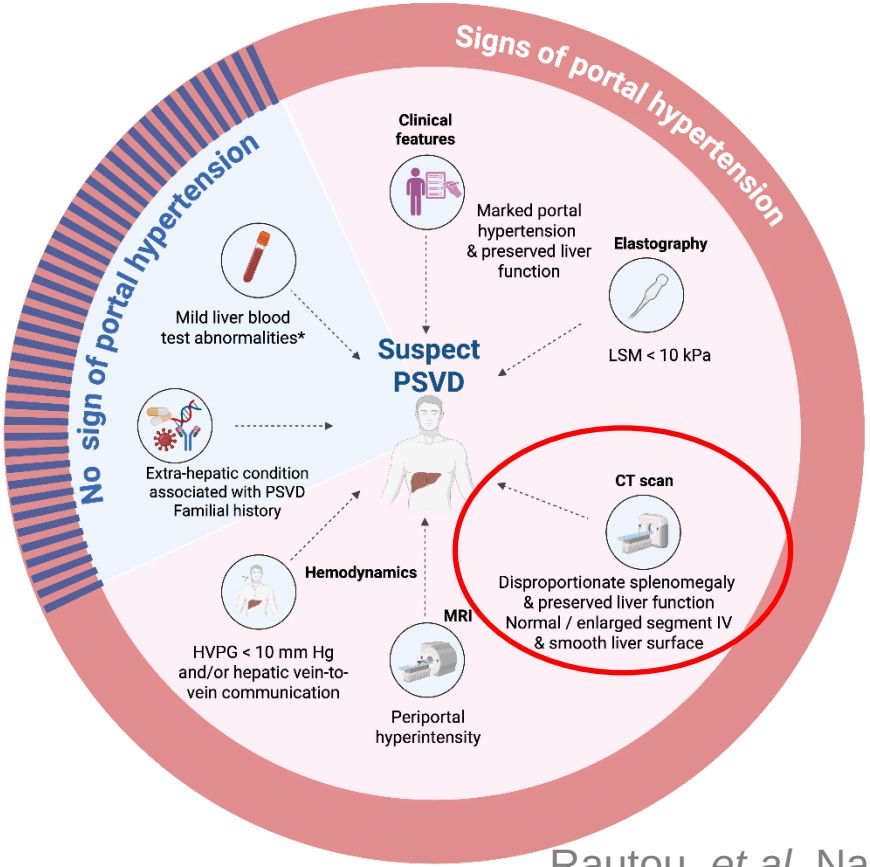
- Laharie Eur J Gastroenterol Hepatol 2010
- Seijo Dig Liver Dis 2012
- Sharma J Clin Exp Hepatol 2017
- Ferreira-Silva J, Dig Liver Dis. 2022

PSVD: liver stiffness measurement (TE)



Elkrief, Lazareth *et al.*, Hepatology 2021; EASL guidelines on vascular liver diseases 2026

PSVD: when to suspect?



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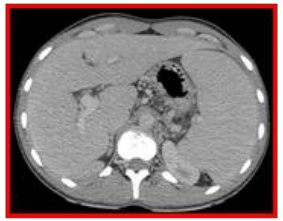
PSVD: CT-scan



50 patients with PSVD

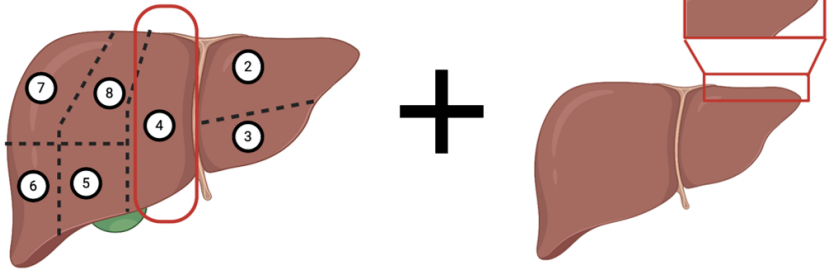
Matched on ascites

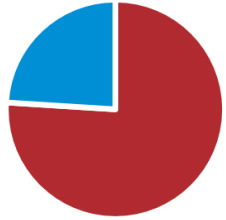
100 patients with cirrhosis

CT features for PSVD diagnosis

No atrophy of segment IV

LSN < 2.5





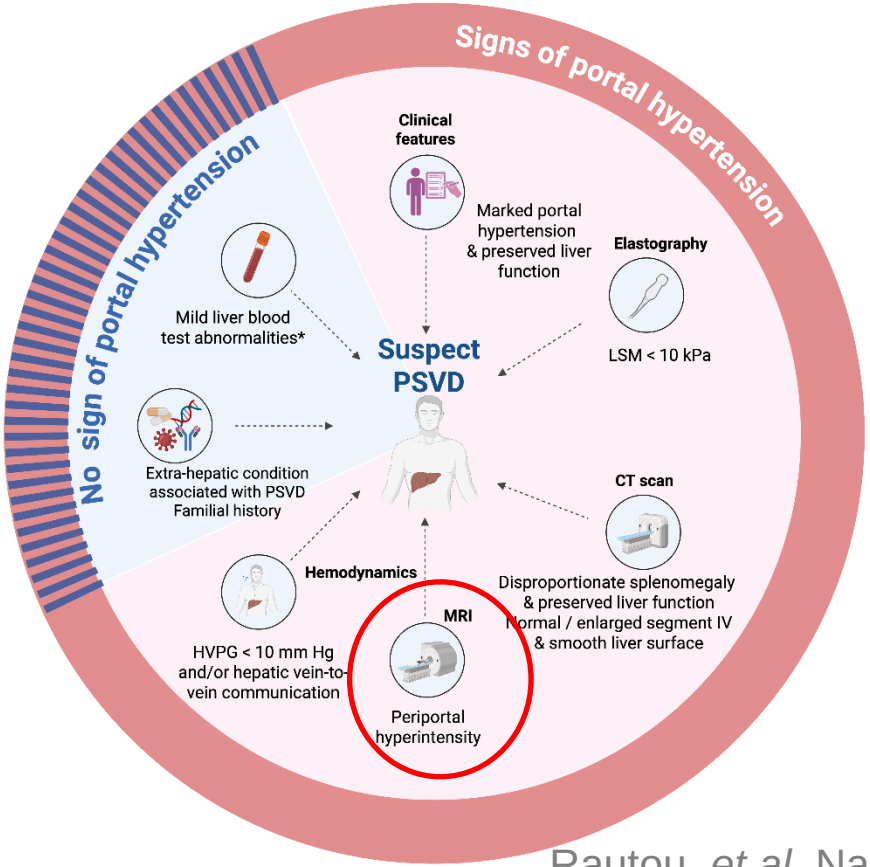
With the features

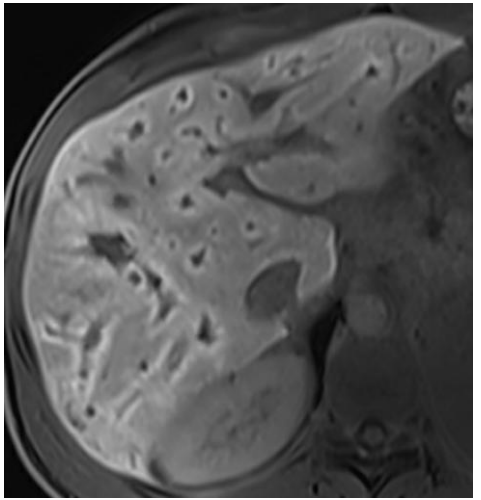
PSVD Cirrhosis

Spe > 90% for the diagnosis of PSVD
in patients with signs of portal hypertension

Valainathan *et al.* Hepatology 2022; Similar results in: Lampichler K *et al.* , Eur Radiol. 2022; Glatard *et al.* Radiology 2012; Kang, *et al.* Abdominal Radiology 2020

PSVD: when to suspect?



