Porto-sinusoidal vascular disorder

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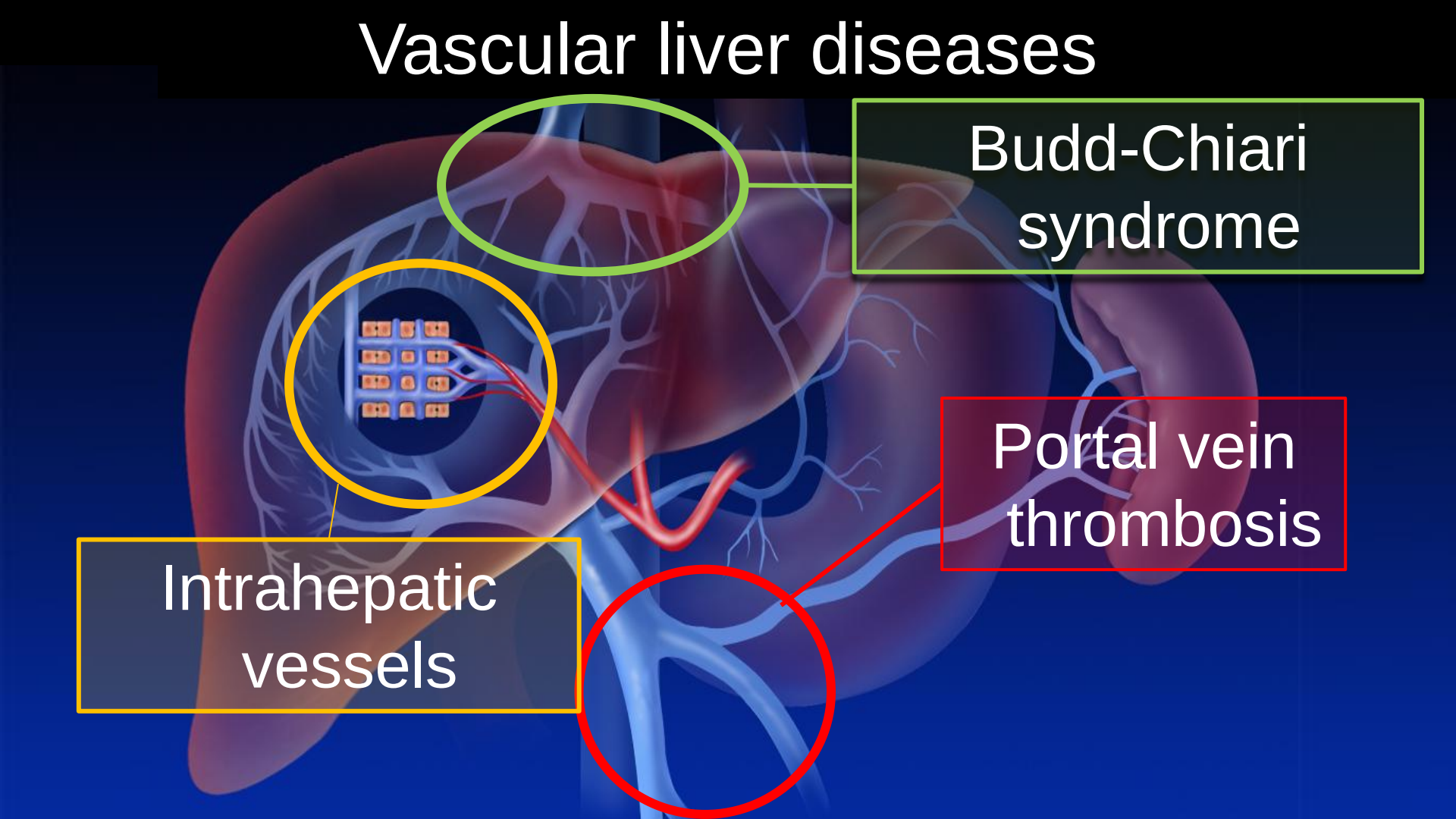
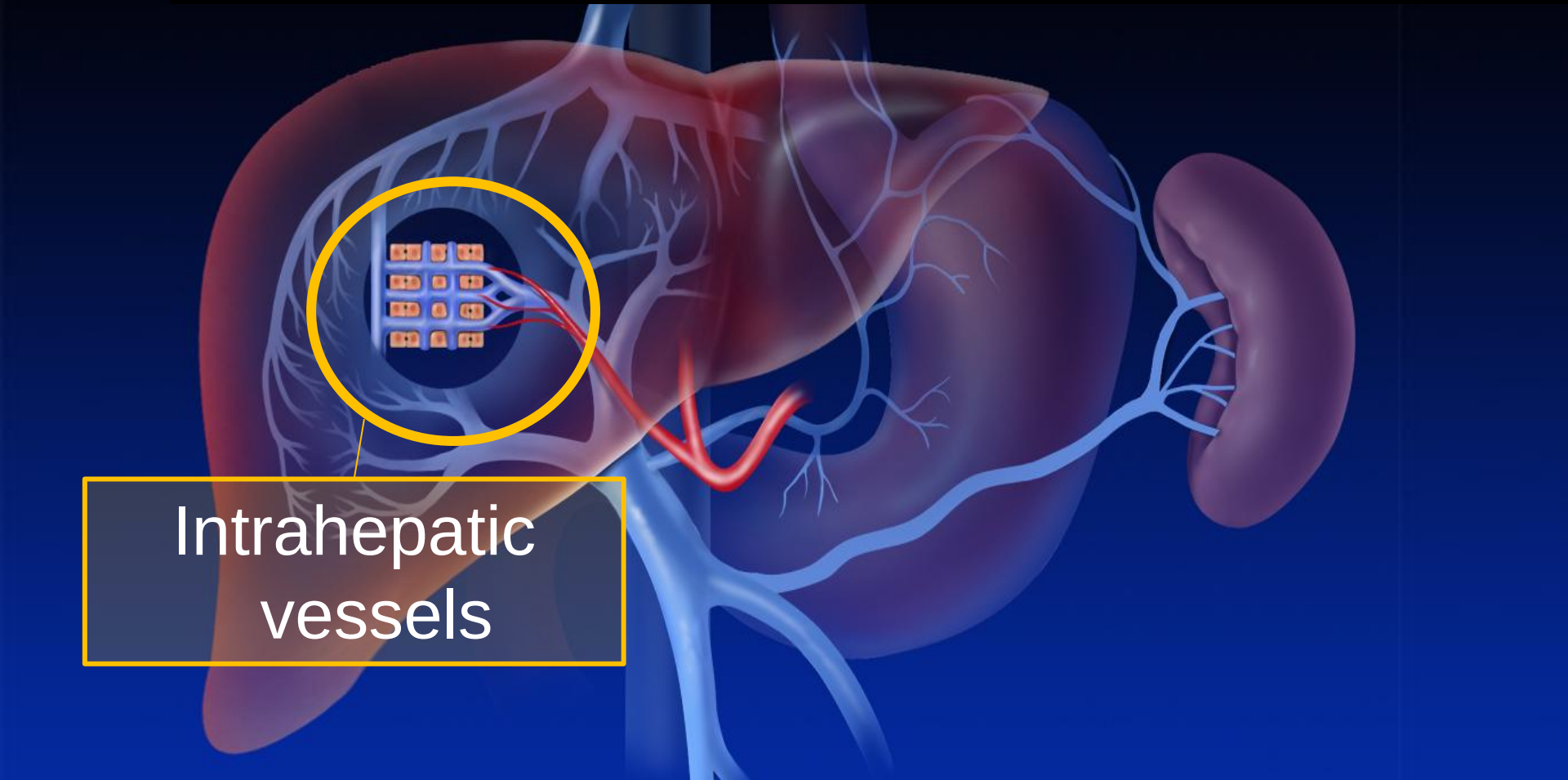
Vascular liver diseases



