ASSISTANCE PUBLIQUE HÔPITAUX DE PARIS

Université Paris Cité

Diseases affecting intrahepatic vessels

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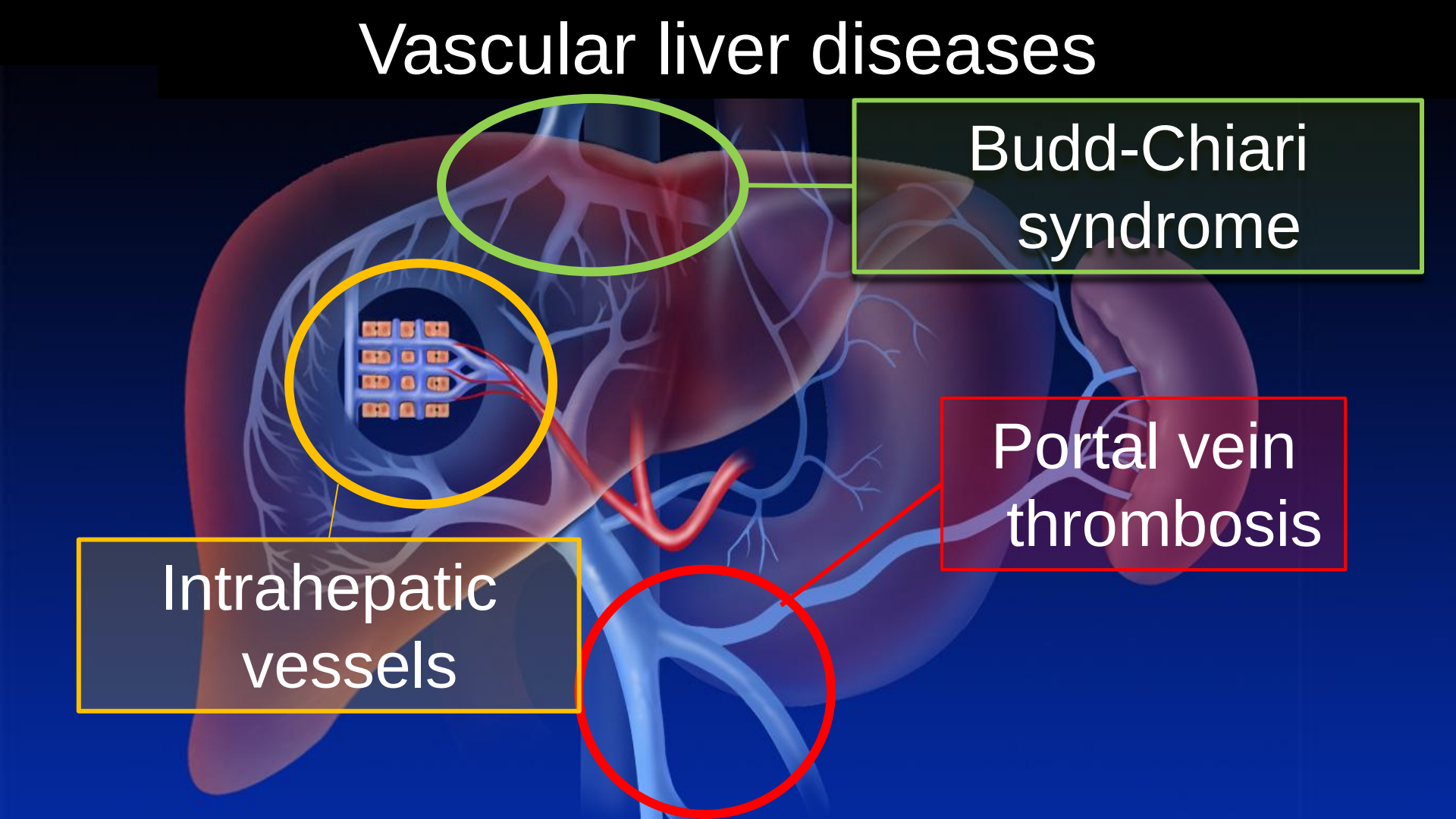
MVF
MALADIES VASCULAIRES DU FOIE
RÉSEAU D'EXPERTISE

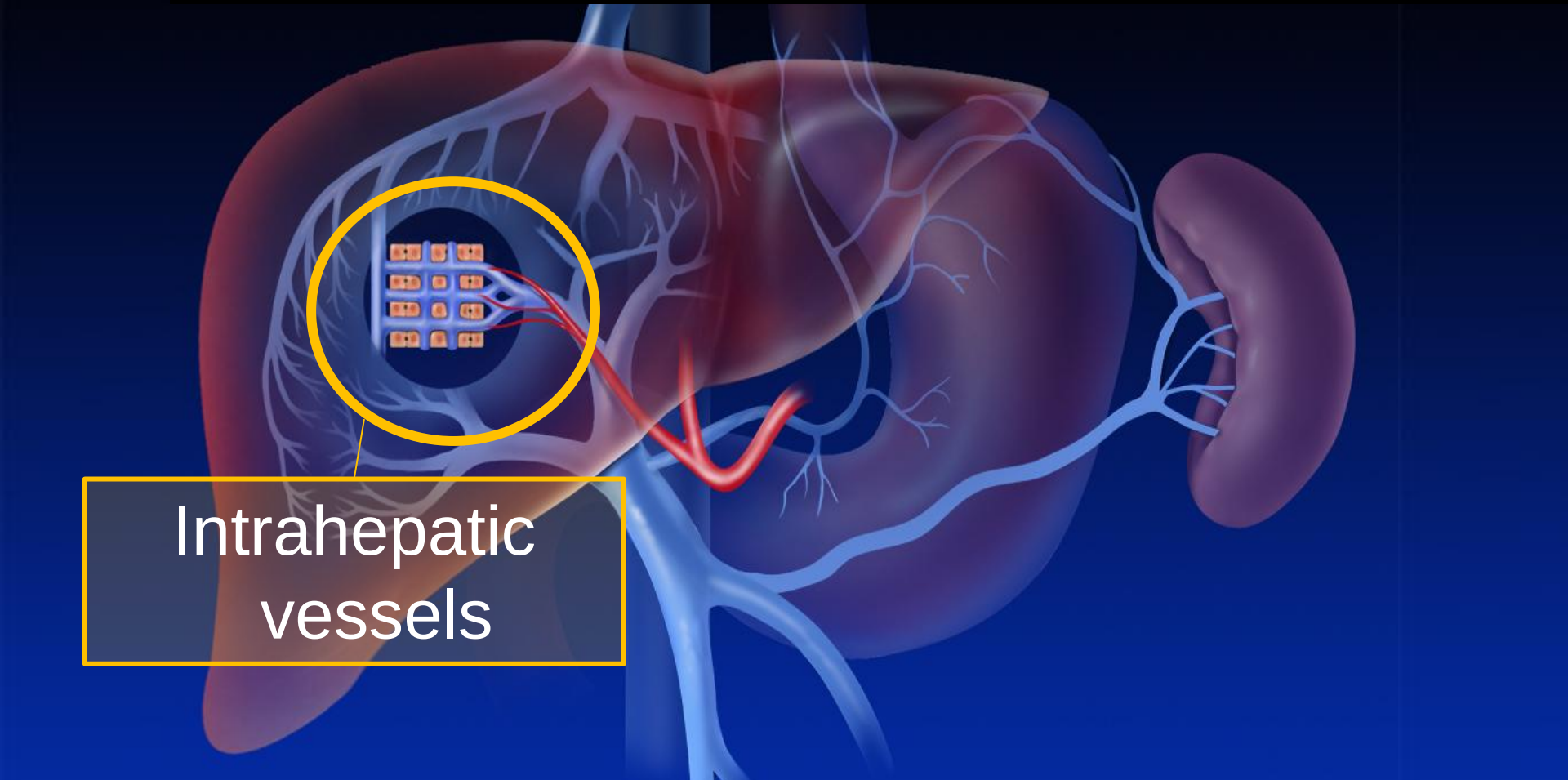
Instituts thématiques Inserm
Institut national de la santé et de la recherche médicale

CRI
CENTRE DE RECHERCHE SUR L'INFLAMMATION

Vascular liver diseases



Vascular liver diseases

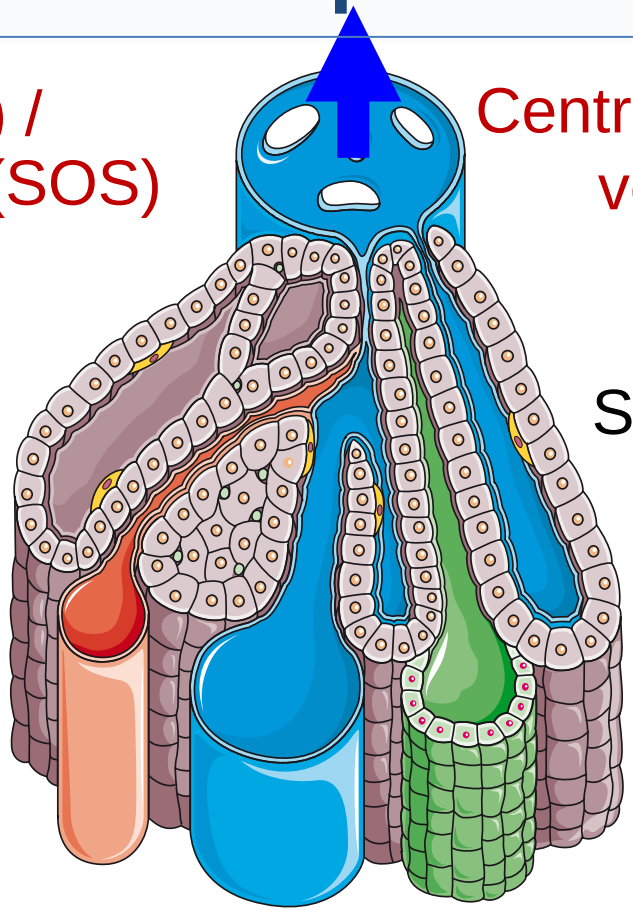


Diseases affecting small intrahepatic vessels

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

Sinusoidal dilation / Peliosis

Portosinusoidal vascular disorder (PSVD)



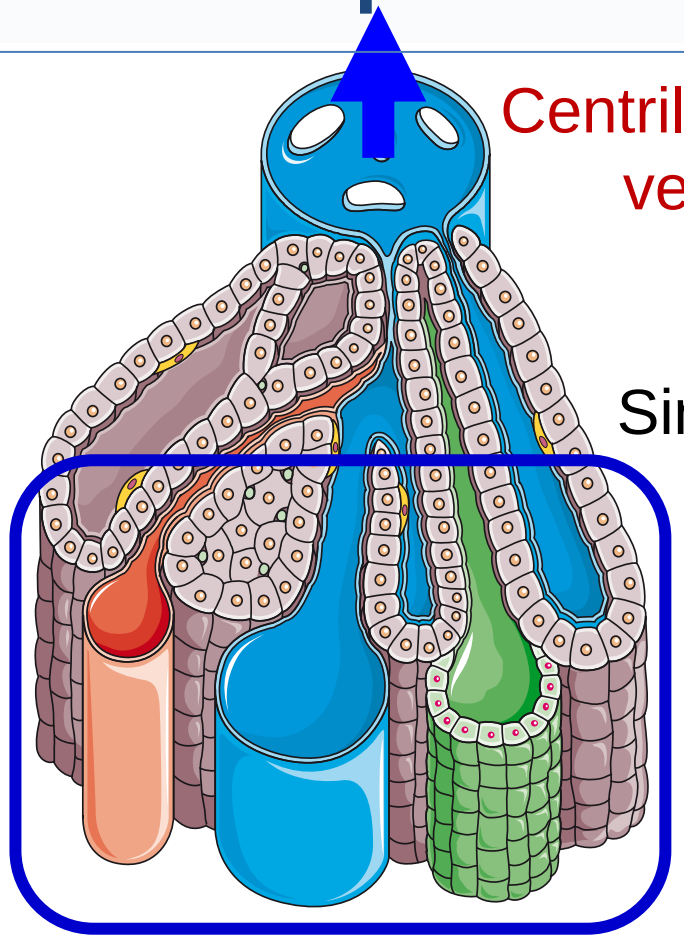
Centrilobular vein

Sinusoids

Portal tract

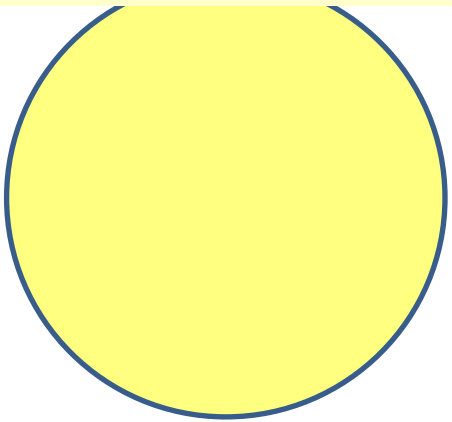
Diseases affecting small intrahepatic vessels

Portosinusoidal vascular disorder (PSVD)