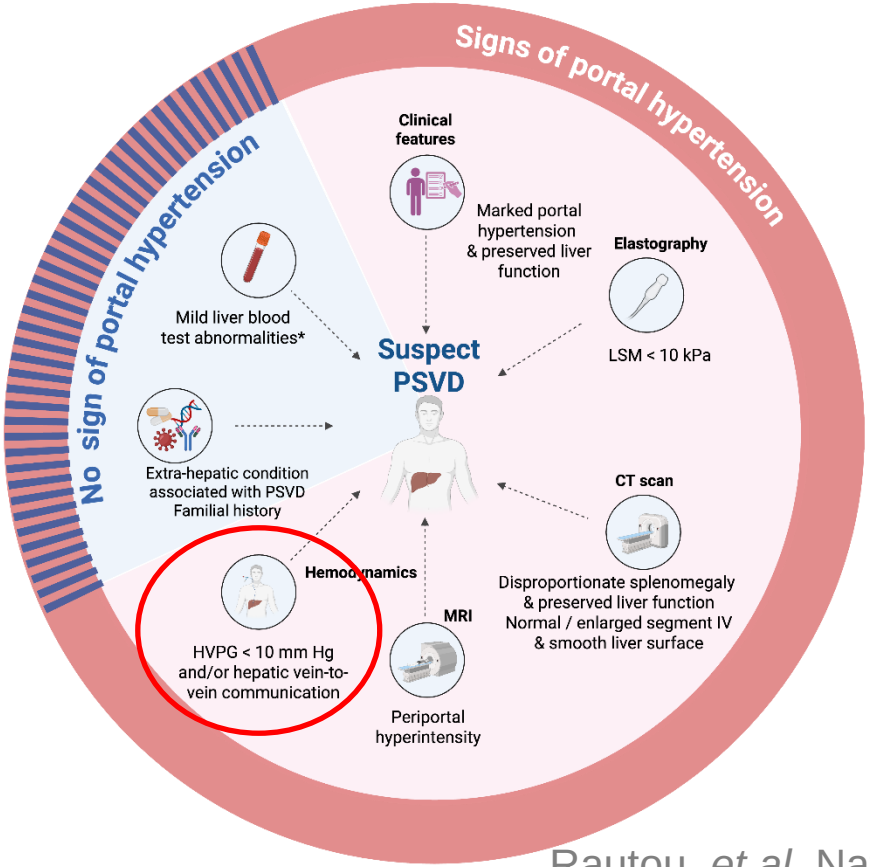


Periportal hyperintensity on hepatobiliary phase

Paris PSVD meeting

Rautou, *et al.* Nat Rev Gastro Hepato 2026

PSVD: when to suspect?



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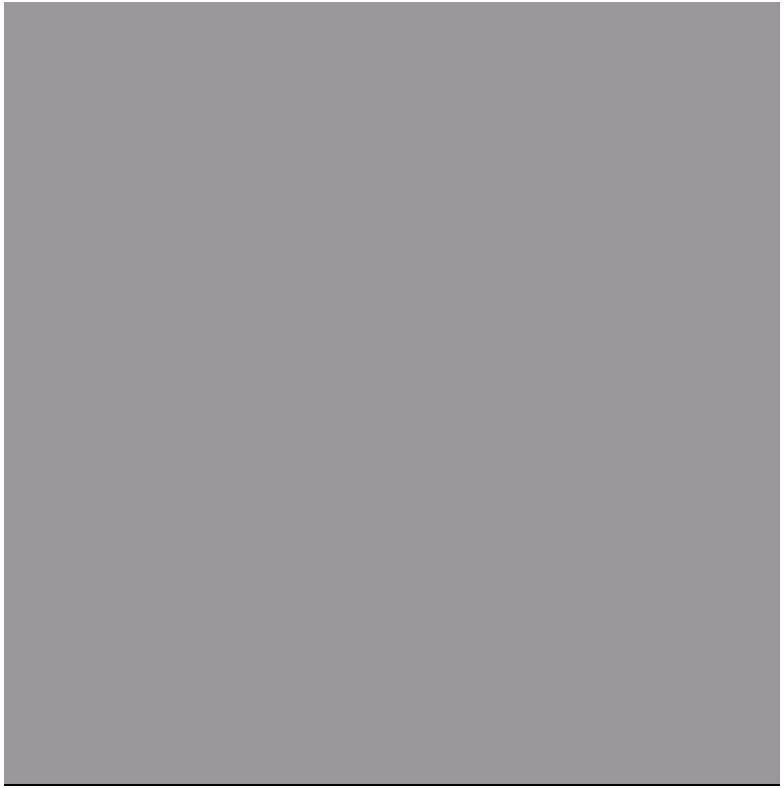
Hepatic veno-venous communications