Vascular liver diseases

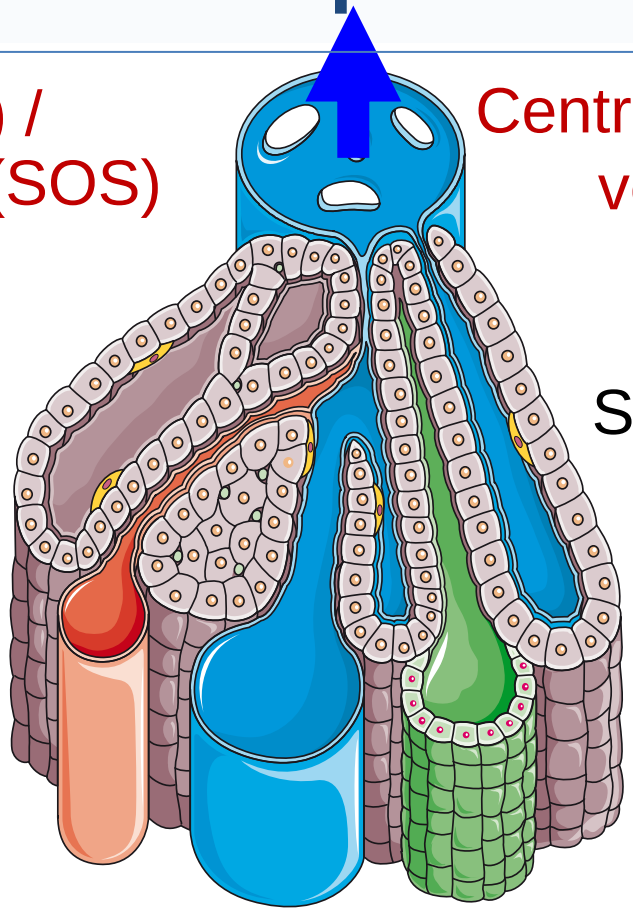


Diseases affecting small intrahepatic vessels

Veno-oclusive disease (VOD) / sinusoidal obstruction syndrome (SOS)

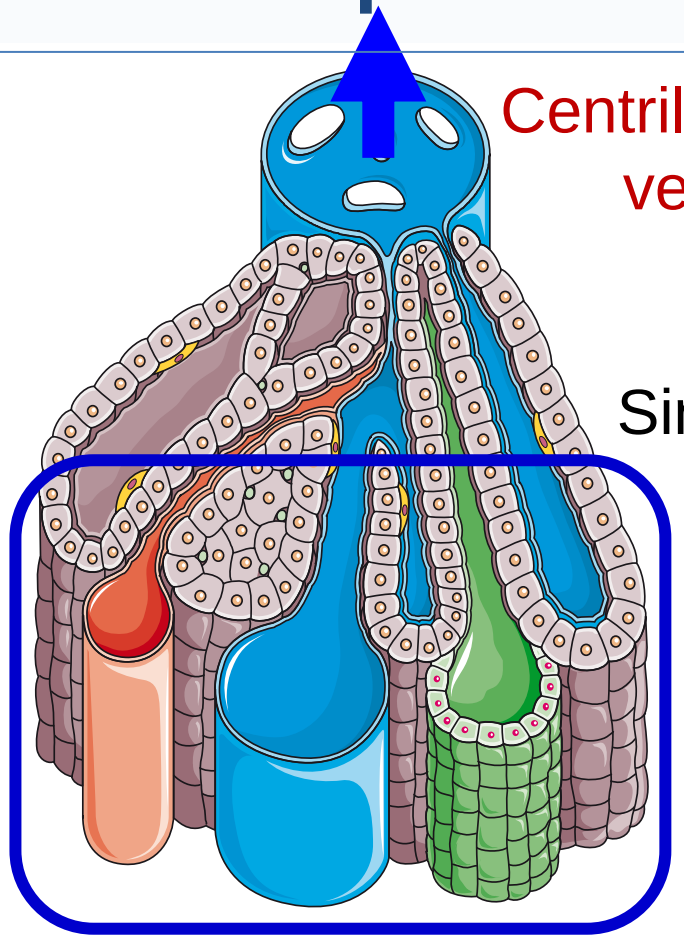
Sinusoidal dilation / Peliosis

Portosinusoidal vascular disorder (PSVD)



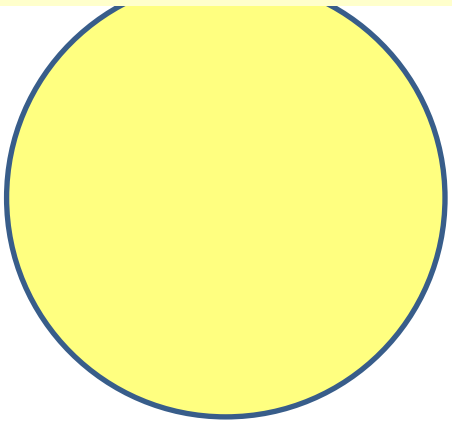
Diseases affecting small intrahepatic vessels

Portosinusoidal vascular disorder (PSVD)