HISTOLOGY

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



Verheij J, Histopathology. 2013
Guido M, Histopathology. 2019
Kmeid M, Diagn Pathol. 2020

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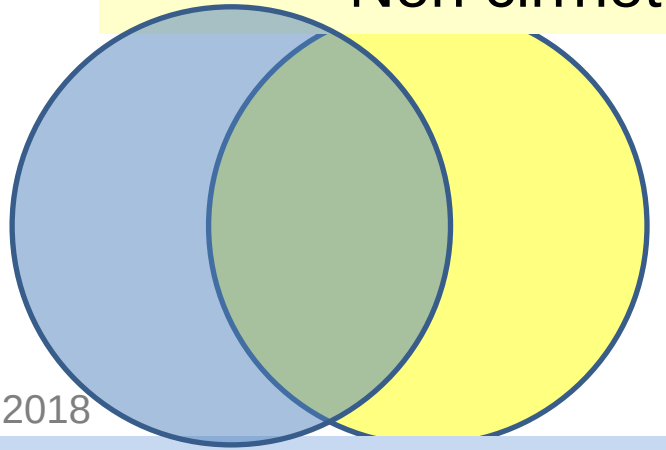


Schouten JN, APT 2012
Schouten JN, Hepatology. 2011
Hernández-Gea V, Hepatology. 2018

Idiopathic portal hypertension
Intrahepatic non cirrhotic portal hypertension

CLINIC

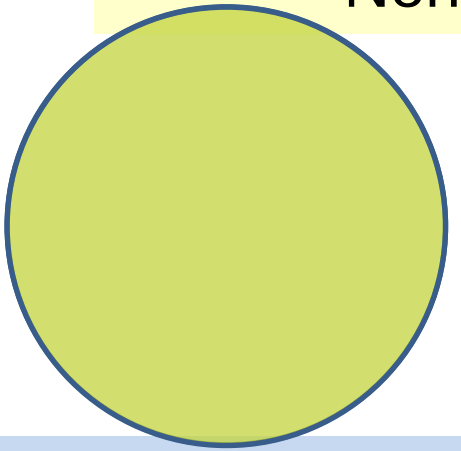
“Porto-sinusoidal vascular disorder” (PSVD)

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)

PSVD: limitations

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

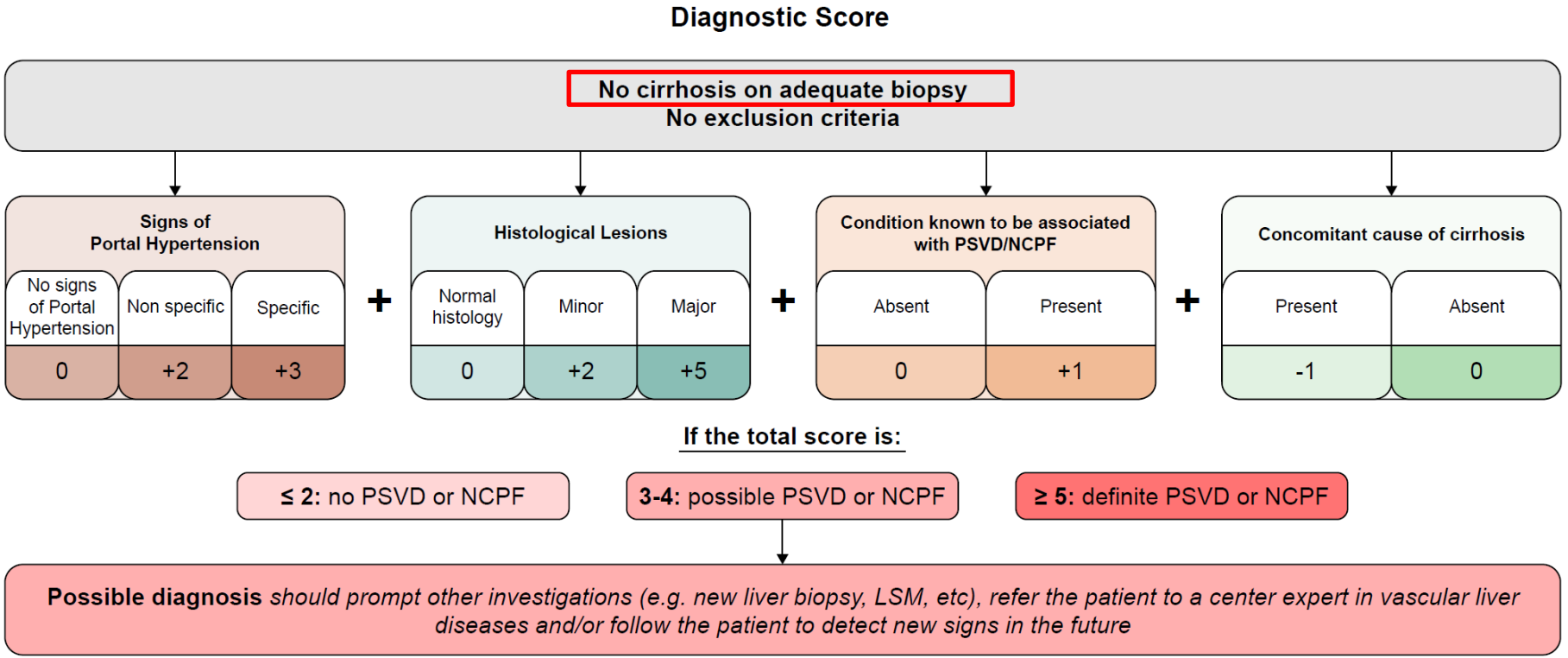
Hernandez-Gea, *et al,* J Hepatol 2026

We restarted from zero!

Porto-sinusoidal vascular disorder

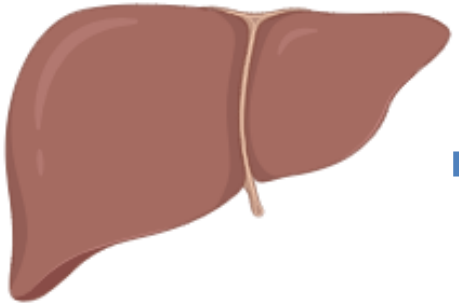
- Definition
- When to suspect?
- Outcome and treatment