Cirrhosis: 3%

PSVD: 30 to 50%

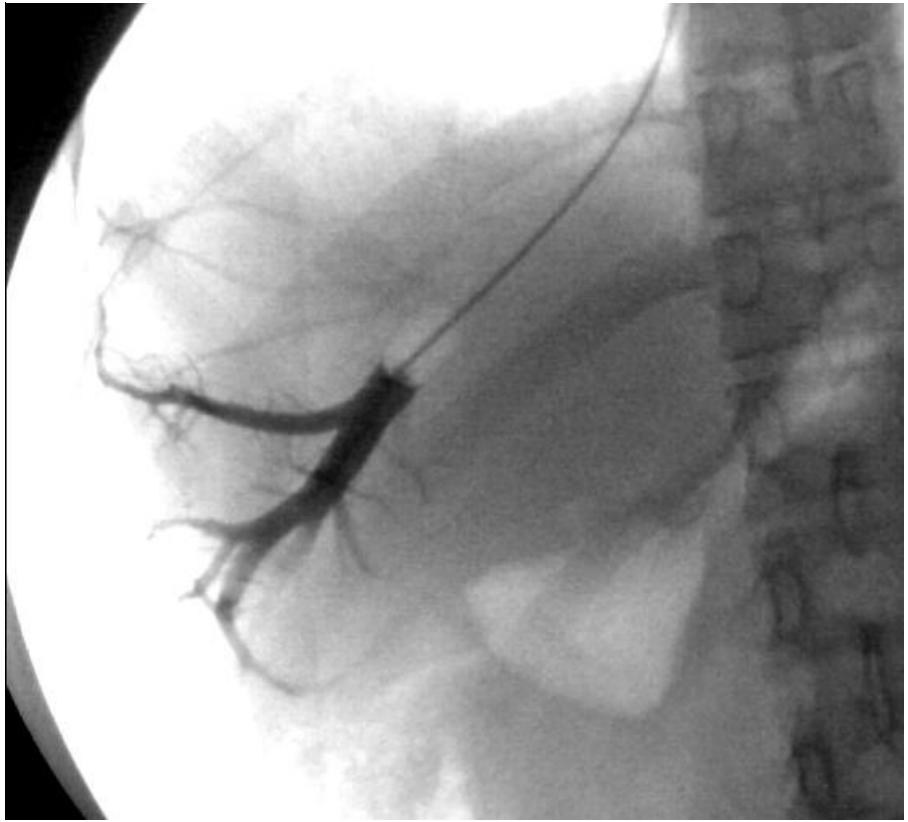


Drilhon et al; ILC 2020

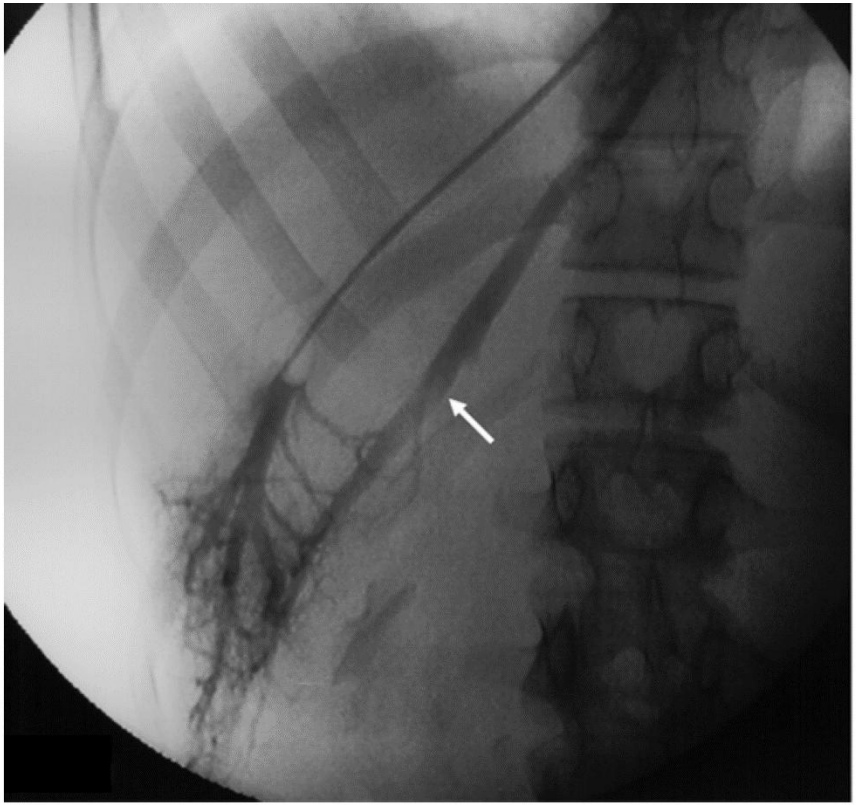
Seijo *et al.* Dig Liv Dis 2012

Hepatic veno-venous communications

Cirrhosis: 3%

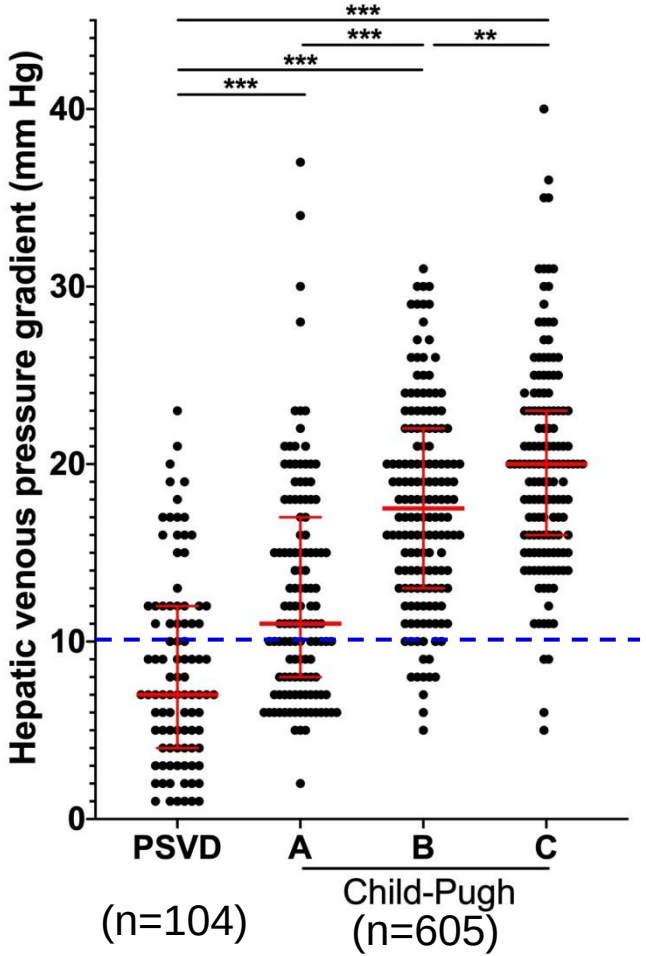


PSVD: 30 to 50%



Drilhon, ILC 2020; Seijo *et al.* Dig Liv Dis 2012

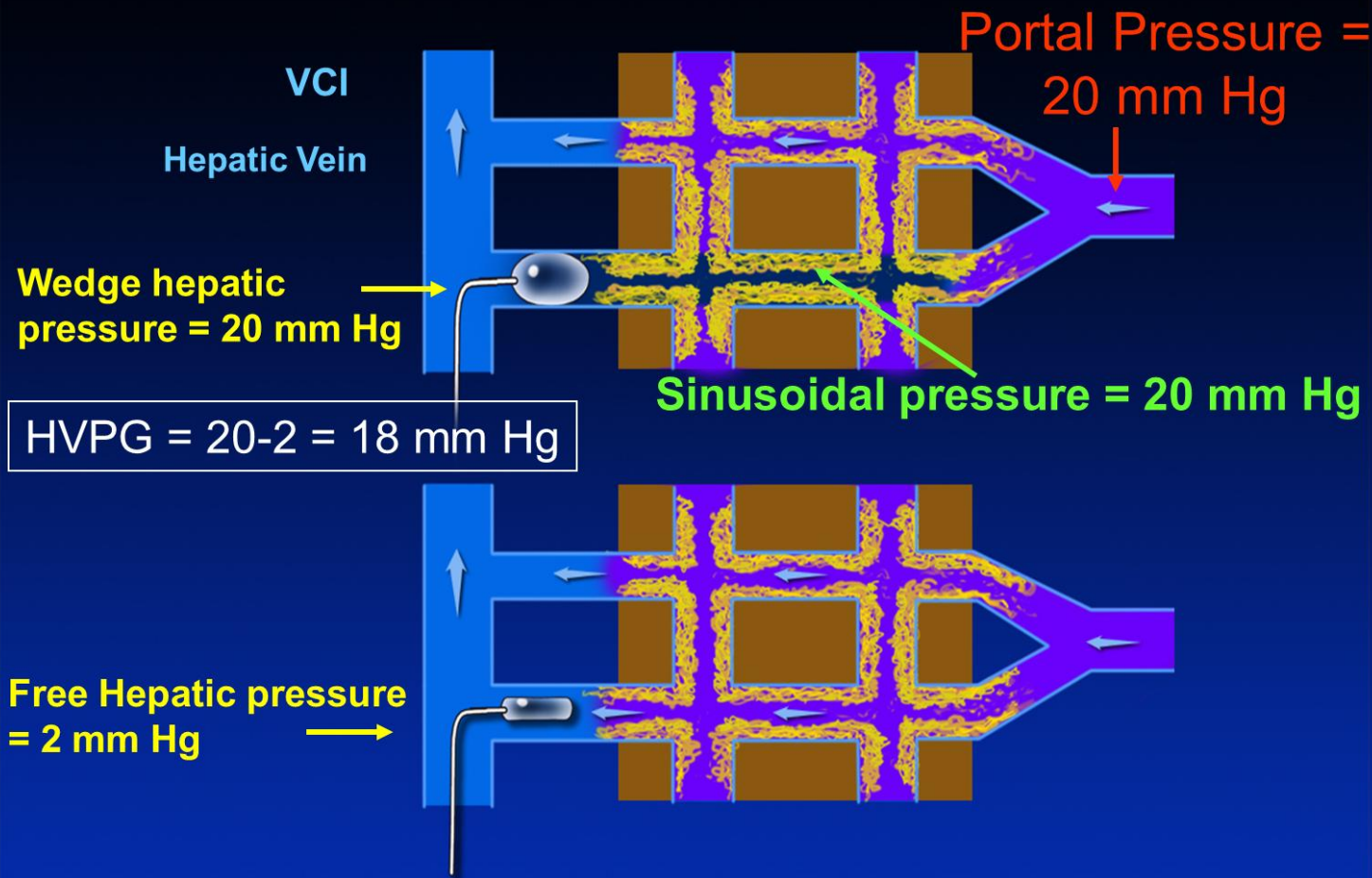
HVPG: PSVD vs. cirrhosis



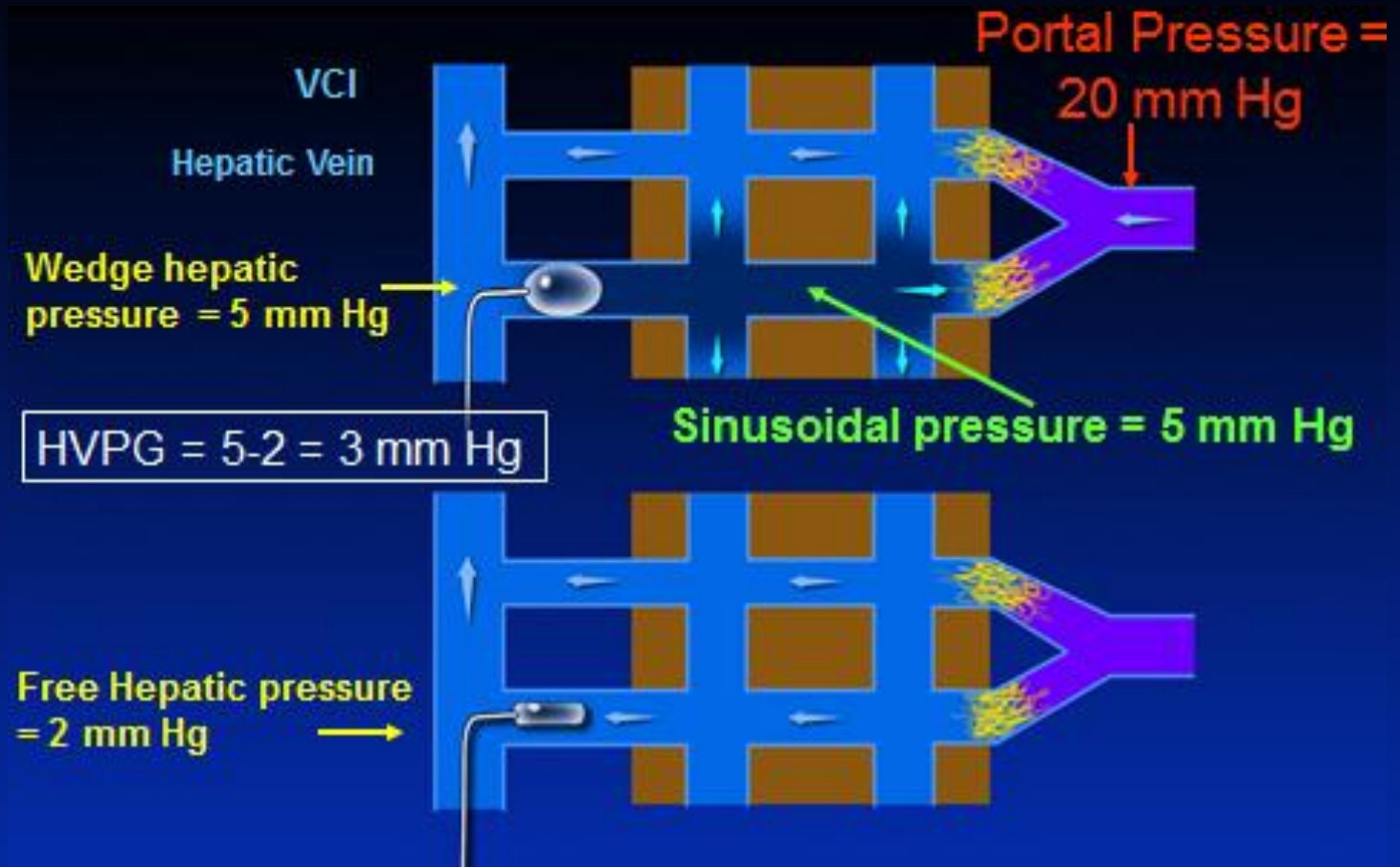
Patients with signs of portal hypertension