HISTOLOGY

- Obliterative portal venopathy
- Nodular regenerative hyperplasia
- Hepatoportal sclerosis
- Non cirrhotic portal fibrosis



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HISTOLOGY

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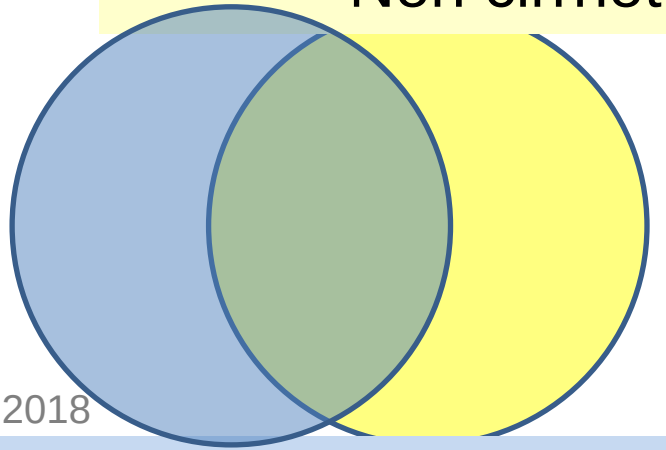


Schouten JN, APT 2012
Schouten JN, Hepatology. 2011
Hernández-Gea V, Hepatology. 2018

Idiopathic portal hypertension
Intrahepatic non cirrhotic portal hypertension

CLINIC

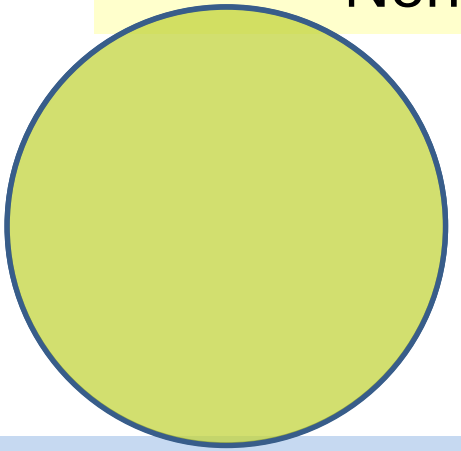
“Porto-sinusoidal vascular disorder” (PSVD)

HISTOLOGY

- Obliterative portal venopathy
- Nodular regenerative hyperplasia
- Hepatoportal sclerosis
- Non cirrhotic portal fibrosis

VALDIG
VASCULAR LIVER DISEASE GROUP

De Gottardi et al,
Lancet Gastro Hepatol 2019



Idiopathic portal hypertension
Intrahepatic non cirrhotic portal hypertension

CLINIC

Porto-sinusoidal vascular disease: proposal and description of a novel entity

Andrea De Gottardi, Pierre-Emmanuel Rautou, Jeoffrey Schouten, Laura Rubbia-Brandt, Frank Leebeek, Jonel Trebicka, Sarwa Darwish Murad, Valérie Vilgrain, Virginia Hernandez-Gea, Filipe Nery, Aurélie Plessier, Annalisa Berzigotti, Paulette Bioulac-Sage, Massimo Primignani, David Semela, Laure Elkrief, Pierre Bedossa, Dominique Valla, Juan Carlos Garcia-Pagan*, on behalf of the VALDIG group*

Lancet Gastroenterol Hepatol
2019; 4: 399–411

Non-cirrhotic portal fibrosis/idiopathic portal hypertension: APASL recommendations for diagnosis and management

Akash Shukla[1] · Don C. Rockey[2] · Patrick S. Kamath[3] · David E. Kleiner[4] · Ankita Singh[1] · Arun Vaidya[1] · Abraham Koshy[5] · Ashish Goel[6] · A. Kadir Dökmeci[7] · Babulal Meena[8] · Cyriac Abby Philips[9] · Chhagan Bihari Sharma[10] · Diana A. Payawal[11] · Dong Joon Kim[12] · Gin-Ho Lo[13] · Guohong Han[14] · Huma Qureshi[15] · Ian R. Wanless[16] · Jidong Jia[17] · Jose D. Sollano[18] · Mamun Al Mahtab[19] · Mark Dhinesh Muthiah[20,34,35] · Mark W. Sonderup[21] · Mendez Sanchez Nahum[22] · Mohamed Ismail Bin Merican[23] · Necati Ormeci[24] · Norifumi Kawada[25] · Rajender Reddy[26] · R. K. Dhiman[27] · Rino Gani[28] · Saeed S. Hameed[29] · Sidharth Harindranath[1] · Wasim Jafri[29] · Xiaolong Qi[30] · Yogesh Kumar Chawla[31] · Yoshihiro Furuichi[32] · Ming-Hua Zheng[33,36] · Shiv Kumar Sarin[8]

Seminar

JOURNAL OF HEPATOLOGY

Baveno VII – Renewing consensus in portal hypertension

Roberto de Franchis[1,*], Jaime Bosch[2,3], Guadalupe Garcia-Tsao[4,5], Thomas Reiberger[6,7], Cristina Ripoll[8], on behalf of the Baveno VII Faculty[§]

Clinical Practice Guidelines

JOURNAL OF HEPATOLOGY

EASL Clinical Practice Guidelines on vascular diseases of the liver☆

HEPATOLOGY

AASLD

HEPATOLOGY, VOL. 73, NO. 1, 2021

Vascular Liver Disorders, Portal Vein Thrombosis, and Procedural Bleeding in Patients With Liver Disease: 2020 Practice Guidance by the American Association for the Study of Liver Diseases

Patrick G. Northup,[1] Juan Carlos Garcia-Pagan,[2–4] Guadalupe Garcia-Tsao,[5,6] Nicolas M. Intagliata,[1] Riccardo A. Superina,[7] Lara N. Roberts,[8] Ton Lisman,[9] and Dominique C. Valla[10,11]

“Porto-sinusoidal vascular disorder” (PSVD)

“Porto-sinusoidal vascular disorder” (PSVD)

Biopsy > 20 mm **w/o cirrhosis**	+	**Specific sign of PH** (varices, portosys. collat, variceal bleeding)
Biopsy > 20 mm **w/o cirrhosis**	+	**Specific histological lesion** (obliterative portal venopathy, NRH)
Biopsy > 20 mm **w/o cirrhosis**	+	1 non specific sign of PH and 1 non specific histological lesion

PSVD: limitations

- Based on expert opinion
- Histology not specific
- Exclusion criteria to revisit

Hernandez-Gea, *et al,* J Hepatol 2026; In Press

We restarted from zero!