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

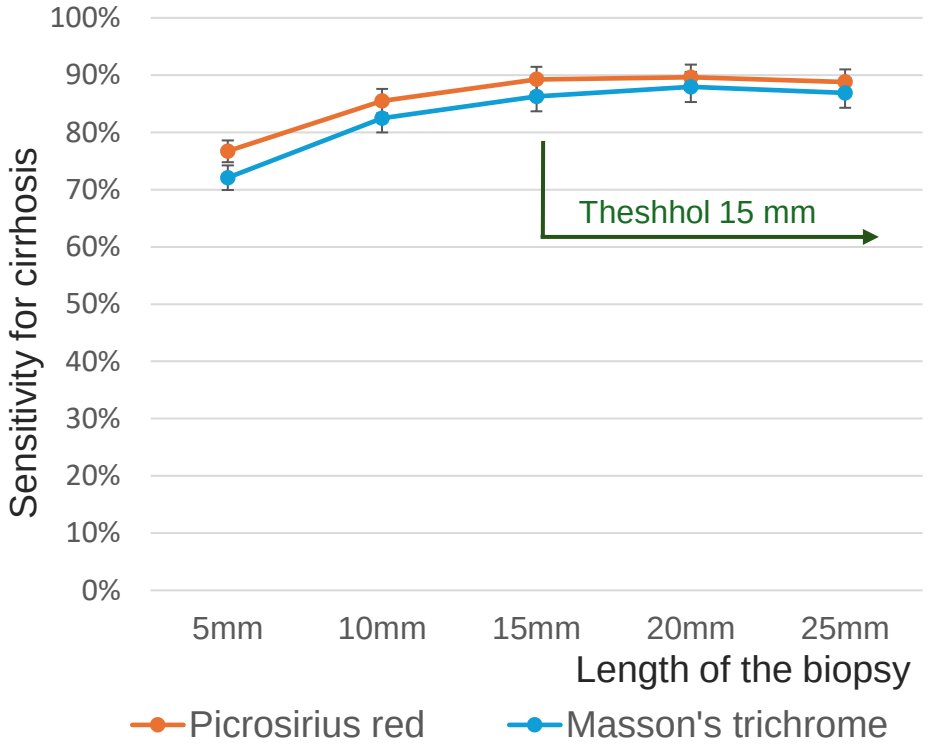
No cirrhosis

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

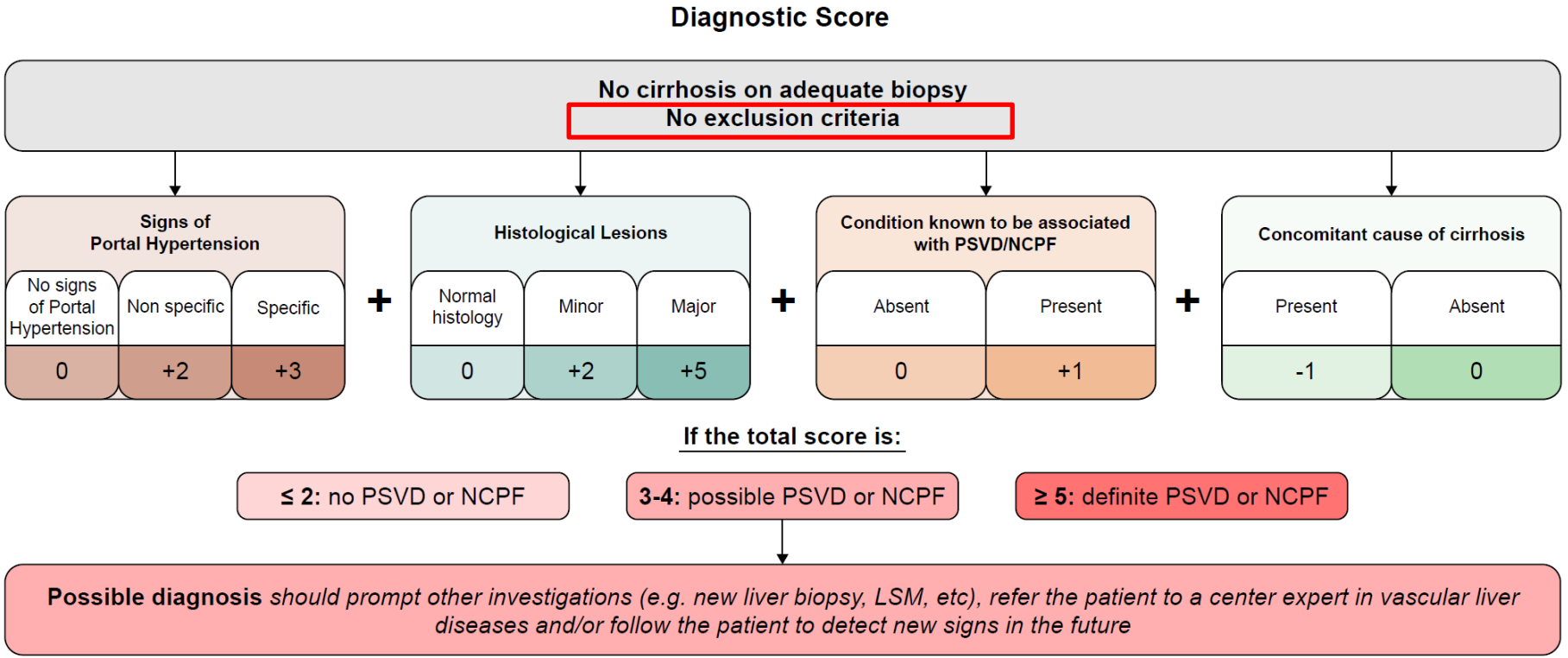


Sensitivity for cirrhosis: 89%

- 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

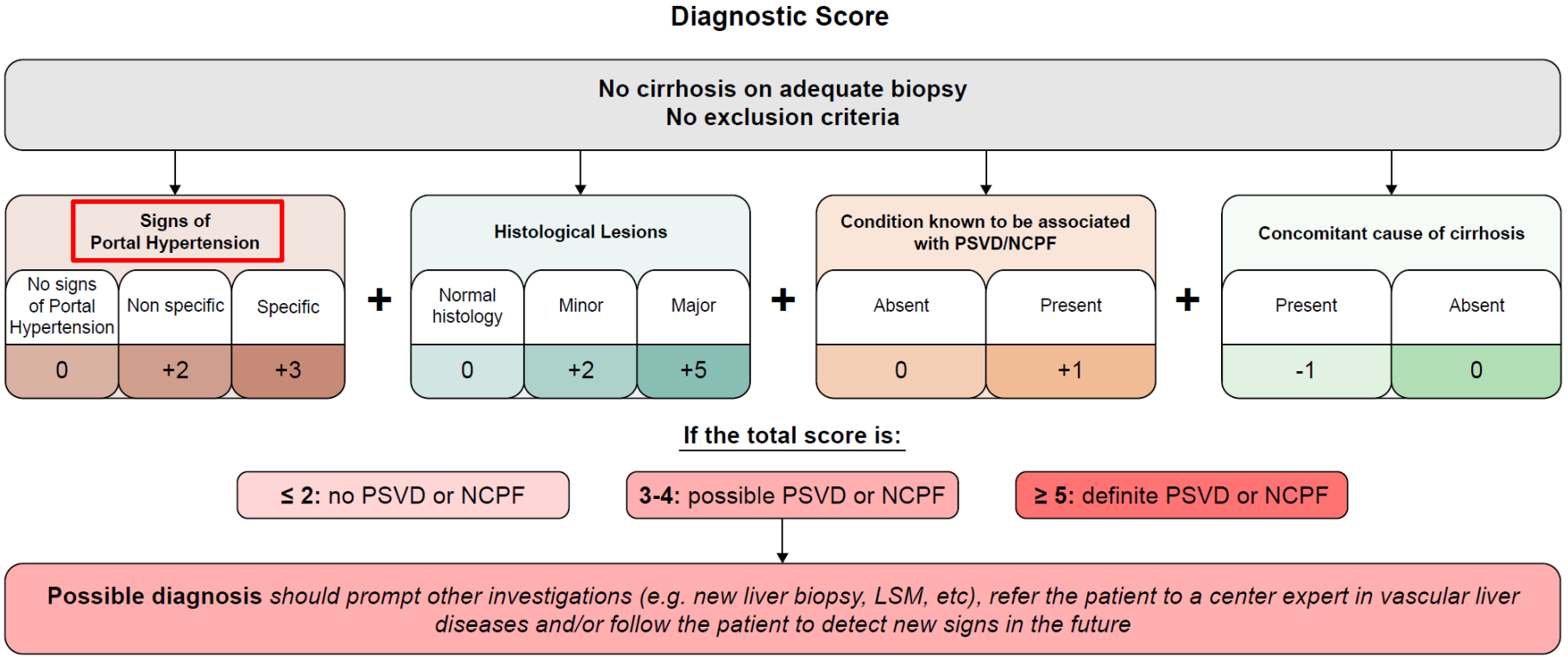
Exclusion criteria

	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

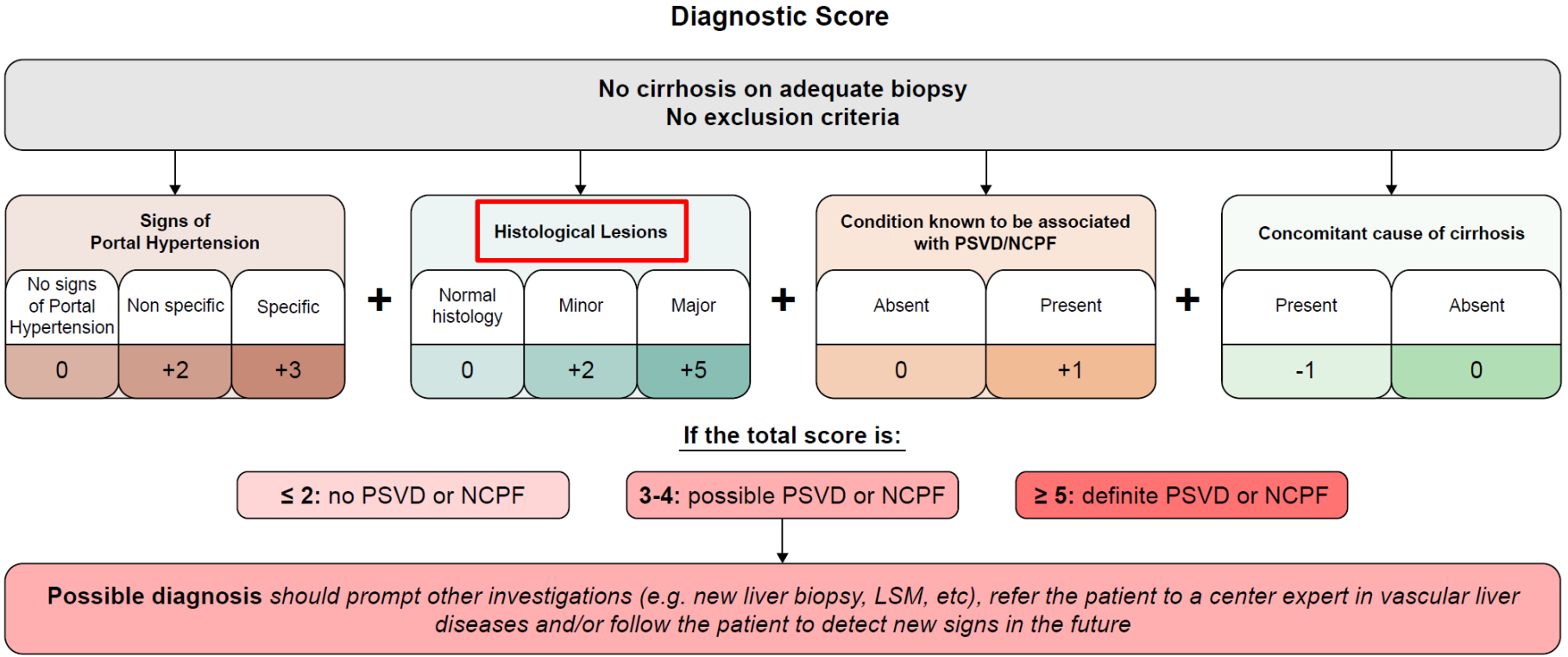
Specific signs of portal hypertension:

- ✓ 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

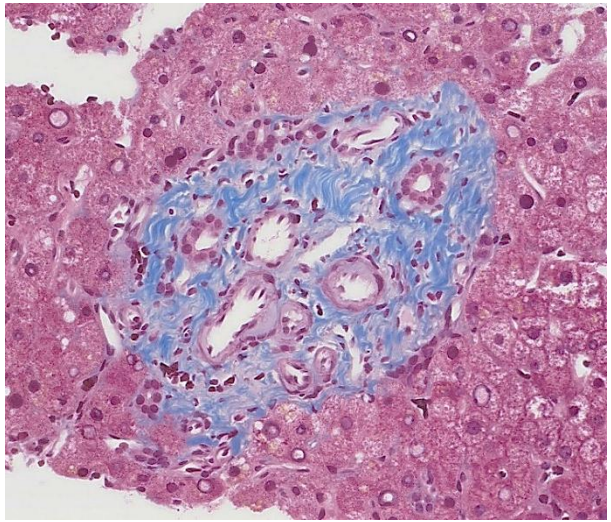
PSVD diagnostic criteria 2026



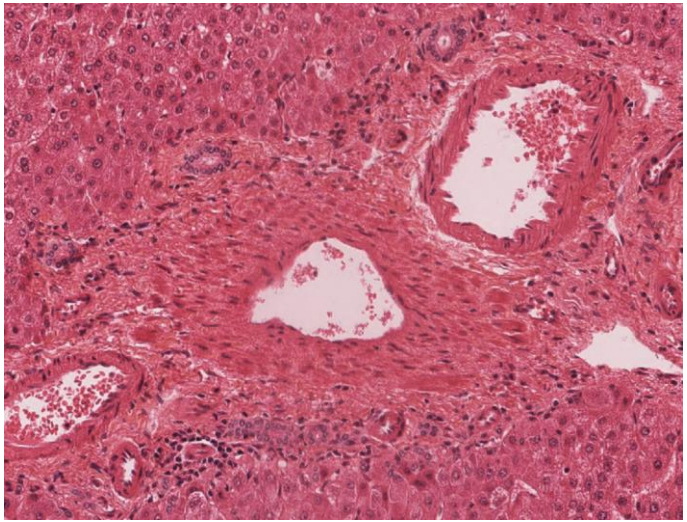
Hernandez-Gea, *et al,* J Hepatol 2026; Baveno VIII

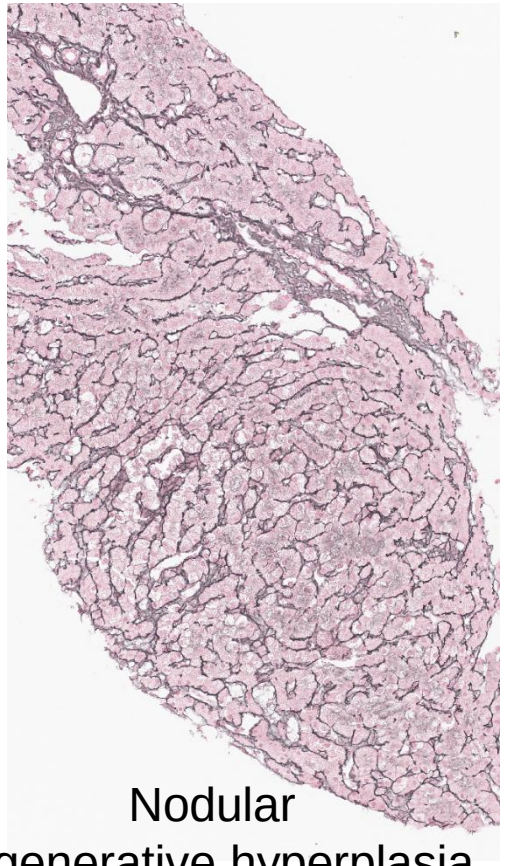
PSVD: major criteria

PV stenosis ≥ 50% of portal tracts



Muscularized portal vein

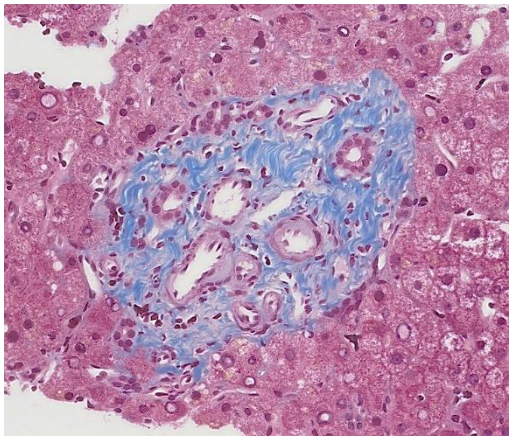




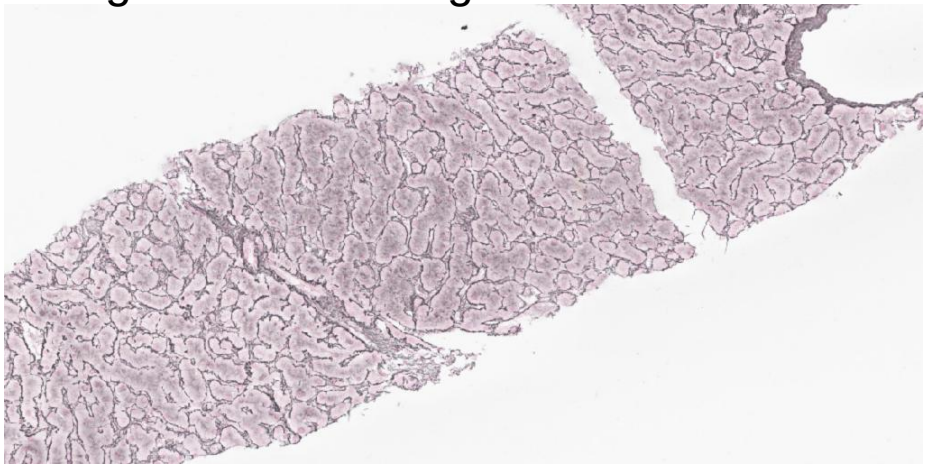
Nodular regenerative hyperplasia

PSVD: minor criteria

PV stenosis 25-49% of portal tracts



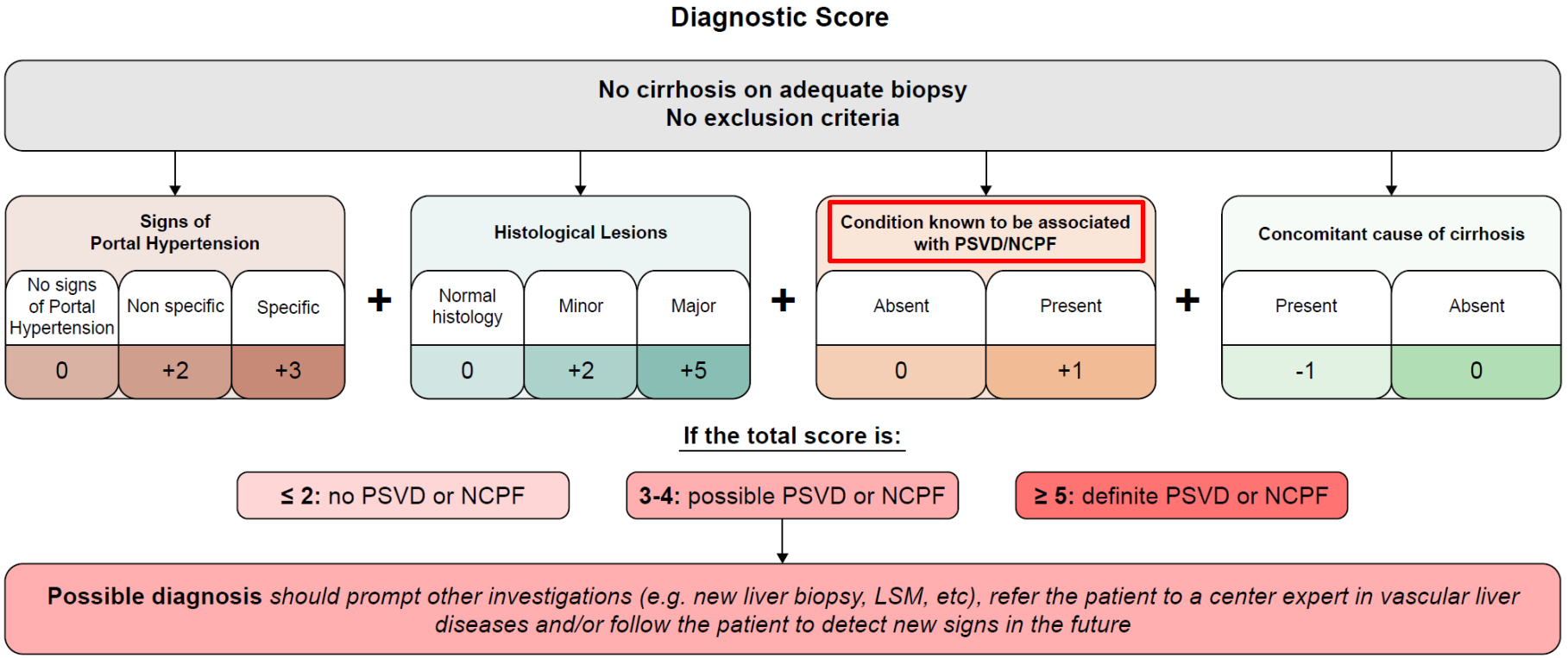
Regenerative changes without obvious NRH