Drilhon *et al*, ILC 2020

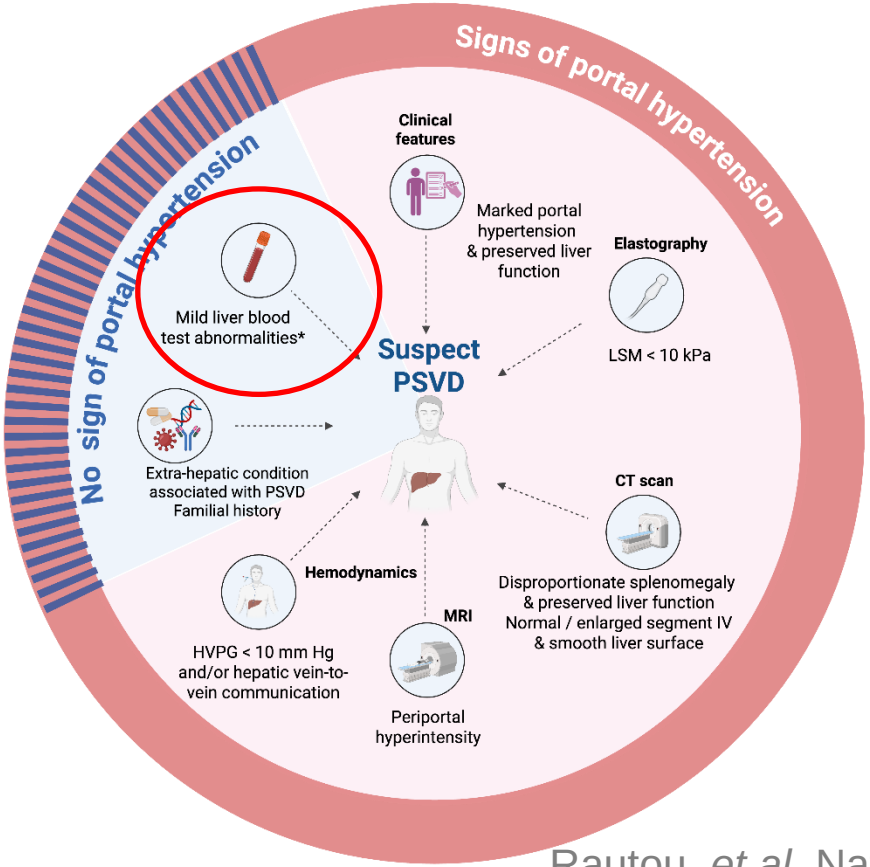
Hepatic hemodynamics: cirrhosis



Hepatic hemodynamics: PSVD



PSVD: when to suspect?



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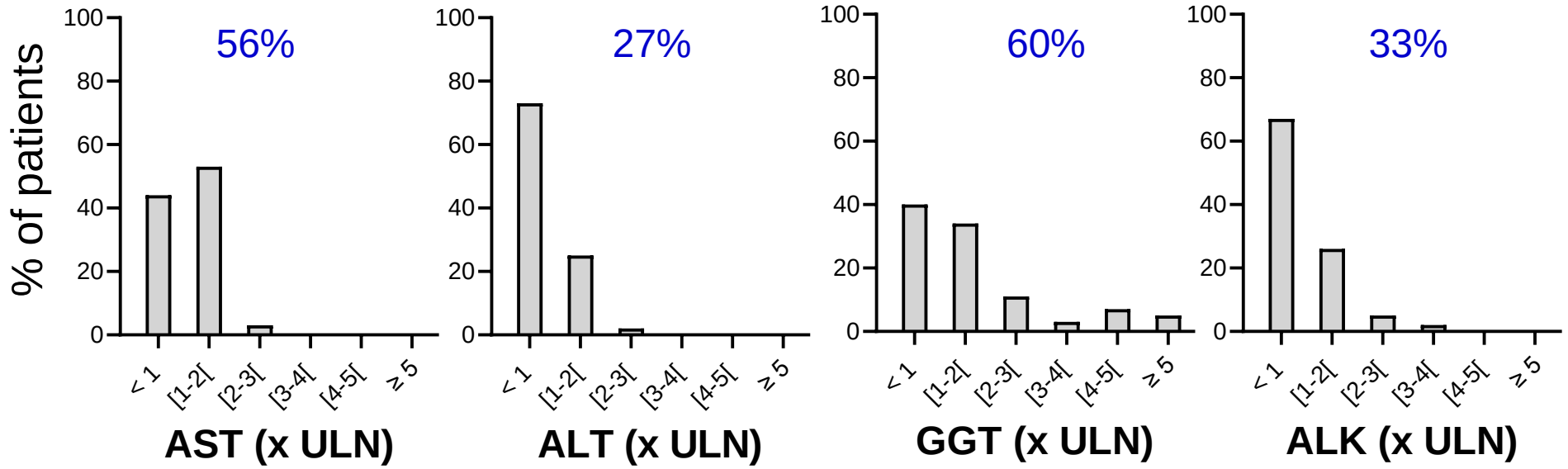
PSVD: liver blood test

Abnormal values:



Data at inclusion in APIS trial (NCT04007289)

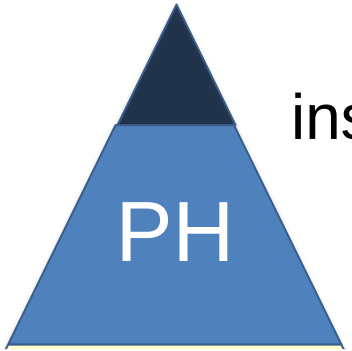
Similar in Magaz*, Giudicelli-Lett* *et al.* J Hepatol 2024

PSVD: manifestations

Abnormal liver blood tests (n=1271)

Guido, *et al.* Liv Int 2016; Barge, *et al.* Liv Int 2016;
Even higher in: Pugliese, Liv Int 2022, Pugliese J HEP REP 2024