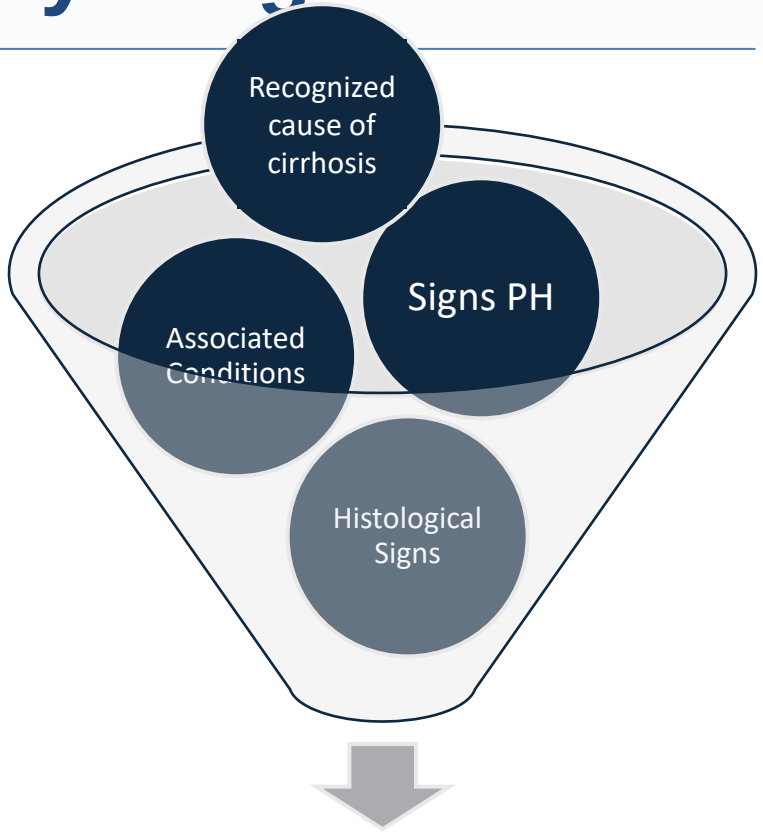
A global initiative

Scientific societies agreed to join this initiative and to designate international experts in adult and pediatric hepatology and pathology to collaborate in the process.