Abnormal distribution of vascular structures

All 3 minor criteria = 1 major

PSVD diagnostic criteria 2026



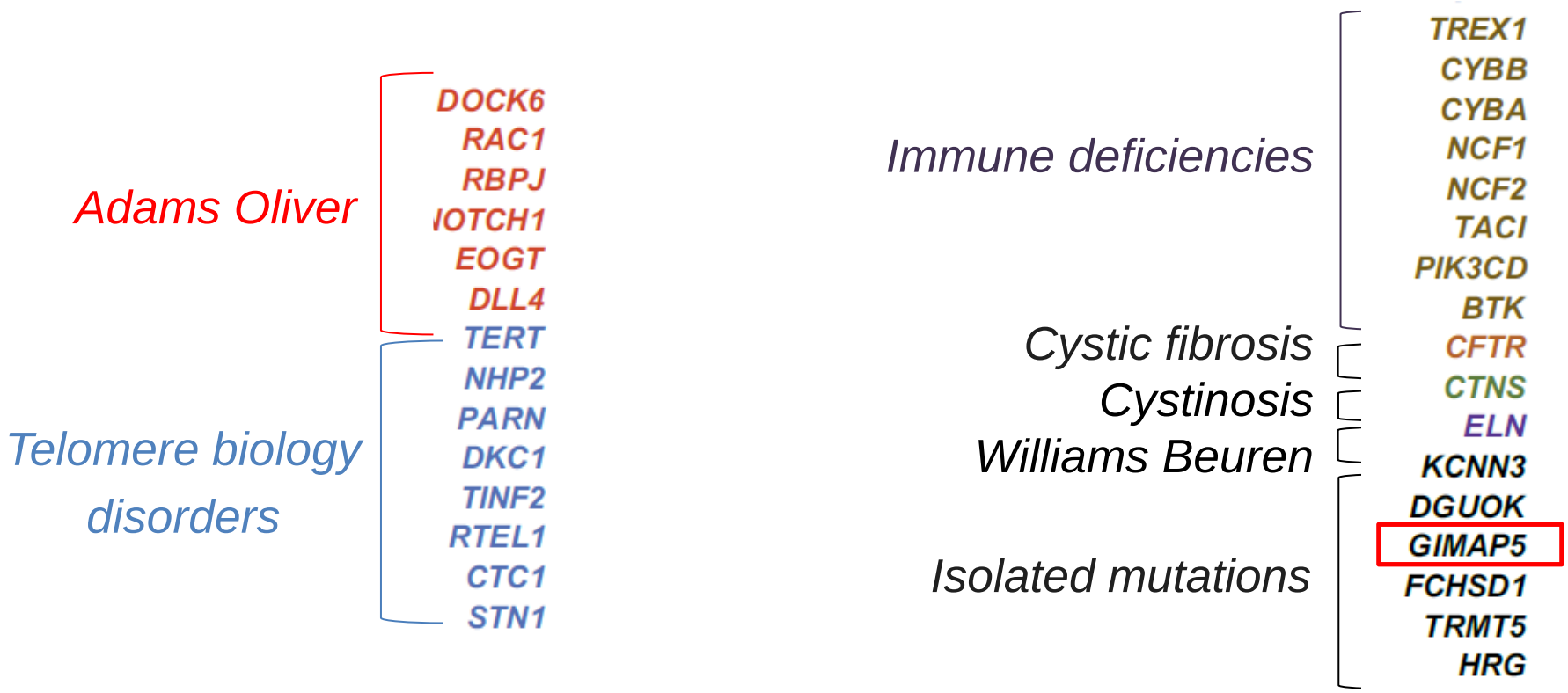
Hernandez-Gea, *et al,* J Hepatol 2026; Baveno VIII

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%**

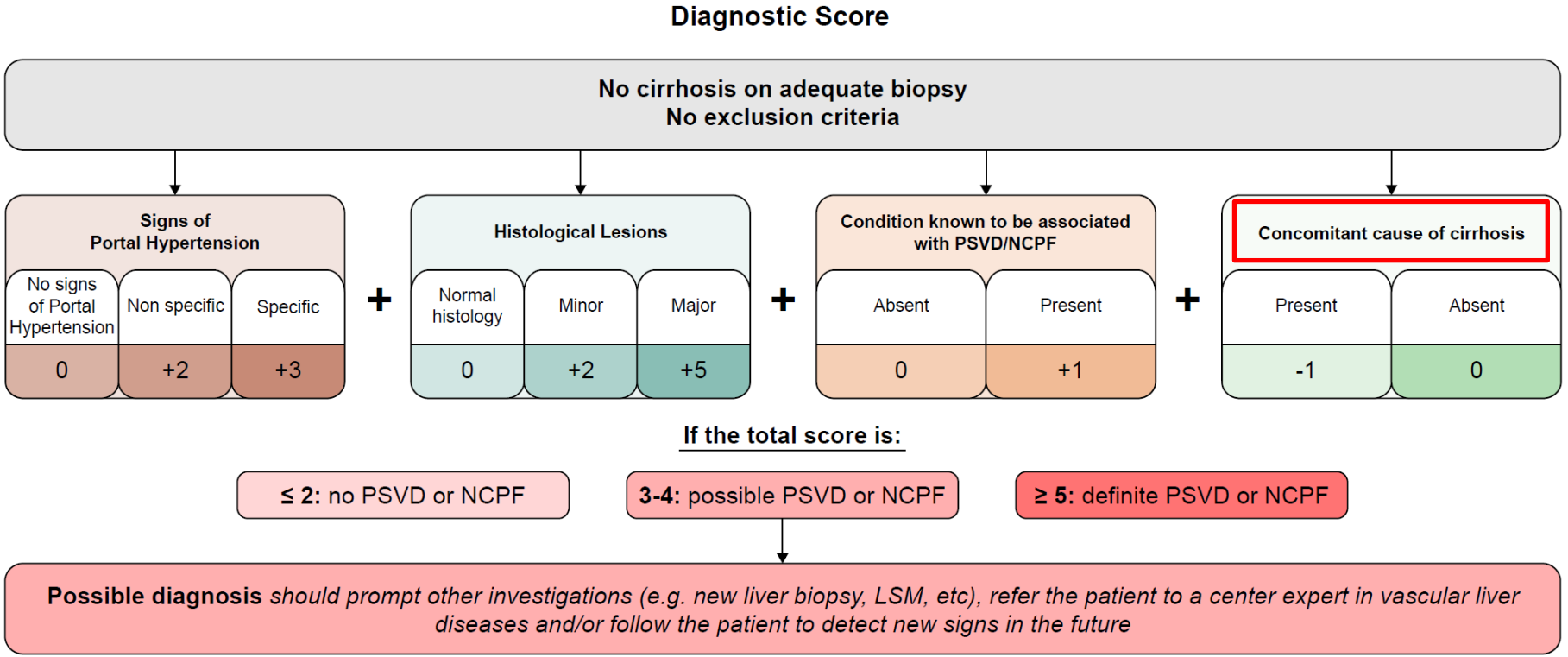
Magaz *et al.*,
J Hepatol 2025

Genetic forms of PSVD



Ciriaci N*, Bertin L*, Rautou *Hepatology*, *2026*

PSVD diagnostic criteria 2026

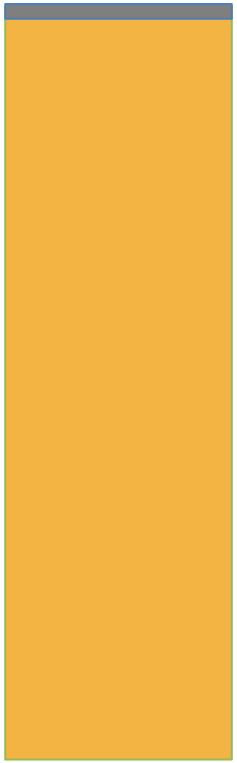


Hernandez-Gea, *et al,* J Hepatol 2026; Baveno VIII

Porto-sinusoidal vascular disorder

- Definition
- When to suspect?
- Outcome and treatment

PSVD among cirrhosis

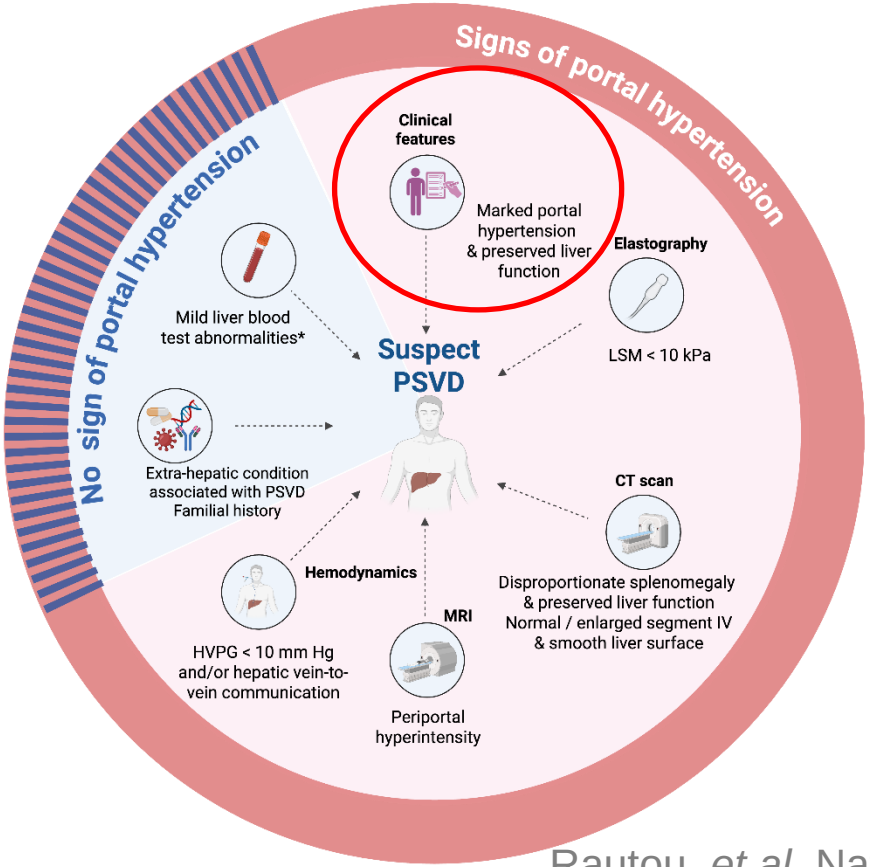


PSVD
0.4%

Cirrhosis
99.6%

➔ Screening tools needed

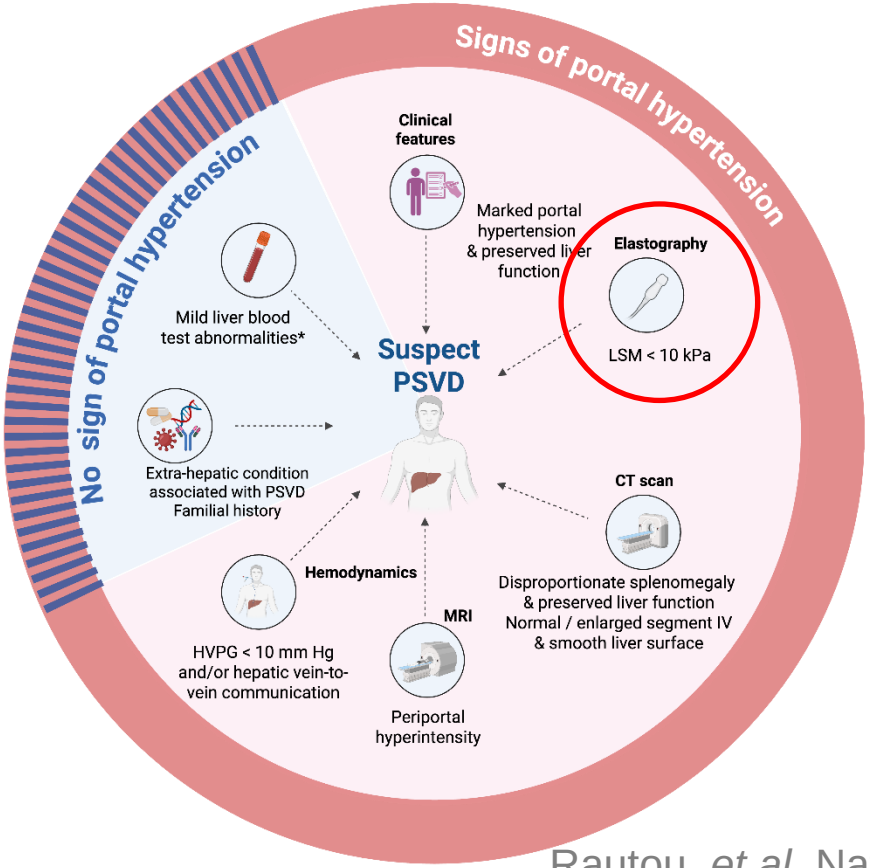
PSVD: when to suspect?