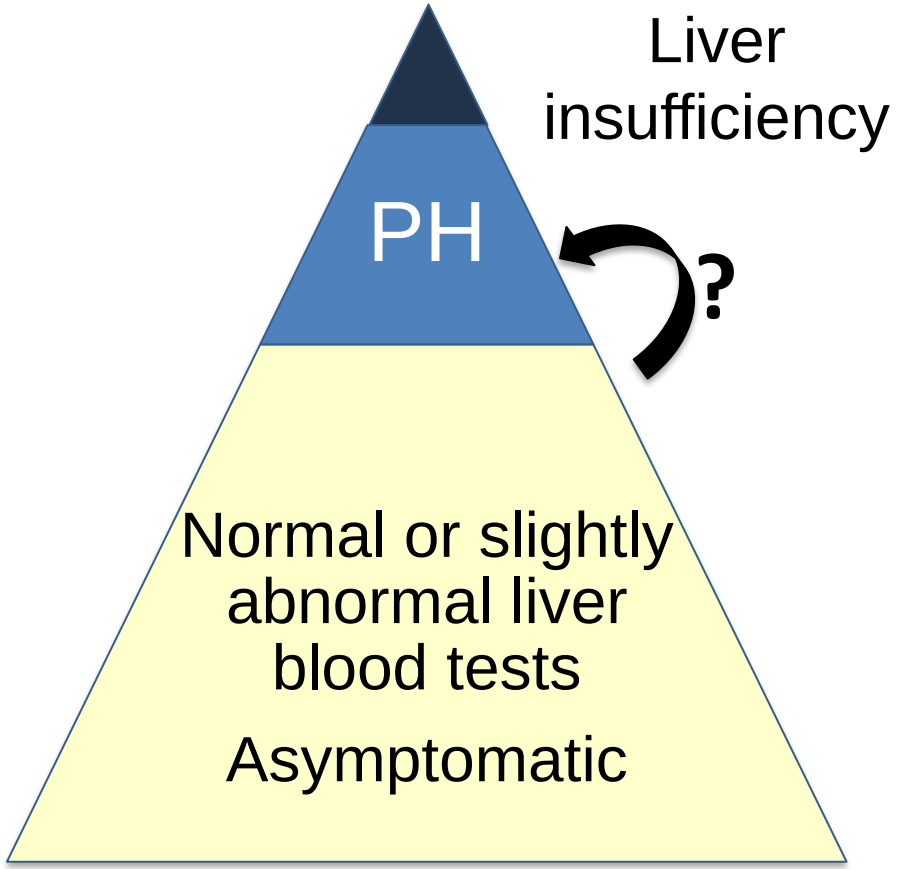
PSVD: manifestations



Liver insufficiency

Elkrief & Rautou, Liv Int 2016

PSVD: manifestations



Elkrief & Rautou, Liv Int 2016

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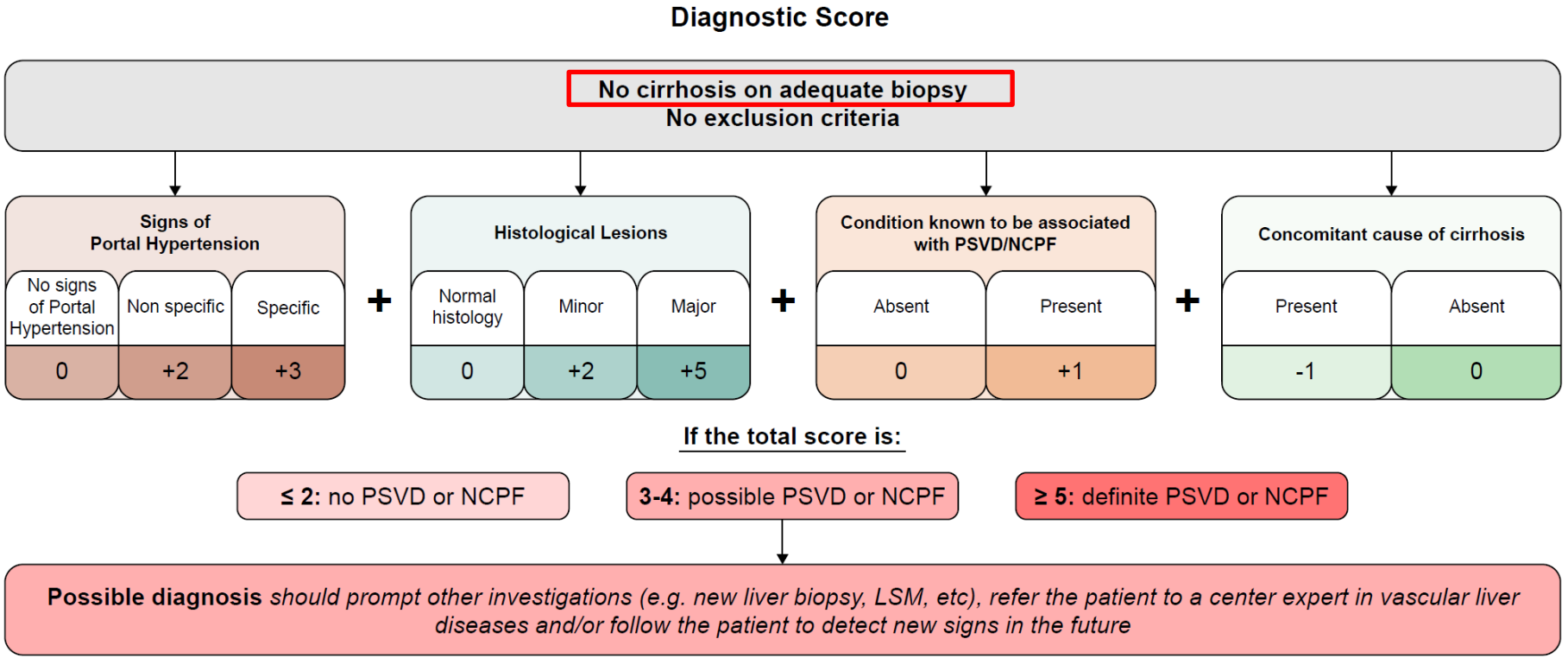
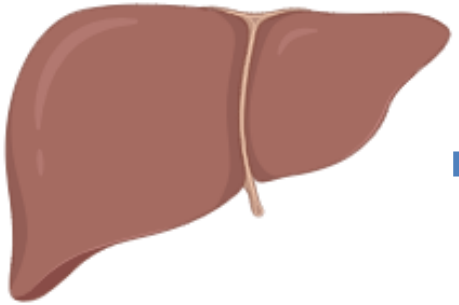
PSVD diagnostic criteria 2026