PARIS PSVD MEETING

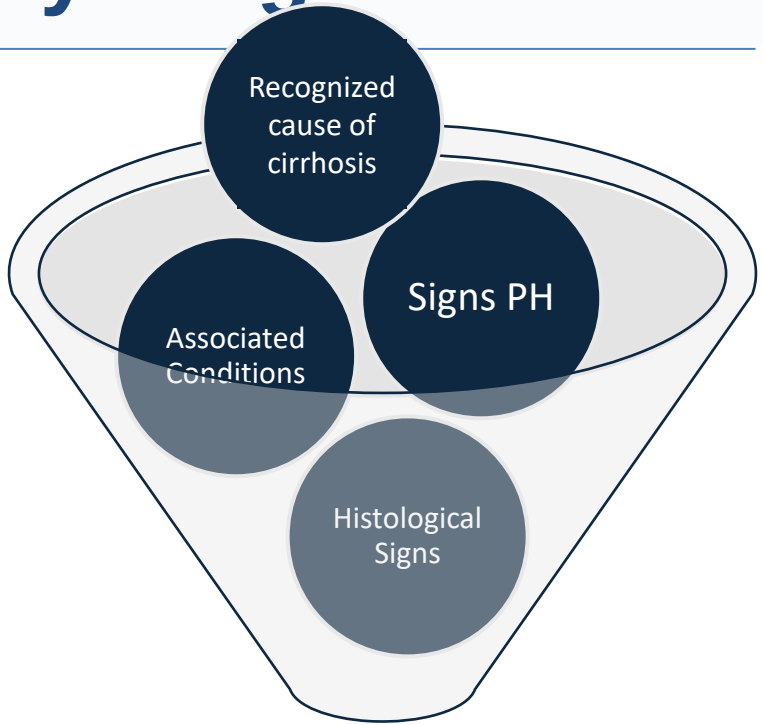
We revisited everything

No Exclusion Criteria & Absence of cirrhosis



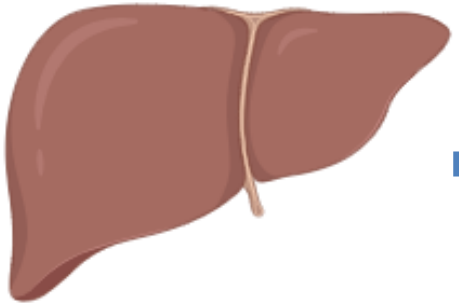
We revisited everything

No Exclusion Criteria &
Absence of cirrhosis



DIAGNOSIS

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

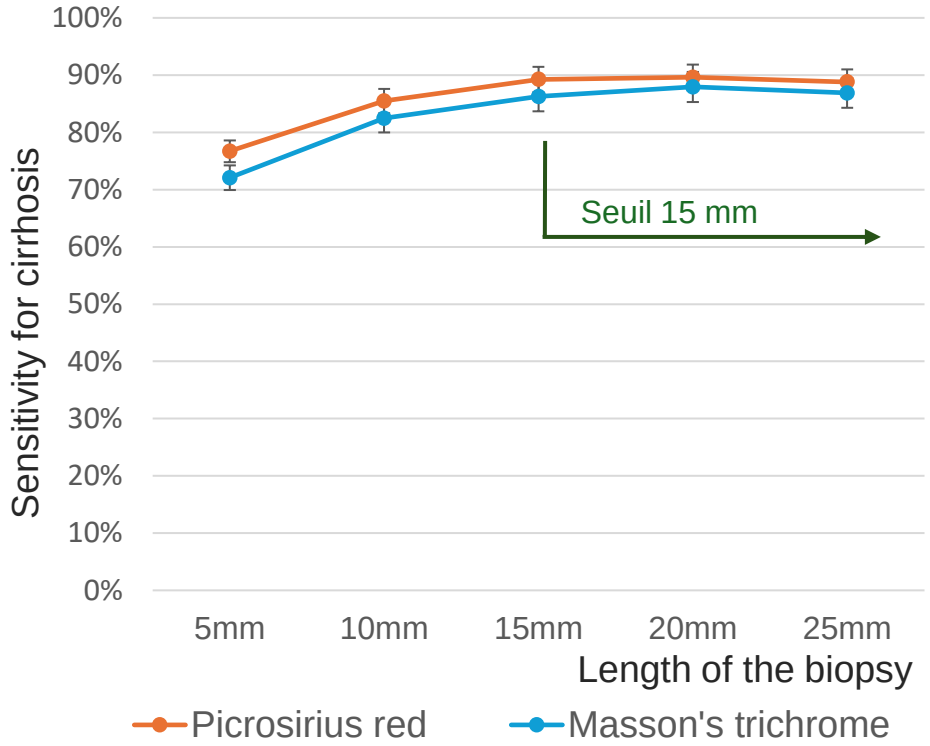
No cirrhosis

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



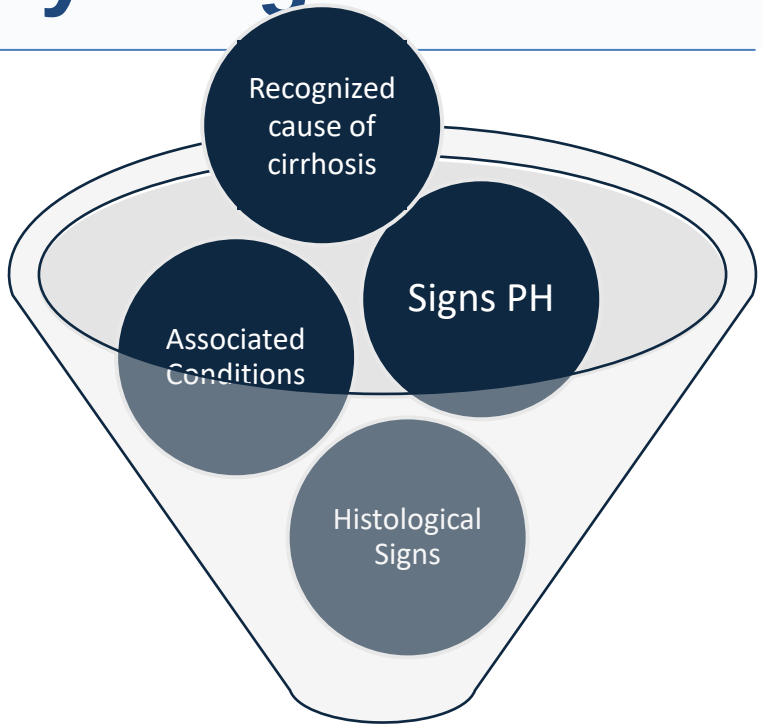
Sensitivity for cirrhosis: 89%

- 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*

We revisited everything

No Exclusion Criteria **&**
Absence of cirrhosis



DIAGNOSIS

Exclusion criteria

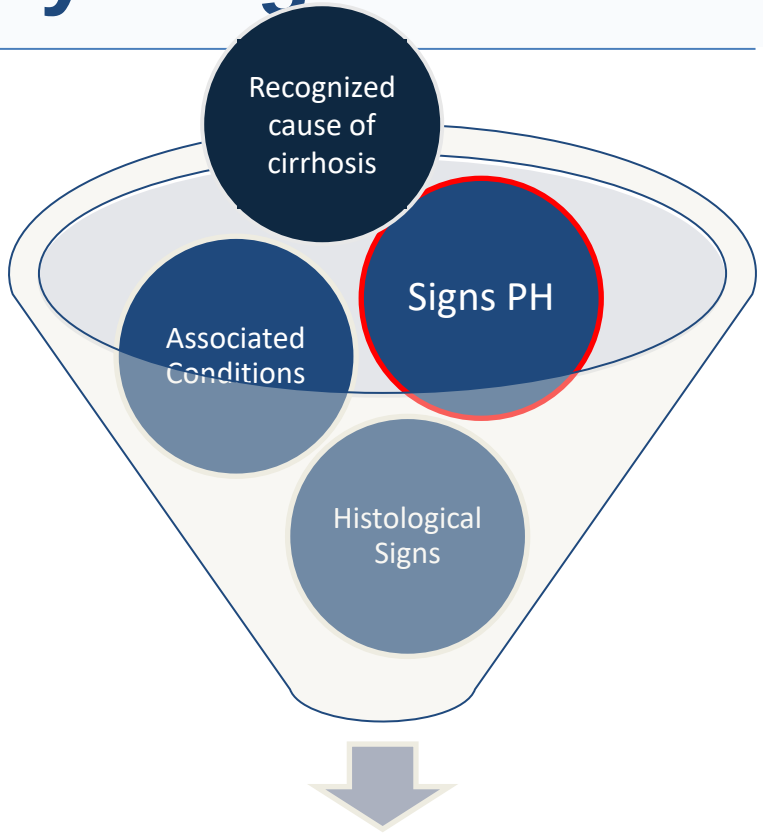
	Exclusion Criteria
1	Abernethy syndrome
2	Budd Chiari syndrome or hepatic venous outflow obstruction
3	Cardiac failure or Fontan Associated Liver Disease
4	Cholestatic liver diseases
5	Congenital hepatic fibrosis
6	Gaucher disease
7	Hepatic amyloidosis
8	Hereditary hemorrhagic telagiectasia
9	Hypervitaminosis 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

We revisited everything

No Exclusion Criteria & Absence of cirrhosis



PSVD: signs of portal hypertension

Specific signs of portal hypertension:

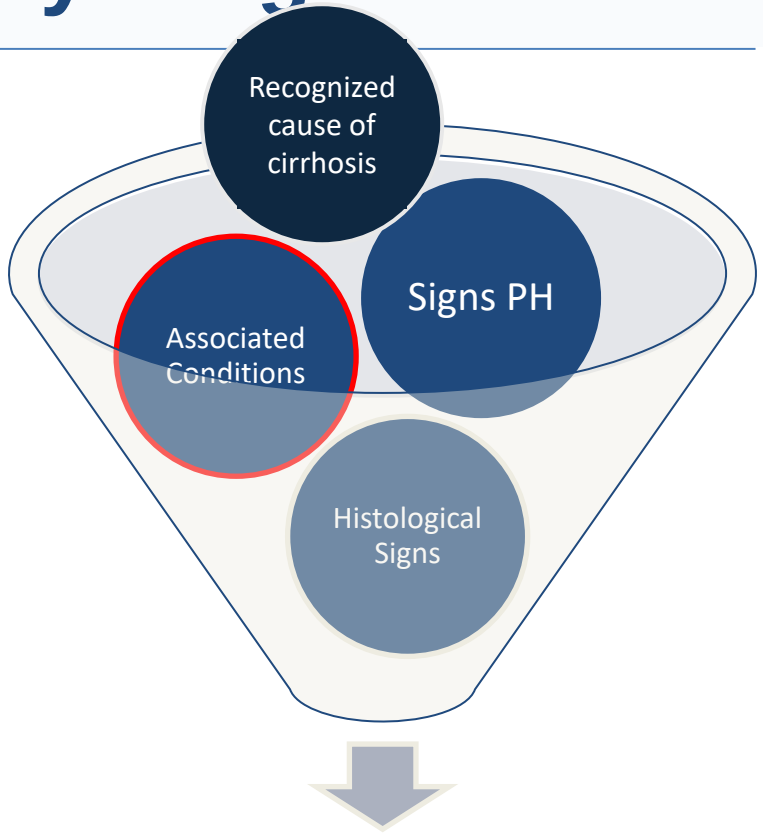
- ✓ Gastrointestinal bleeding due to varices
- ✓ Medium / large oesophageal varices
- ✓ Gastric varices
- ✓ Portosystemic shunt
- ✓ Spleen stiffness > 40 kPa in the absence of MPN