Paris PSVD meeting

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

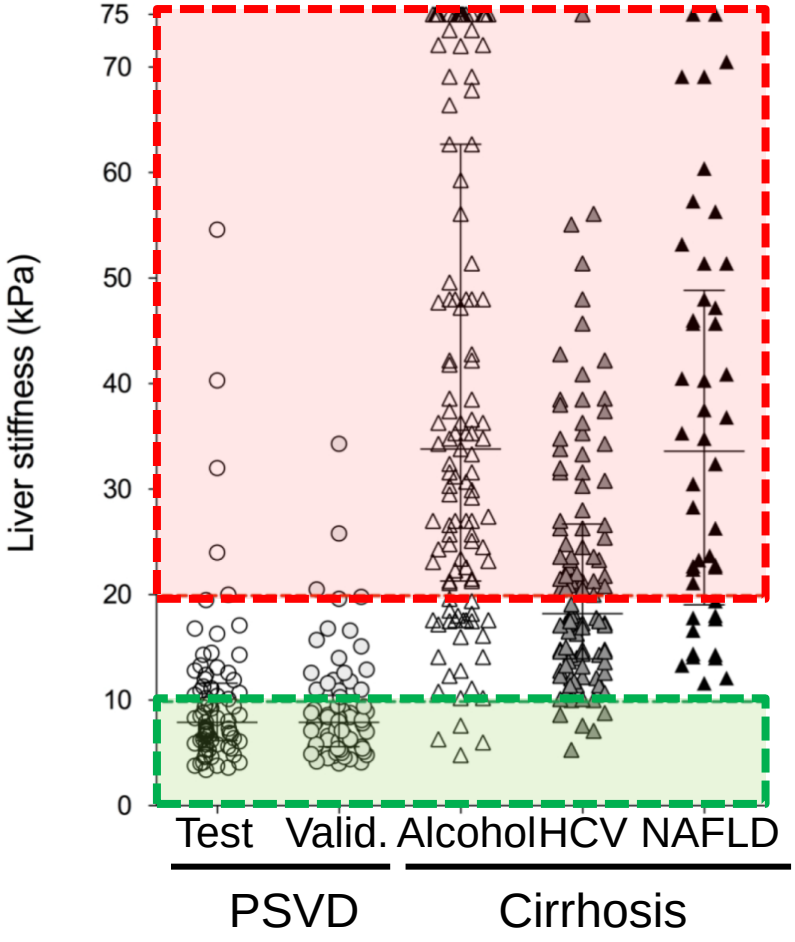
PSVD: when to suspect?



Paris PSVD meeting

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

PSVD: liver stiffness measurement (TE)



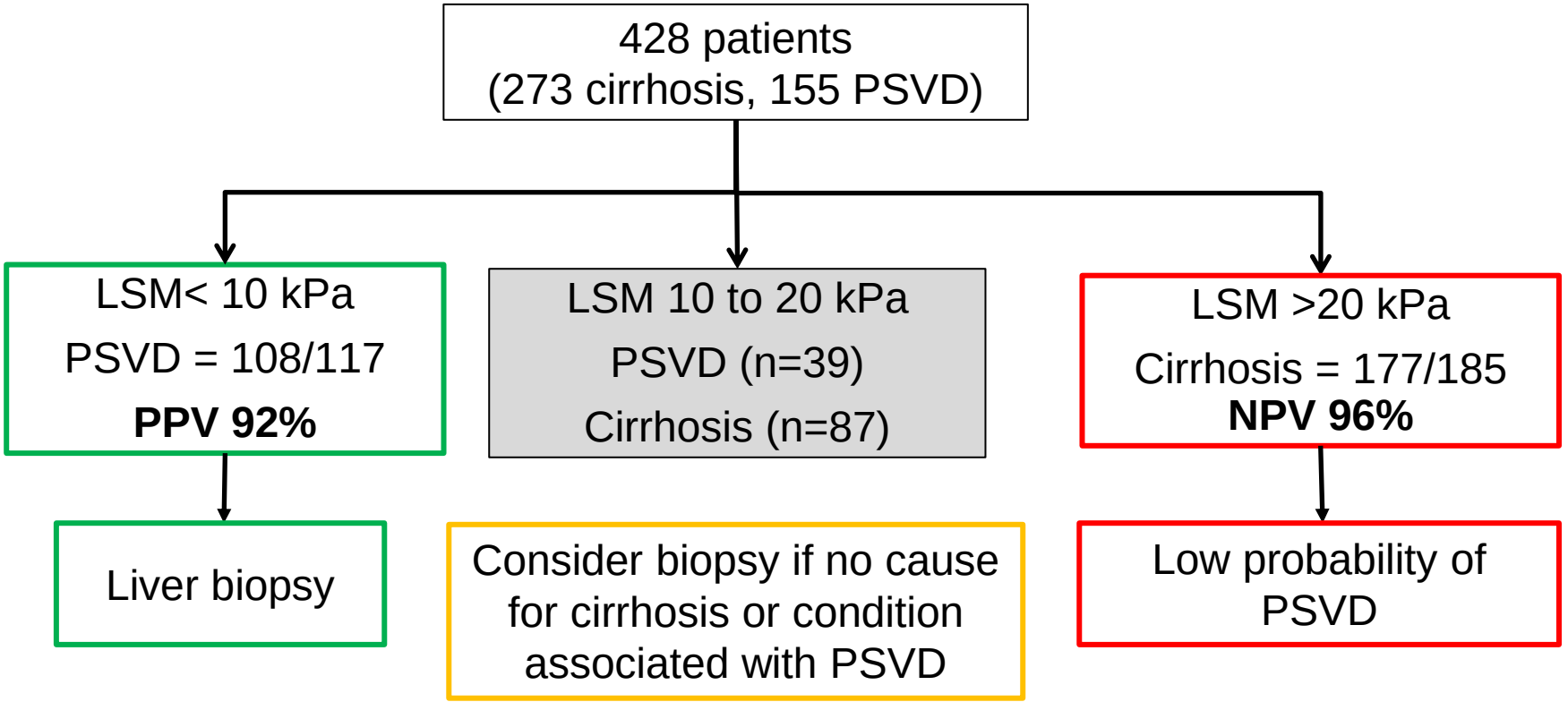
Patients with signs of portal hypertension
155 PSVD, 273 cirrhosis

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

Similar results in:

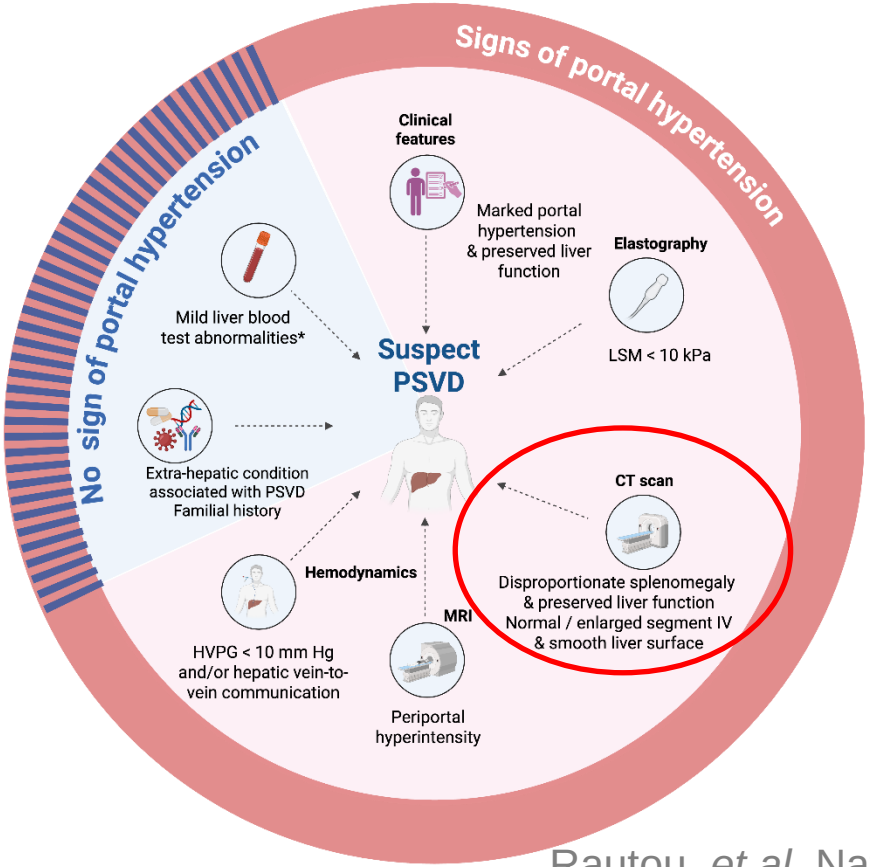
- Laharie Eur J Gastroenterol Hepatol 2010
- Seijo Dig Liver Dis 2012
- Sharma J Clin Exp Hepatol 2017
- Ferreira-Silva J, Dig Liver Dis. 2022

PSVD: liver stiffness measurement (TE)



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

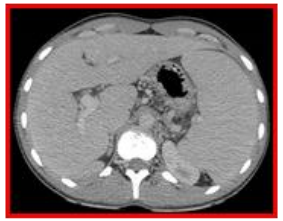
PSVD: when to suspect?