Hernandez-Gea, *et al,* J Hepatol 2026; Baveno VIII

PSVD: 15 mm is enough to rule out cirrhosis



24 explants:
- ✓ 12 PSVD
- ✓ 12 cirrhosis

36 000 virtual liver biopsies

Length:
5 mm, 10 mm, 15 mm, 20 mm, 25 mm, 5+10 mm, 5+5+5 mm

Staining:
Masson's trichrome
Picrosirius red

Width:
1 mm : percutaneous
572 µm : transjugular

Cirrhosis

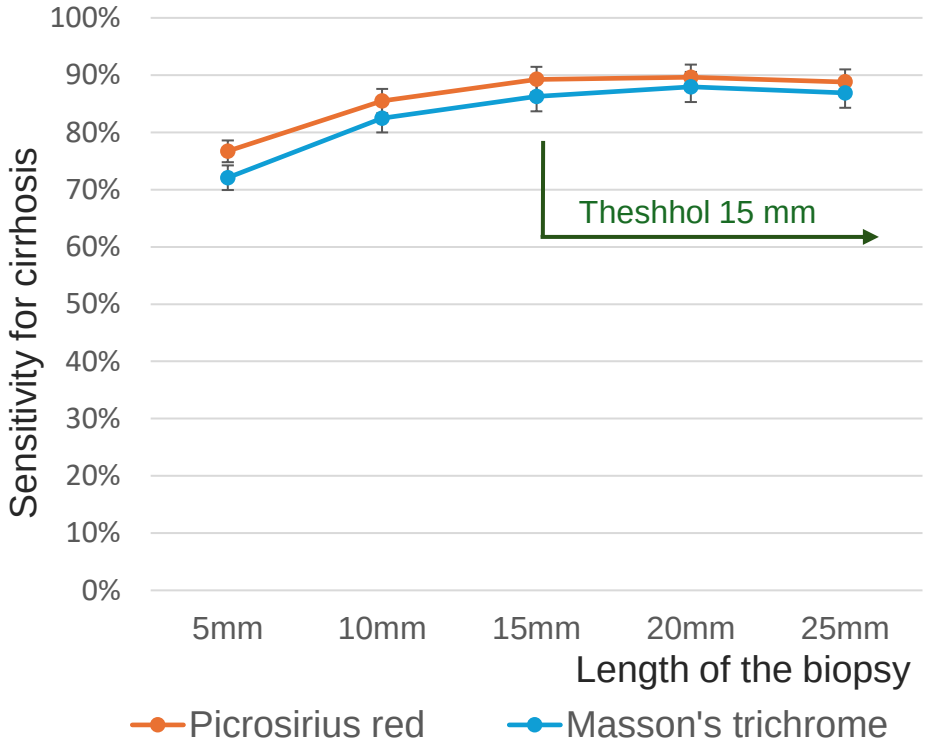
No cirrhosis

Each biopsy evaluated by an expert pathologist

Validation cohort: 2nd pathologist

PSVD: 15 mm is enough to rule out cirrhosis

Quality criteria for liver biopsy in cases of suspected PSVD

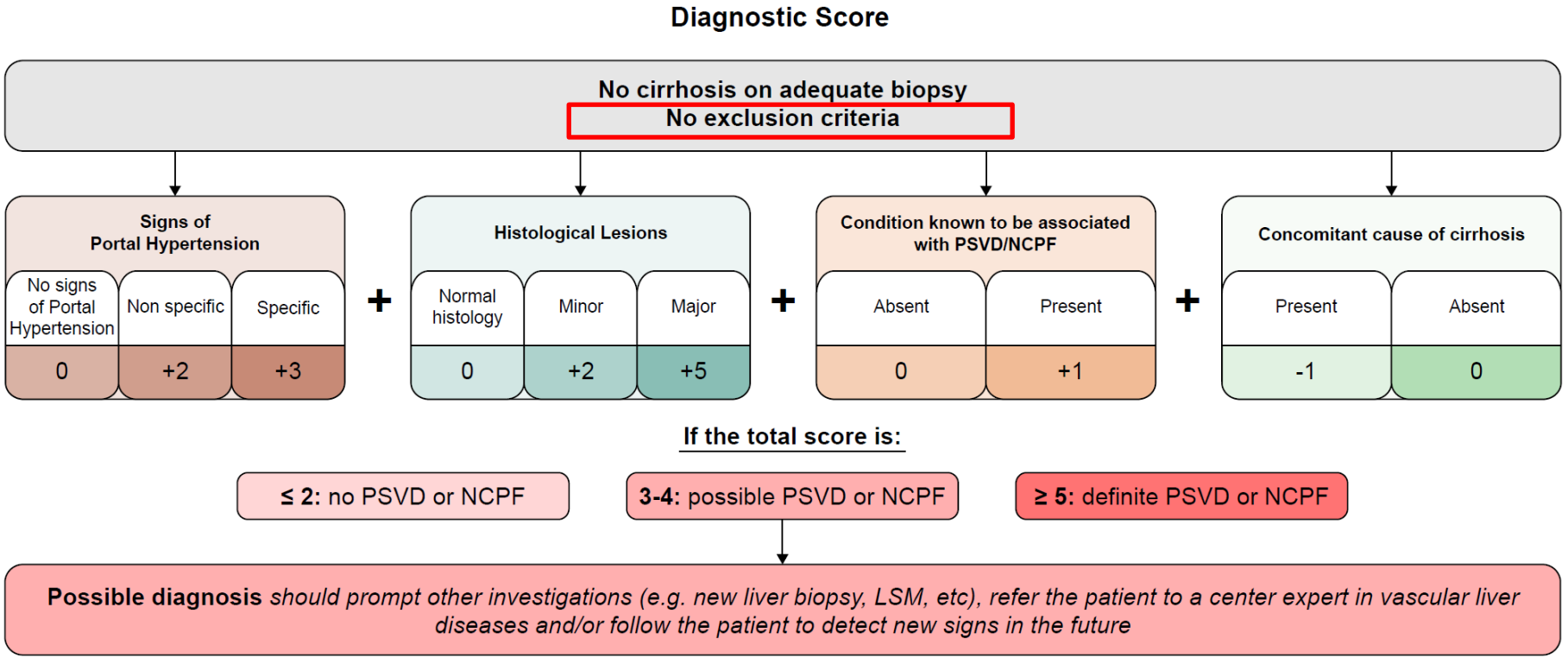


Sensitivity for cirrhosis: 89%

- plateau from 15 mm
- same with ≥1 fragment de 10 mm
- picrosirius red > Masson’s trichrome
- TJ route: sensitivity slightly < than percutaneous, but other information

De Broucker *et al.* JHEP REP *2025*

PSVD diagnostic criteria 2026



Hernandez-Gea, *et al,* J Hepatol 2026; Baveno VIII

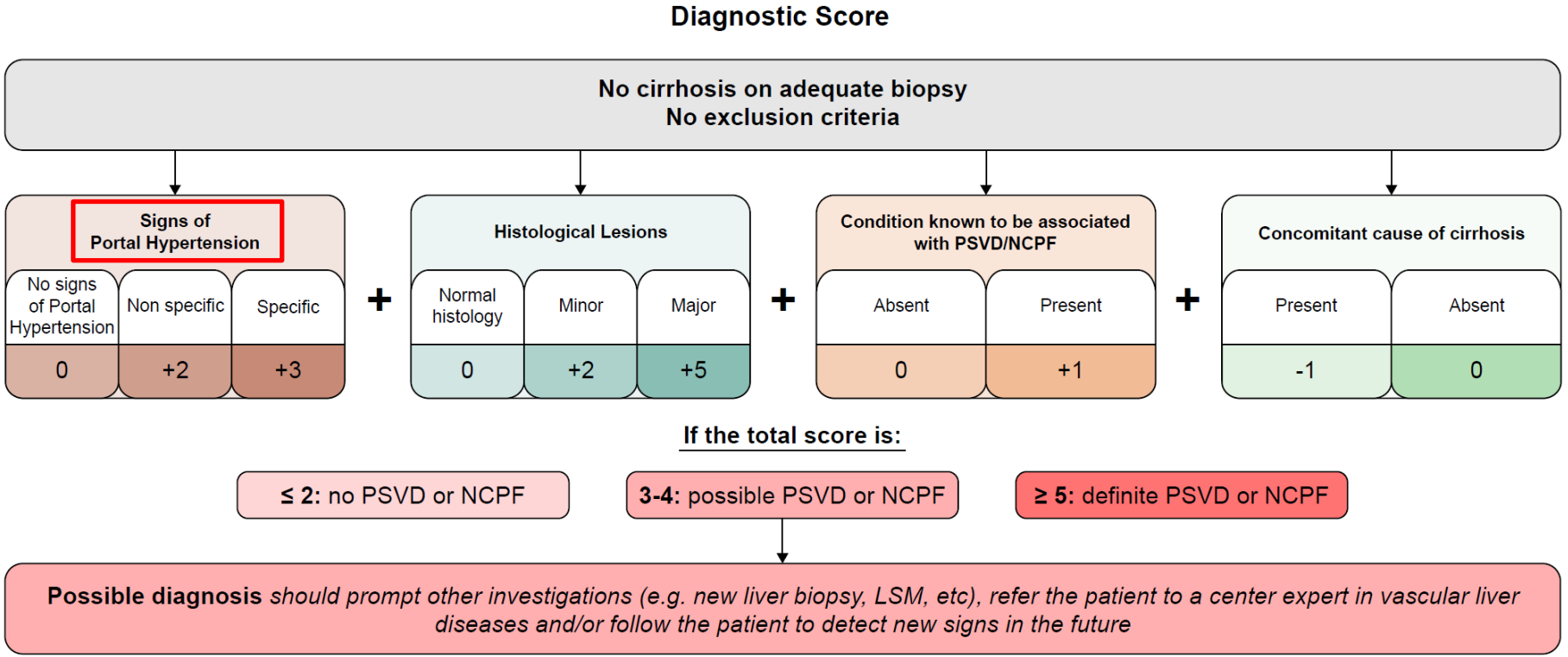
Exclusion criteria

	Exclusion Criteria
1	Abernethy syndrome
2	Budd Chiari syndrome or hepatic venous outflow obstruction
3	Cardiac failure or Fontan Associated Liver Disease
4	Cholestatic liver diseases
5	Congenital hepatic fibrosis
6	Gaucher disease
7	Hepatic amyloidosis
8	Hereditary hemorrhagic telagiectasia
9	Hypervitaminosis A
10	Intra-hepatic arteriovenous shunts
11	Liver infiltration by tumor cells
12	Peliosis hepatis
13	Schistosomiasis**
14	SOS in the last 6 months*

Not excluded anymore:

- portal cavernoma
- history of bone marrow transplant

PSVD diagnostic criteria 2026



Hernandez-Gea, *et al,* J Hepatol 2026; Baveno VIII

PSVD: signs of portal hypertension

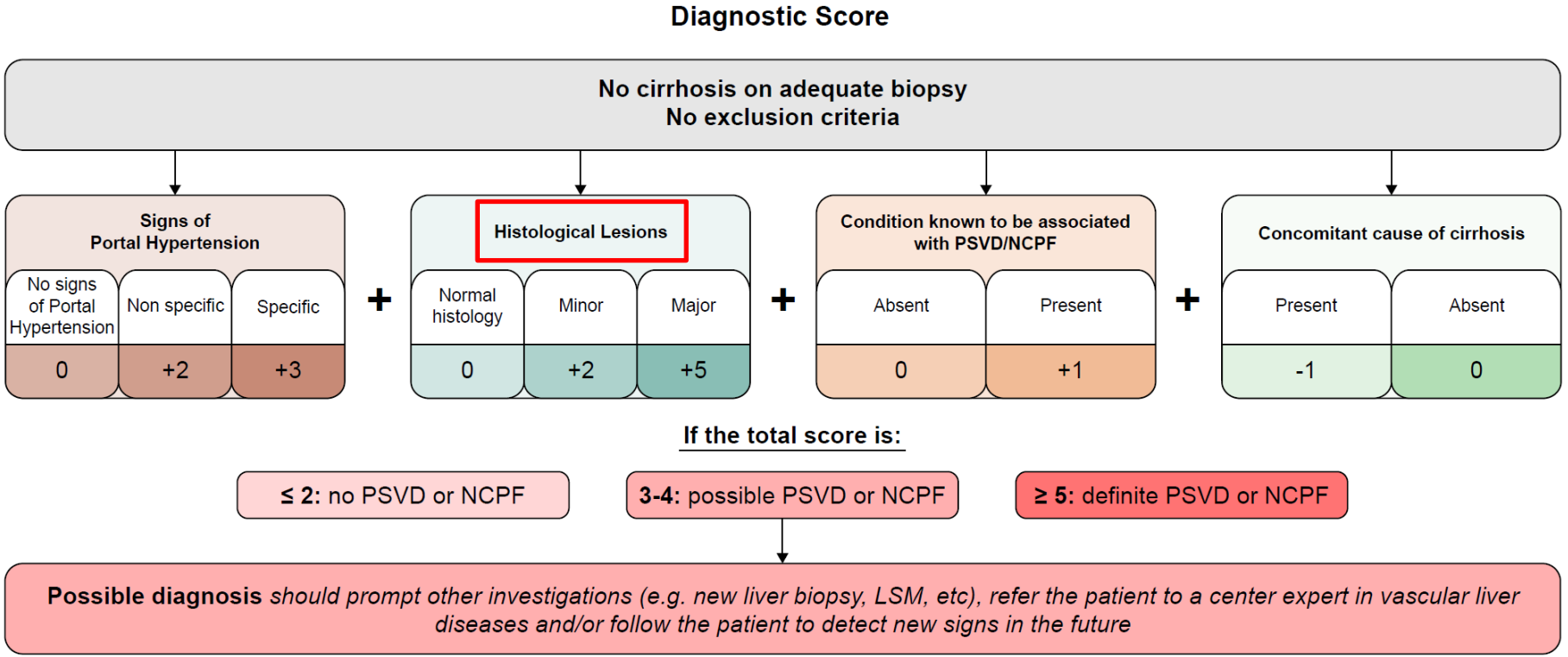
Specific signs of portal hypertension:

- ✓ Gastrointestinal bleeding due to varices
- ✓ Medium / large oesophageal varices
- ✓ Gastric varices
- ✓ Portosystemic shunt
- ✓ Spleen stiffness > 40 kPa in the absence of MPN

Non specific signs of portal hypertension

- ✓ Clinical Ascites
- ✓ Small Varices
- ✓ Low platelet count
- ✓ Splenomegaly

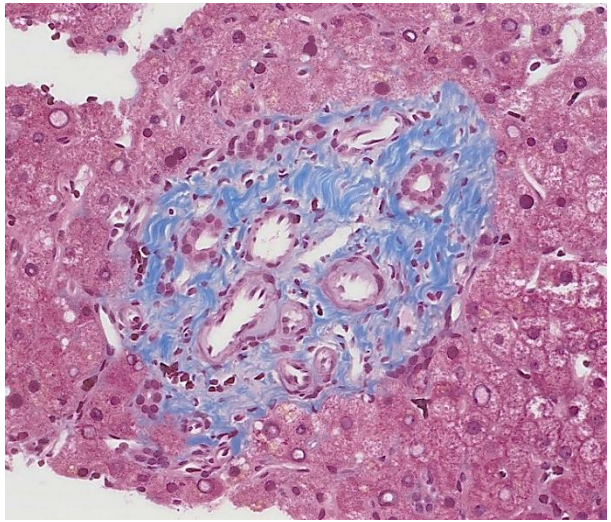
PSVD diagnostic criteria 2026



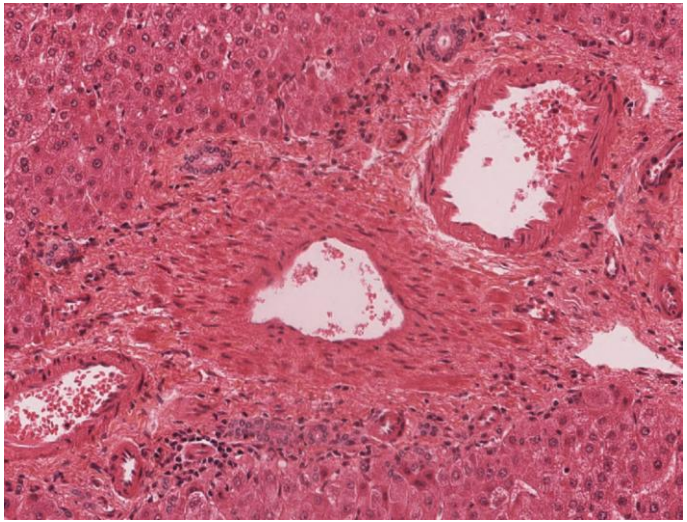
Hernandez-Gea, *et al,* J Hepatol 2026; Baveno VIII