Non specific signs of portal hypertension

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

We revisited everything

No Exclusion Criteria & Absence of cirrhosis



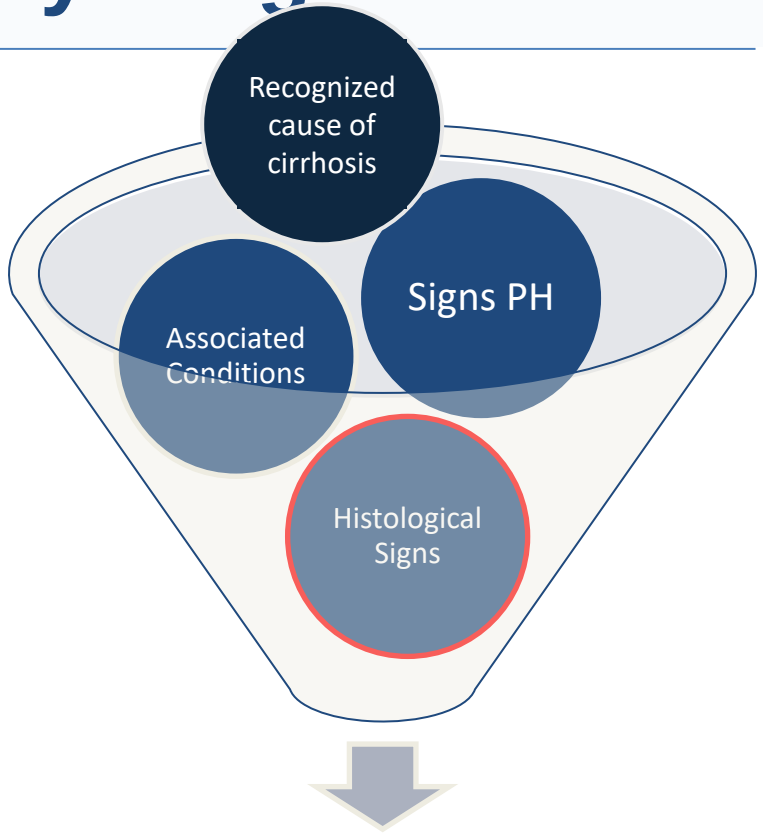
Conditions associated with PSVD (>50% vote)

	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

We revisited everything

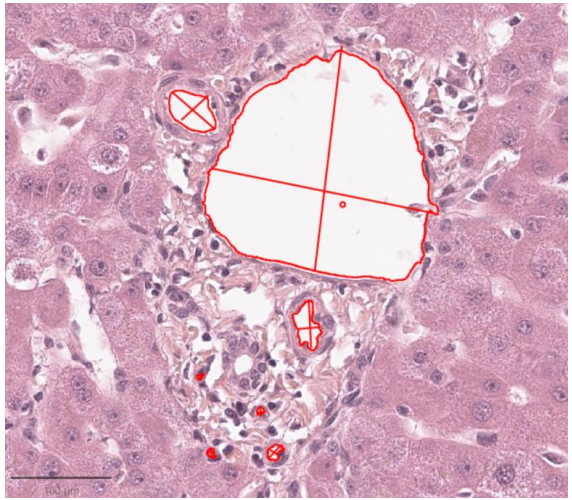
No Exclusion Criteria & Absence of cirrhosis



HOLOGIC
HOLOGIC

PSVD: identification of histological lesions

1. Normal liver vessels



2. Reproducibility of histological lesions

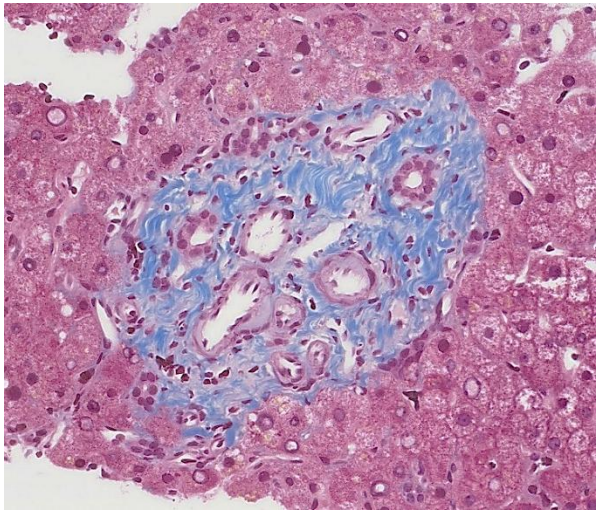
3. Lesions associated with PSVD

PSVD: atlas of histological lesions

Qualitative and quantitative (when possible) description of all elementary histological features & Illustrative pictures

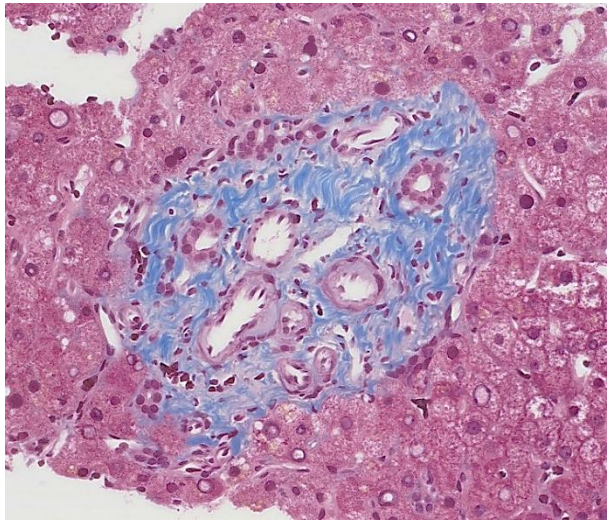
Discussed, updated & validated by pathologists WP1&2 multi-societal