Paris PSVD meeting

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

PSVD: CT-scan



50 patients with PSVD

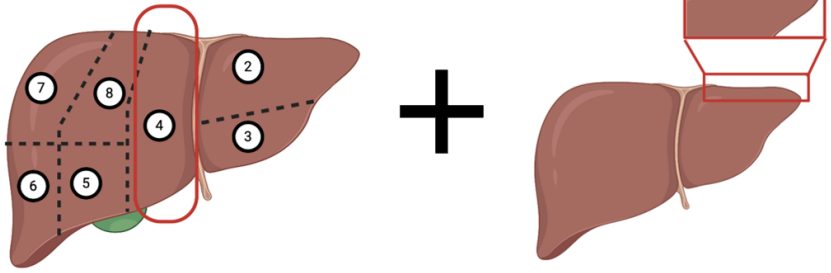
Matched on ascites

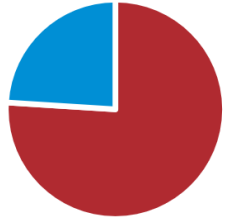
100 patients with cirrhosis

CT features for PSVD diagnosis

No atrophy of segment IV

LSN < 2.5





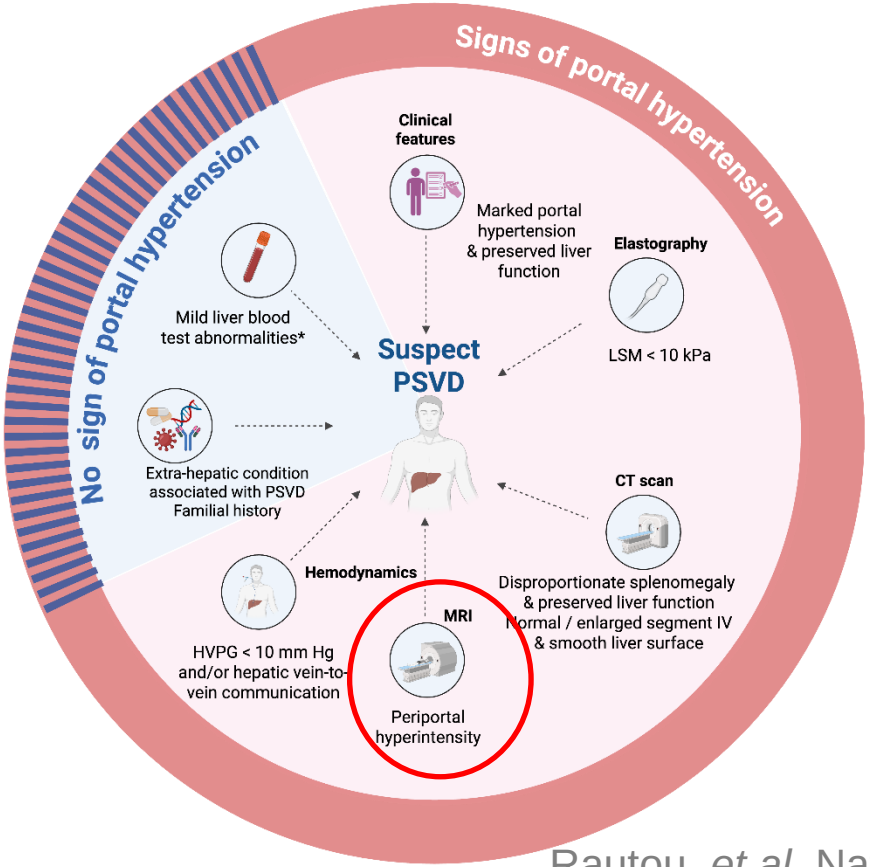
With the features

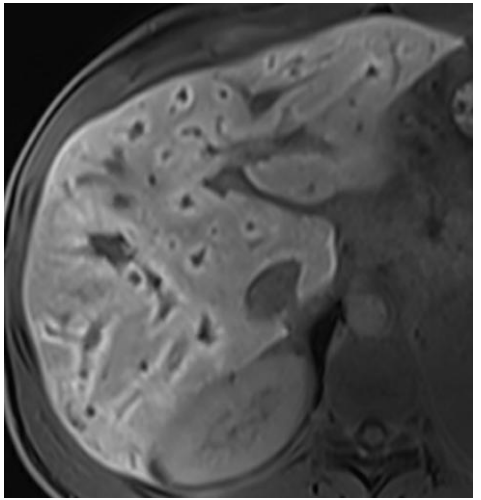
PSVD Cirrhosis

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?



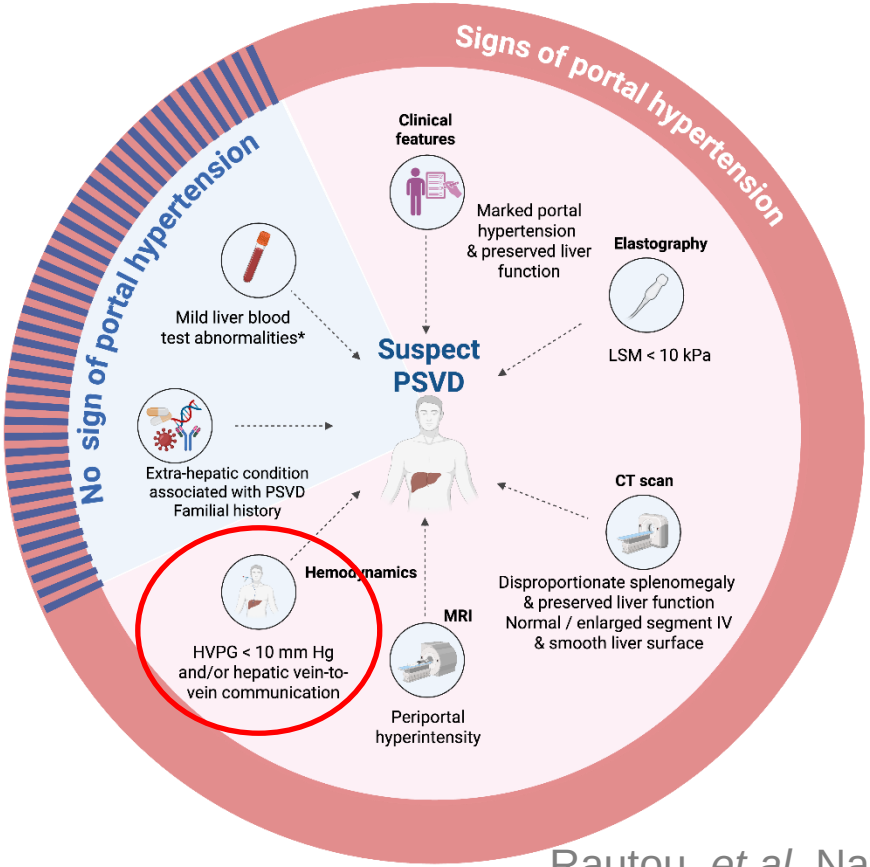


Periportal hyperintensity on hepatobiliary phase

Paris PSVD meeting

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

PSVD: when to suspect?



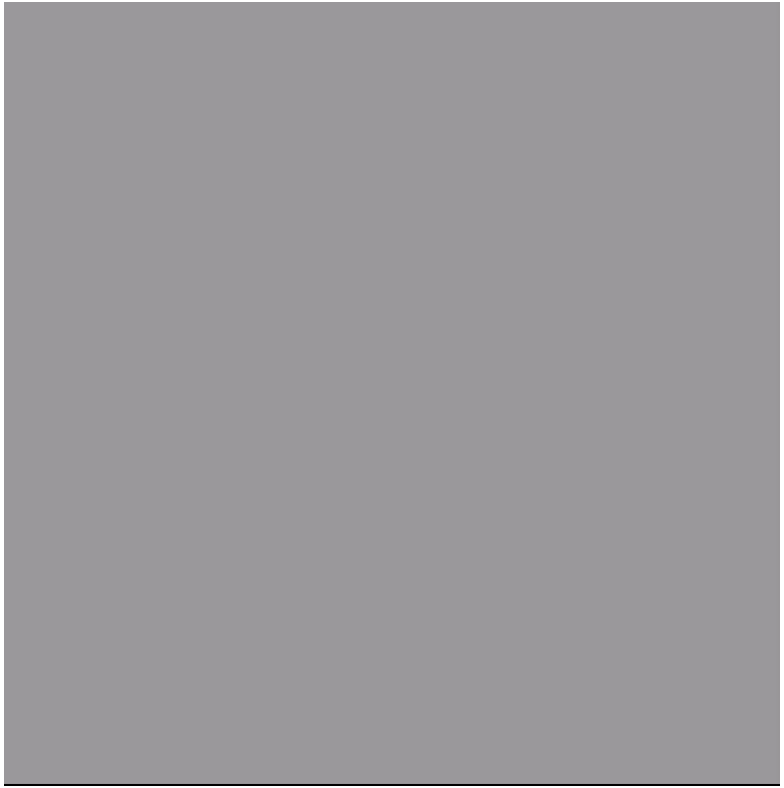
Paris PSVD meeting

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

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

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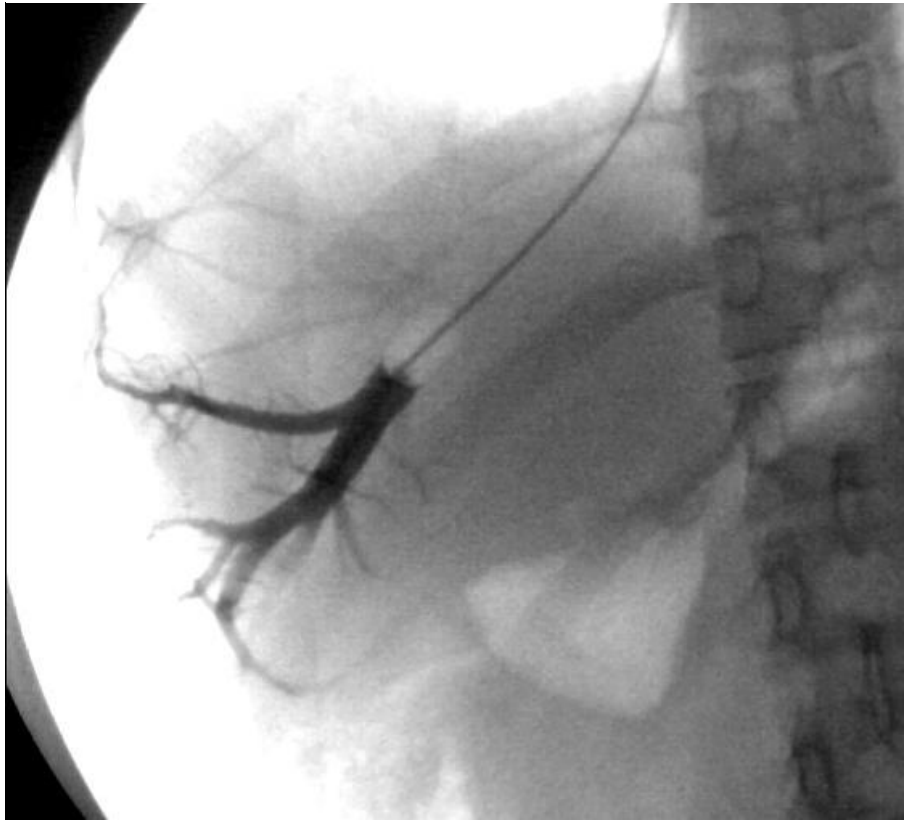
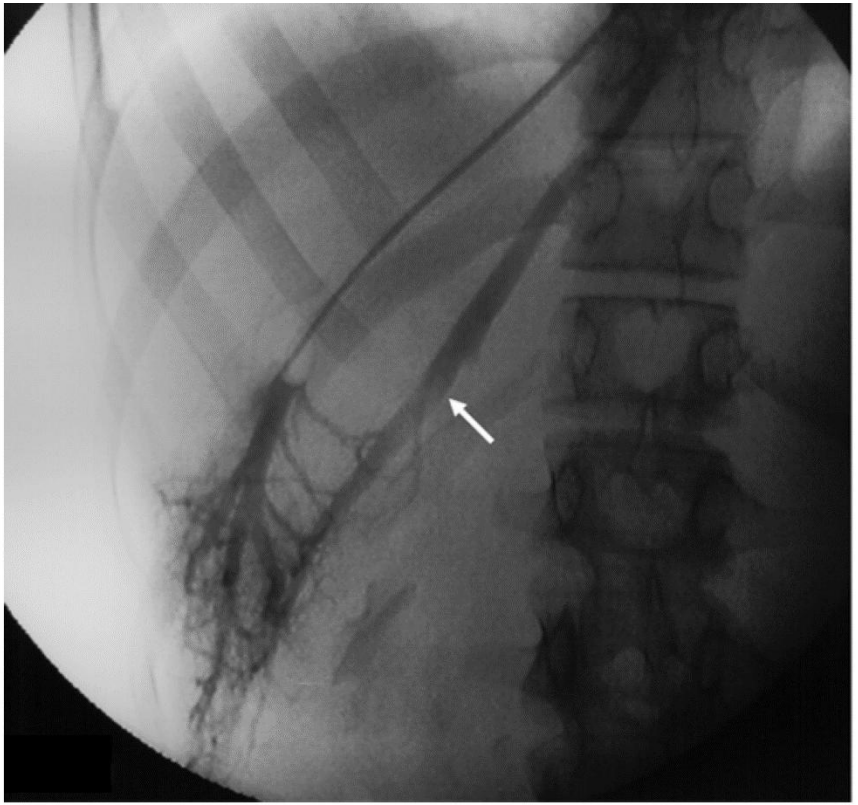
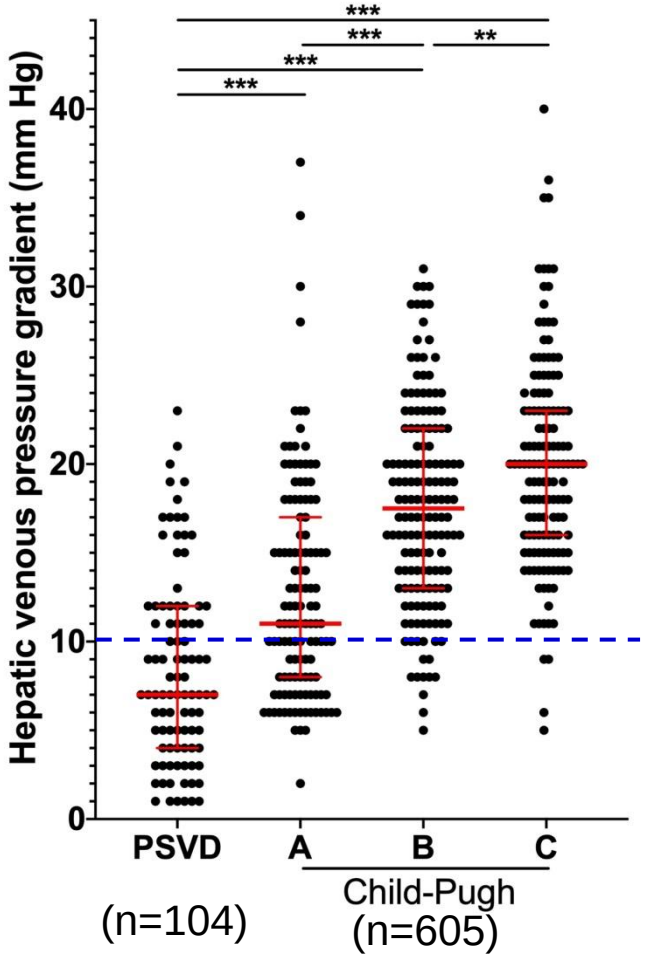