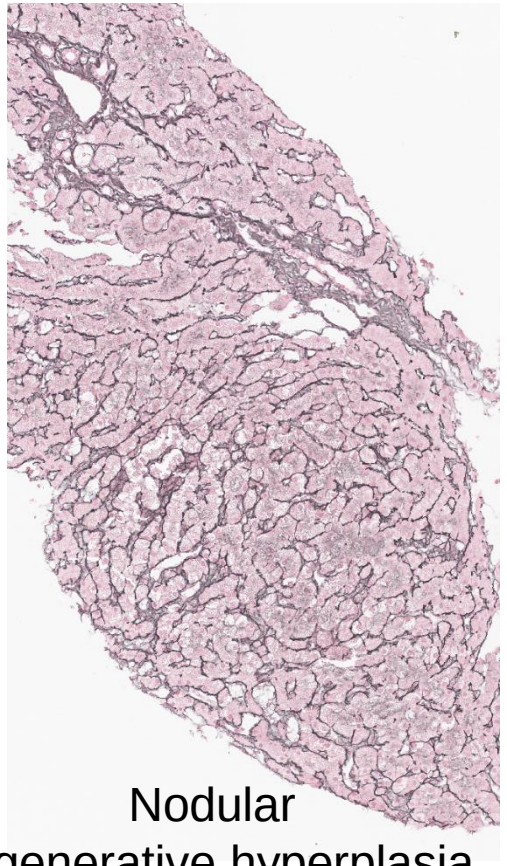
PSVD: major criteria

PV stenosis ≥ 50% of portal tracts



Muscularized portal vein

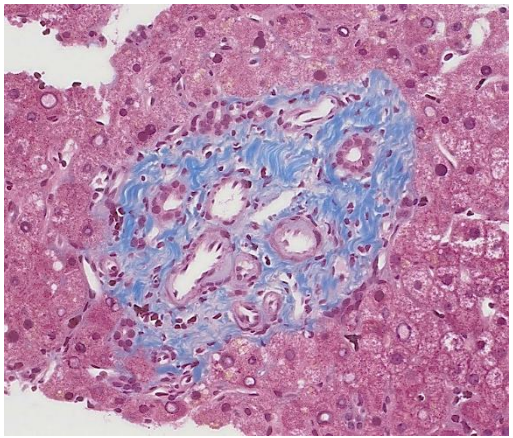




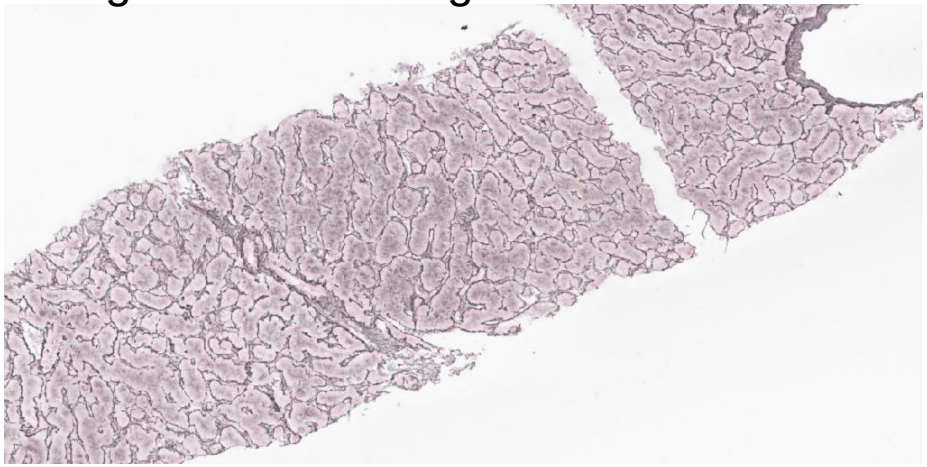
Nodular regenerative hyperplasia

PSVD: minor criteria

PV stenosis 25-49% of portal tracts



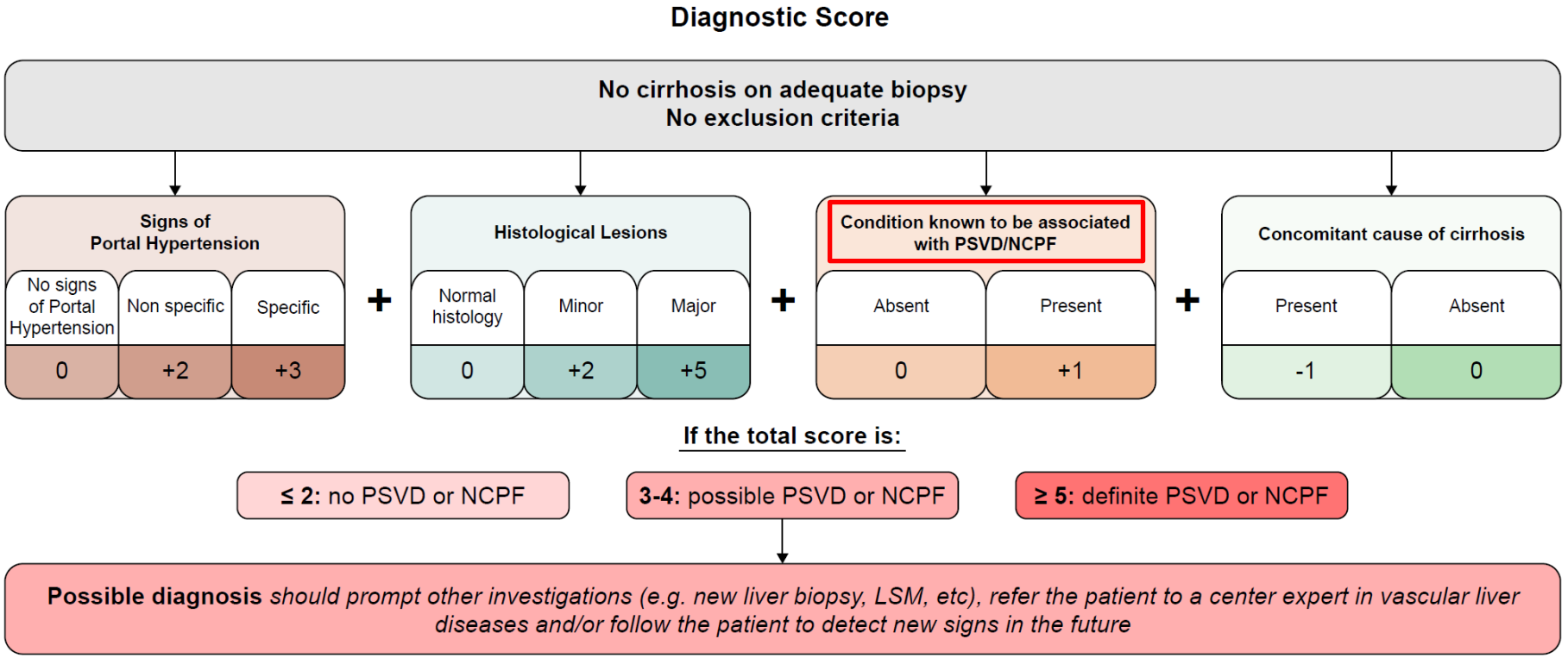
Regenerative changes without obvious NRH



Abnormal distribution of vascular structures

All 3 minor criteria = 1 major

PSVD diagnostic criteria 2026



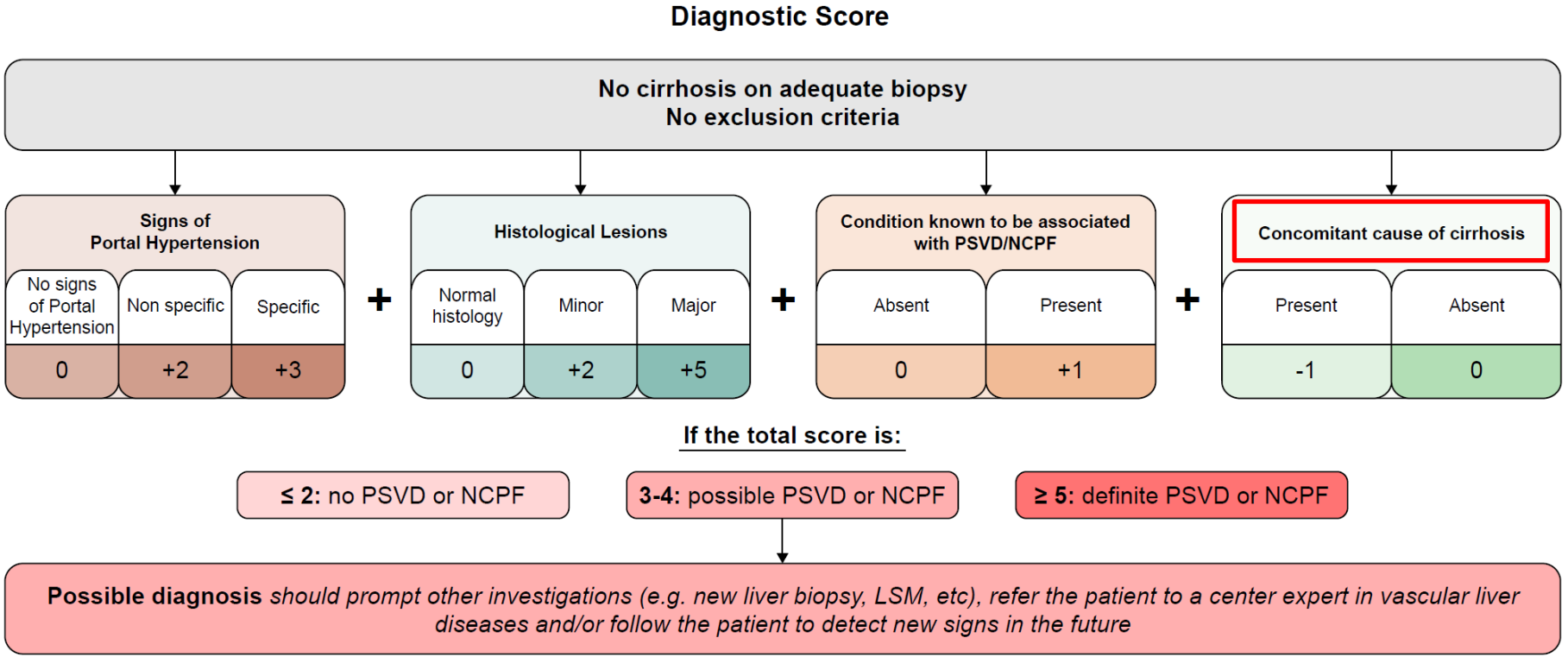
Hernandez-Gea, *et al,* J Hepatol 2026; Baveno VIII

Conditions associated with PSVD (>50% vote)

	n = 587
None identified	**32 %**
Immunological disorder • Common variable immune deficiecy commun variable • IBD • Others	**32 %** 7 % 6 % 19 %
Blood disorder • Myeloproliferative disorder • Other	**9 %** 3 % 6%
HIV infection	**8 %**
Drug exposure • Azathioprine • Oxaliplatin	**15 %** 8 % 7 %
Prothrombotic state	**9 %**
Familial / genetic	**4%**

Magaz *et al.*,
J Hepatol 2025

PSVD diagnostic criteria 2026



Hernandez-Gea, *et al,* J Hepatol 2026; Baveno VIII

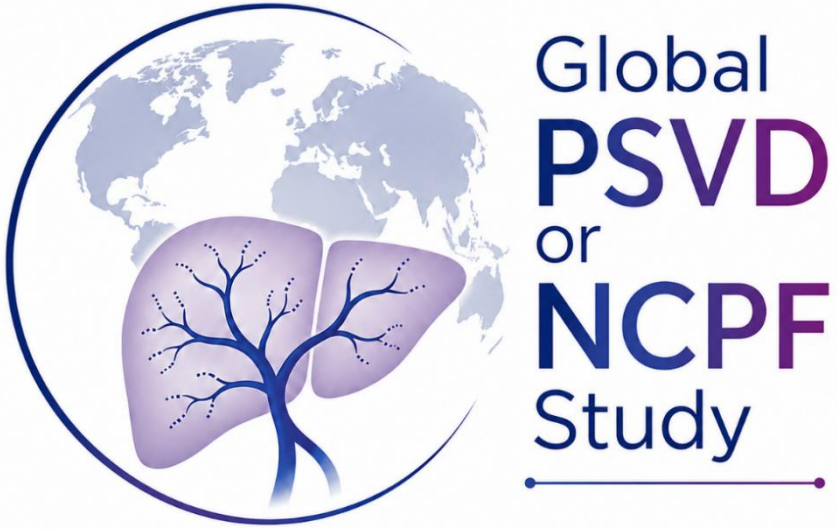
Conclusion & Next step

Summary

✓ **Suspect PSVD:**
- Unexplained liver blood test abnormalities
- No liver insufficiency
- Low liver stiffness
- Low HVPG, hepatic collaterals
- Normal or ↑ segment IV + smooth surface

✓ **Diagnosis:**
- Liver biopsy
- New diagnostic score

Global vision



Objective: understand diversity of PSVD worldwide

COST ACTION

European vascular liver diseases network (EURO-VALDI-NET)

6 working groups

Start Date: 30/10/2024

End Date: 29/10/2028

Pierre-Emmanuel Rautou (Chair)
Virginia Hernandez-Gea (Vice-Chair)
Laure Elkrief (Grant holder scientific representative)
Sarwa Darwish-Murad (Grant Awarding Coordinator)
Filipe Andrade (Science Communication Coordinator

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Reference center for vascular liver diseases

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