Lesion	Qualitative definition	Quantitative definition
Portal venule (PV) stenosis	Complete obliteration of PV branches or reduced luminal size, with or without wall thickening	To define reduced luminal size, evaluate PV axis relative to the accompanying hepatic arteriole (HA): PV < 1 HA (small axis) or PV < 2 HA (long axis)

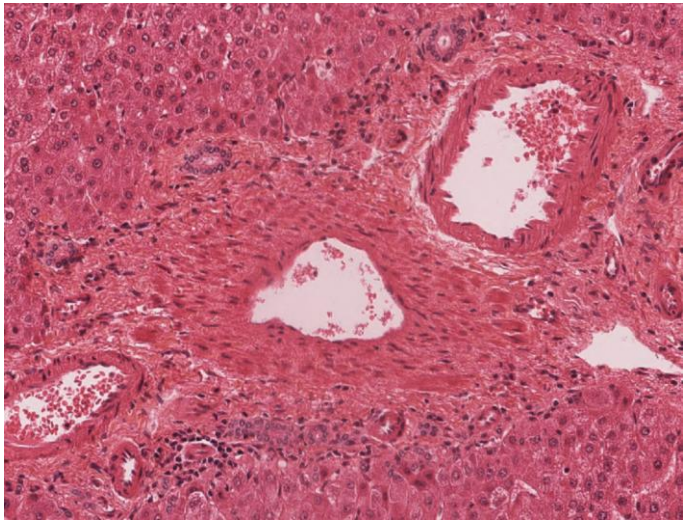


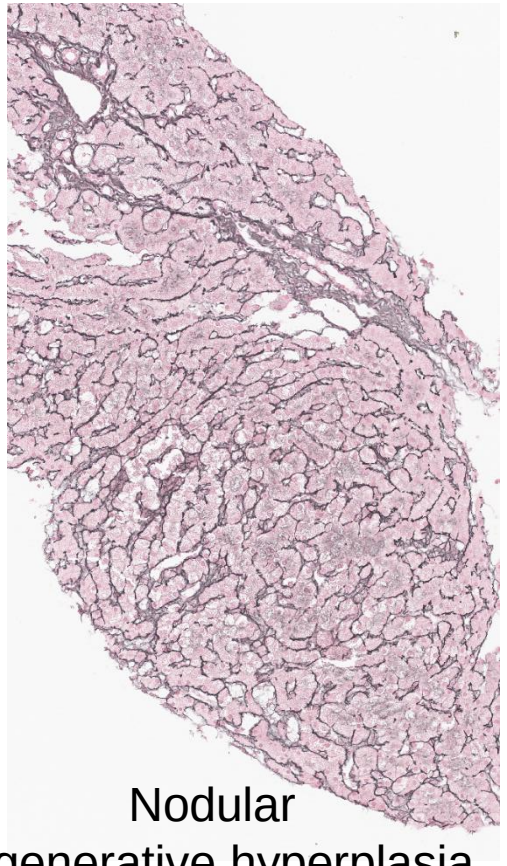
PSVD: major criteria

PV stenosis ≥ 50% of portal tracts



Muscularized portal vein

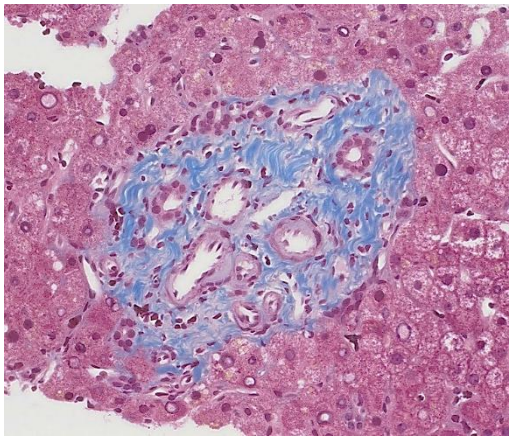




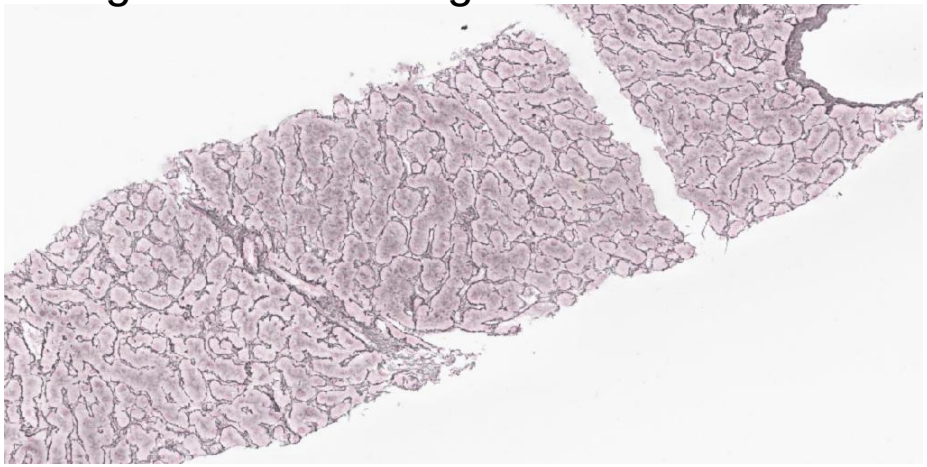
Nodular regenerative hyperplasia

PSVD: minor criteria

PV stenosis 25-49% of portal tracts



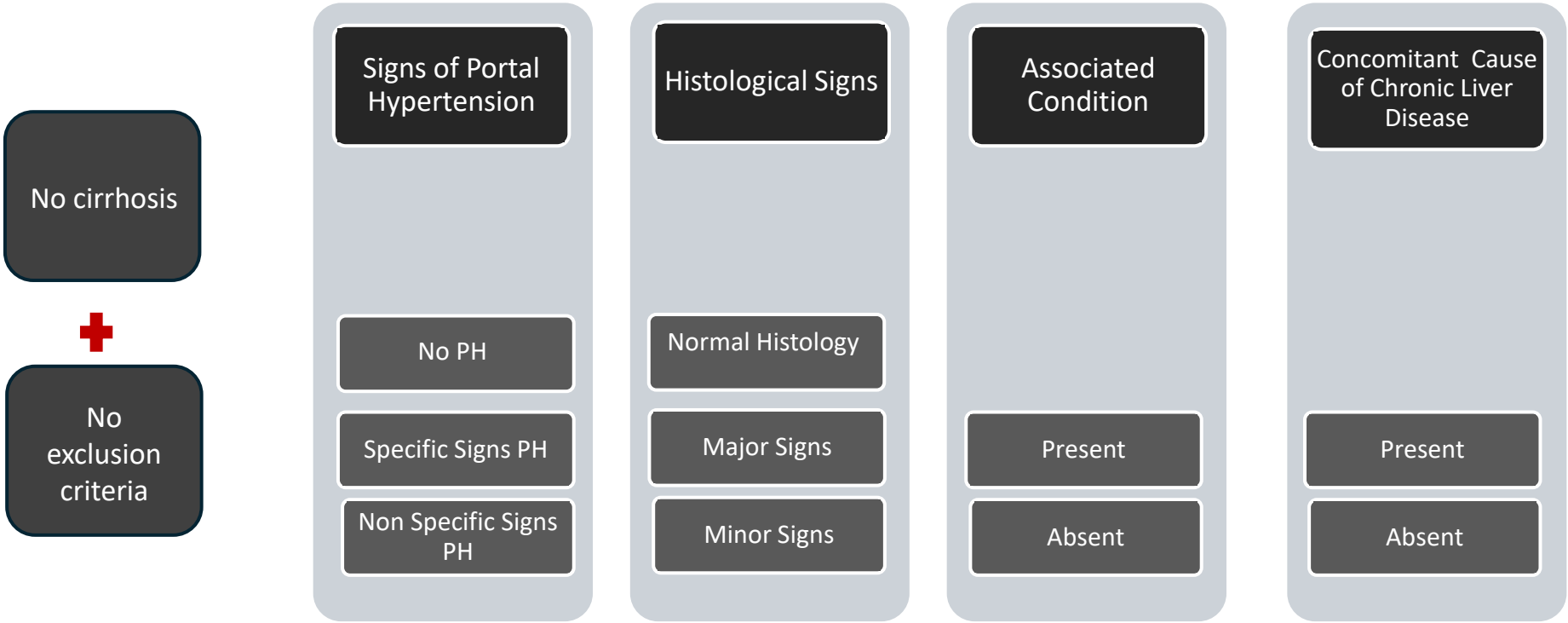
Regenerative changes without obvious NRH



Abnormal distribution of vascular structures