PSVD: 30 to 50%



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

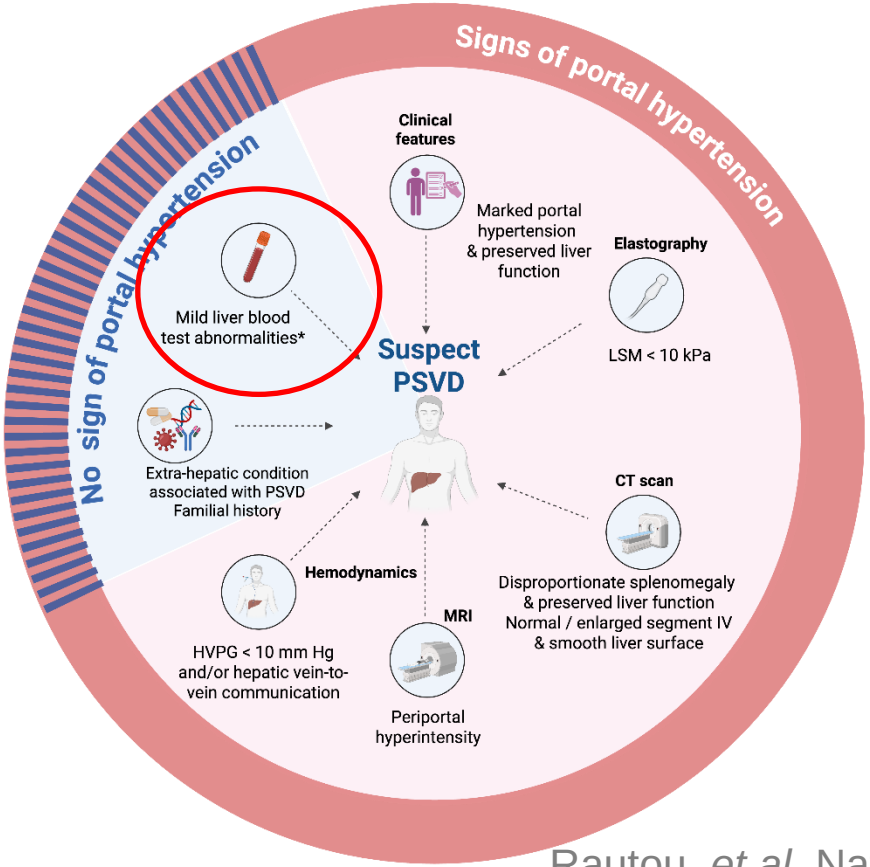
HVPG: PSVD vs. cirrhosis



Patients with signs of portal hypertension

Drilhon *et al*, ILC 2020

PSVD: when to suspect?

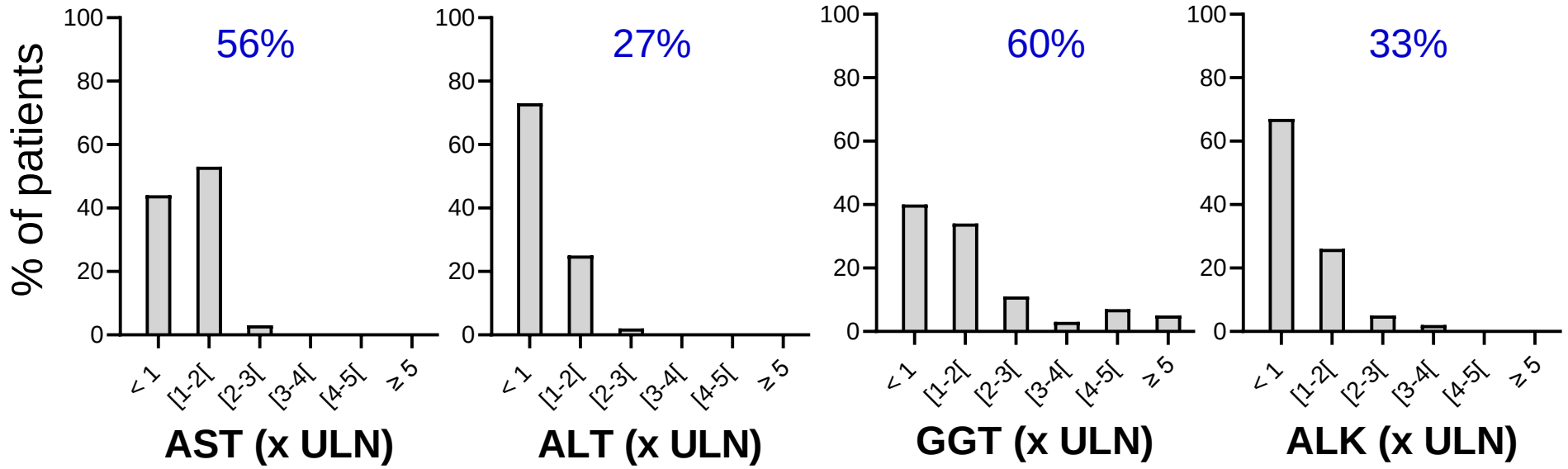


Paris PSVD meeting

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

PSVD: liver blood test

Abnormal values:



Data at inclusion in APIS trial (NCT04007289)

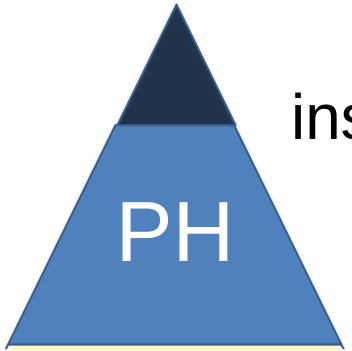
Similar in Magaz*, Giudicelli-Lett* *et al.* J Hepatol 2024

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

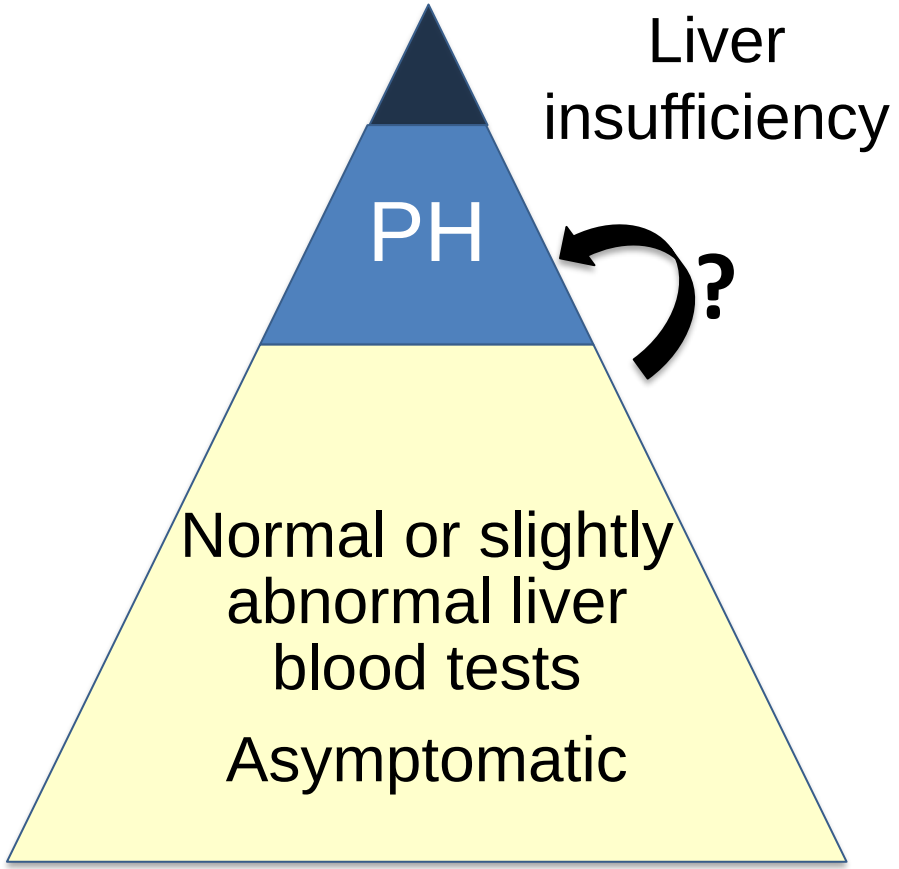
PSVD: manifestations



Liver insufficiency

Elkrief & Rautou, Liv Int 2016

PSVD: manifestations



Elkrief & Rautou, Liv Int 2016

Porto-sinusoidal vascular disorder

- Definition
- When to suspect?
- Outcome and treatment

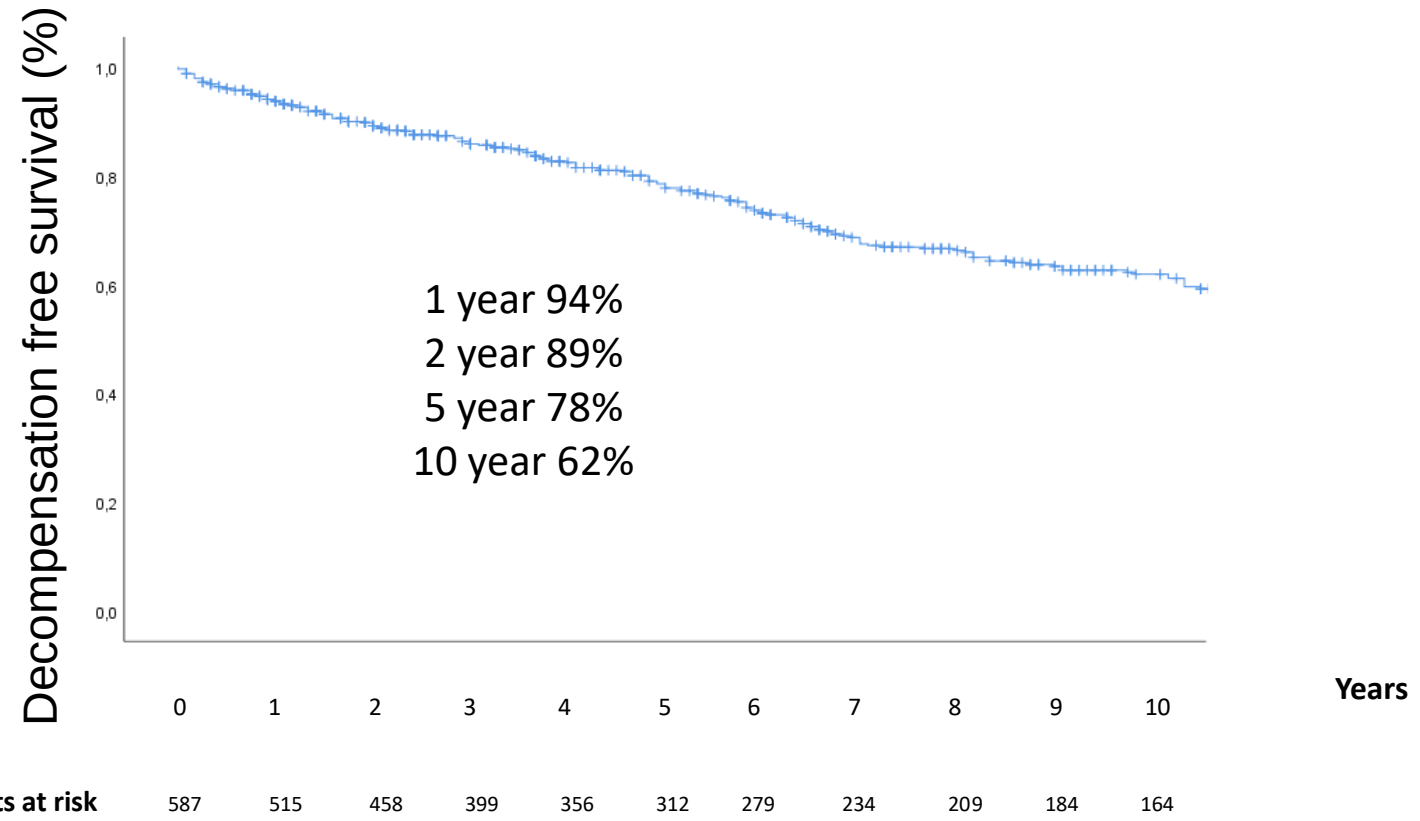
PSVD: outcome & complications

Gastrointestinal bleeding	**15% at 5 years (despite prophylaxis)**
Portal vein thrombosis	**16 % at 5 years**
Ascites	14% at 5 yrs (at the time of GI bleeding and easily controlled)
Hepatic encephalopathy	10%
Hepatocellular carcinoma	0.5%
Liver transplantation	9%
Death	19%