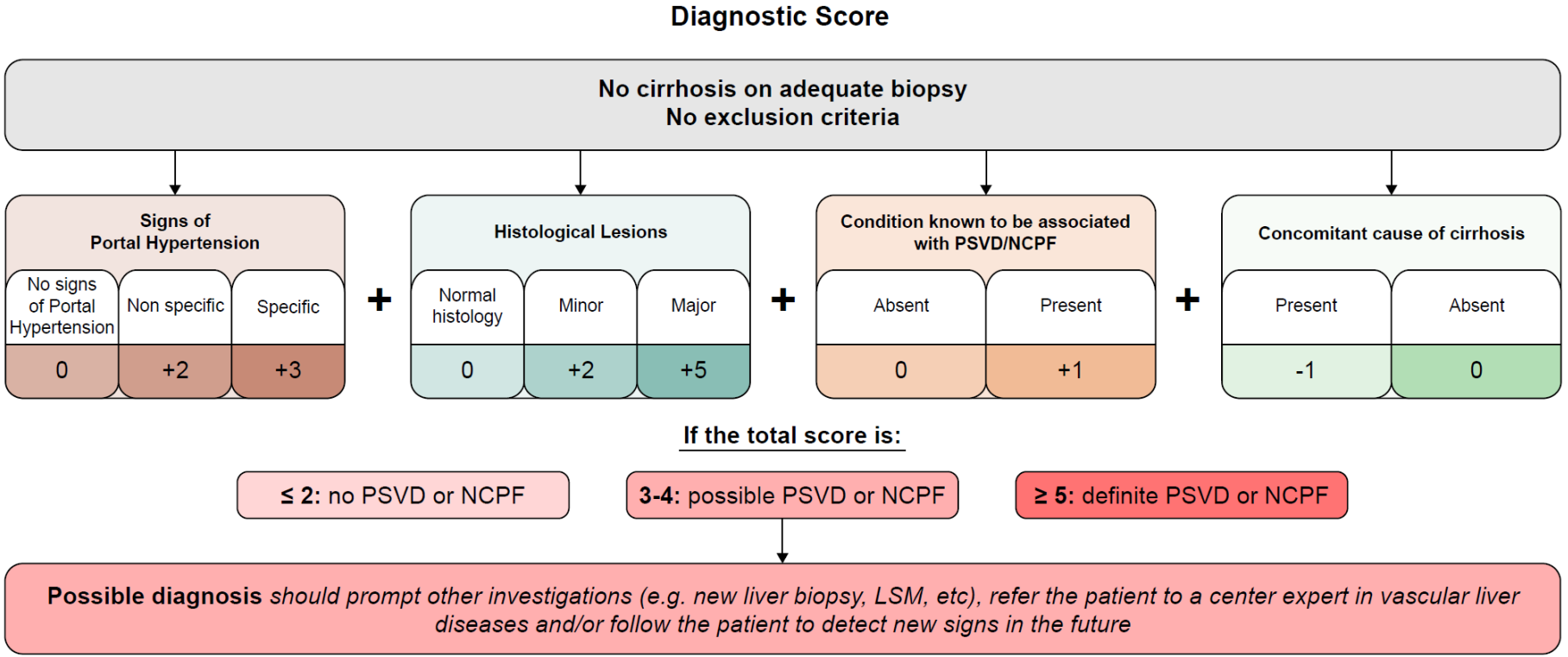
All 3 minor criteria = 1 major

PSVD diagnostic criteria 2026



→ 36 clinical scenarios

PSVD diagnostic criteria 2026



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

Paris PSVD meeting: Dec 2025

What about the name? PSVD or NCPF

Next steps

EASL CONGRESS
Barcelona Spain
27–30 May 2026

JOURNAL OF HEPATOLOGY
EASL
The Home of Liver Research

HEPATOLOGY International
Official Journal of the Asian Pacific Association for the Study of the Liver

HEPATOLOGY

ANNALS OF Hepatology

COST ACTION

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

6 working groups

Start Date: 30/10/2024

End Date: 29/10/2028

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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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