587 patients, 28 centres, median follow-up 6 years

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

PSVD: outcome & complications



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

PSVD: treatment

- Prophylaxis for variceal bleeding ≈ like in cirrhosis
  (Baveno VI)
- Anticoagulation?
- Diuretics if ascites
- Rarely: TIPSS or liver transplantation

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PSVD: conclusion

✓ **Suspect PSVD:**

- Unexplained liver blood test abnormalities
- No liver insufficiency
- Low liver stiffness measurement
- Low HVPG, hepatic collaterals
- Normal or ↑ segment IV + smooth surface

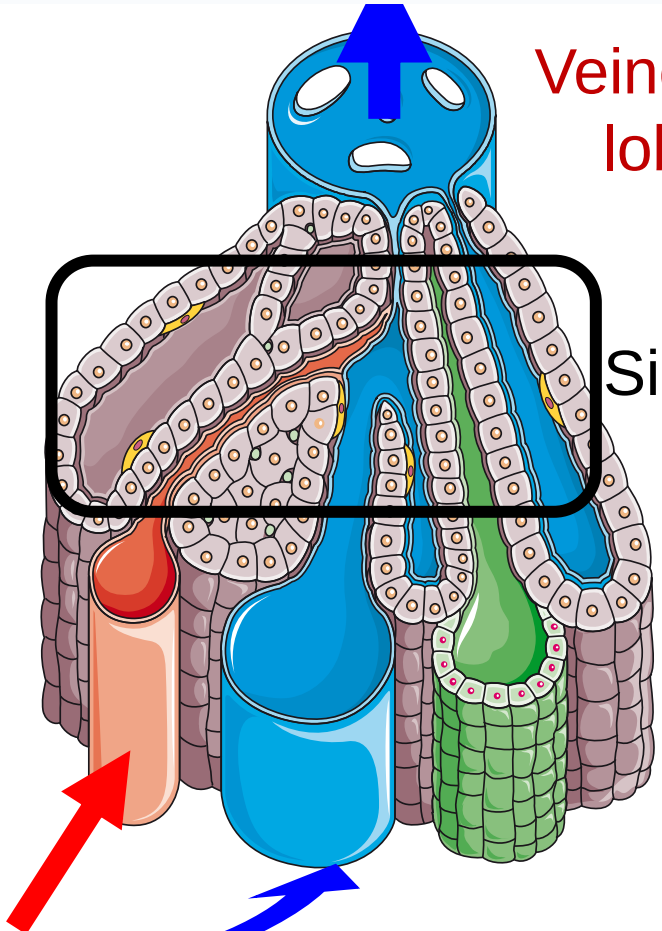
} **≠ signs of portal hypertension**

✓ **Outcome:**

- Portal vein thrombosis
- Variceal bleeding

Diseases affecting small intrahepatic vessels

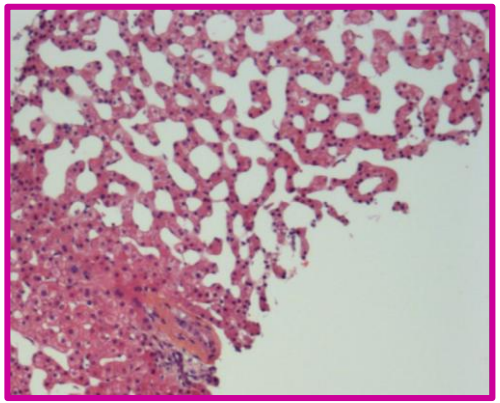
Dilatation sinusoïdale / Péliose



Dilatation sinusoïdale et péliose : définitions

Dilatation sinusoïdale

- > 1 travée hépatocytaire
- dans plusieurs lobules

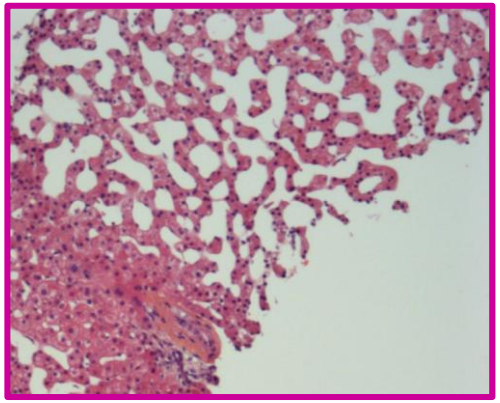


Marzano *et al.*, Hepatology 2015

Dilatation sinusoïdale et péliose : définitions

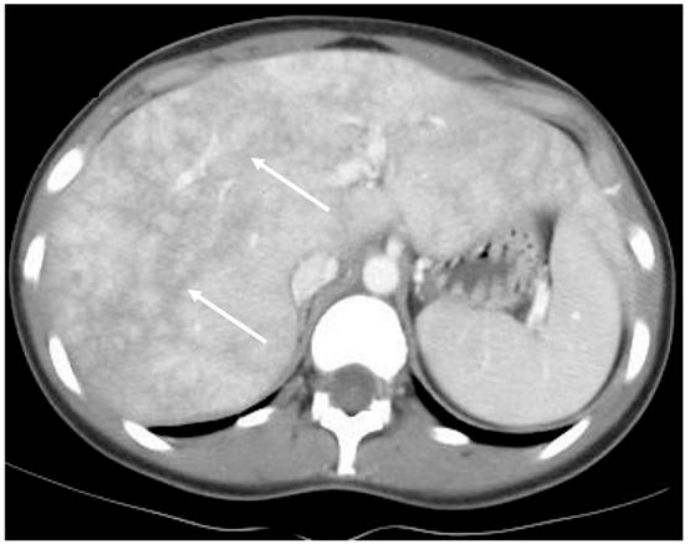
	Dilatation sinusoïdale	Péliose
Atrophie des hépatocytes	Oui	Oui
Rupture de l'endothélium	Non	Oui
Cavités hémorragique	Non	Oui

- > 1 travée hépatocytaire
- dans plusieurs lobules



Marzano *et al.*, Hepatology 2015

Dilatation sinusoïdale : détection



Aspect en mosaïque au temps portal et/ou artériel

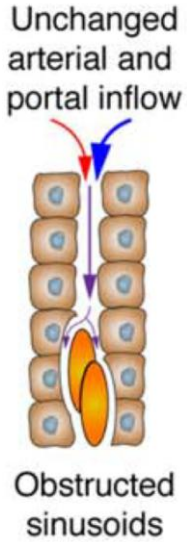
Preuve = biopsie

Ronot, Eur J Radiol 2016; Arrivé Eur J Radiol 2016

Dilatation sinusoïdale : causes

- Dilatation sinusoïdale **obstructive** :

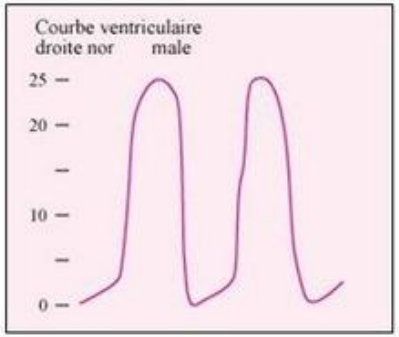
Modification hémodynamiques

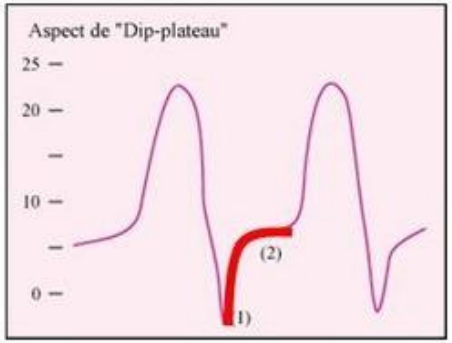


Marzano *et al.*, Hepatology 2015

Dilatation sinusoïdale : causes

- Dilatation sinusoïdale **obstructive** :
  - ✓ Budd-Chiari
  - ✓ Insuffisance cardiaque droite
  - ✓ Péricardite constrictive:
    - ➔ IRM cardiaque
    - ➔ cathétérisme cardiaque : DIP-plateau



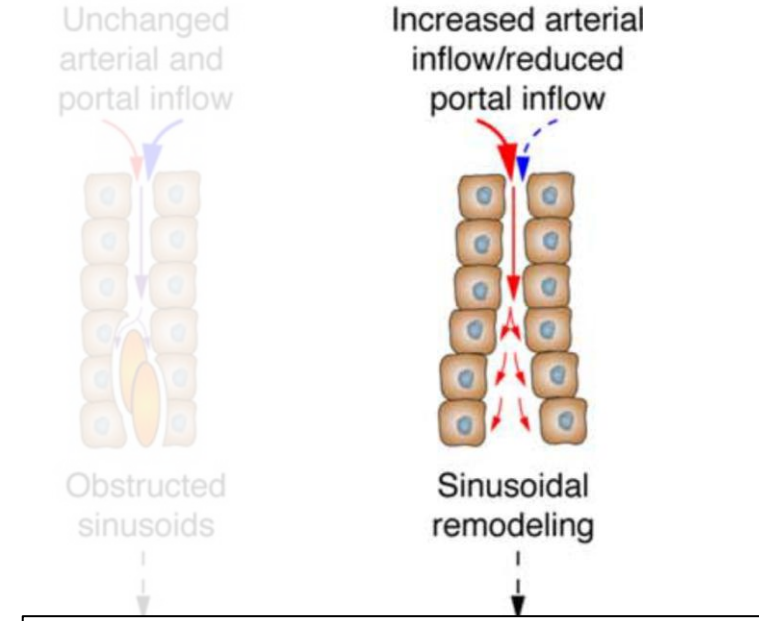


Marzano *et al.*, Hepatology 2015

Dilatation sinusoïdale : causes

- Dilatation sinusoïdale **non obstructive** :

Modification hémodynamiques



- Obstruction de la veine porte
- MVPS
- Shunts portosystémiques congénitaux

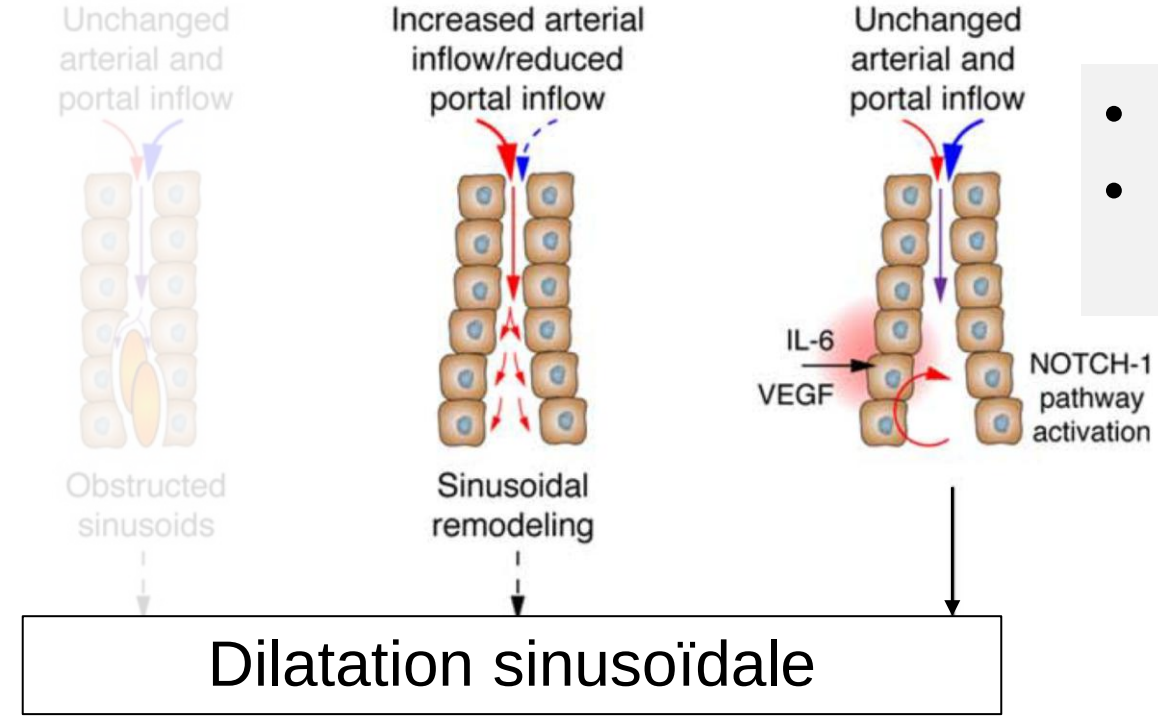
Dilatation sinusoïdale

Marzano *et al.*, Hepatology 2015

Dilatation sinusoïdale : causes

- Dilatation sinusoïdale **non obstructive** :

Modification hémodynamiques | Facteurs solubles



- Néoplasies avec SIRS
- Maladies inflammatoires (infections inclues)

Marzano *et al.*, Hepatology 2015

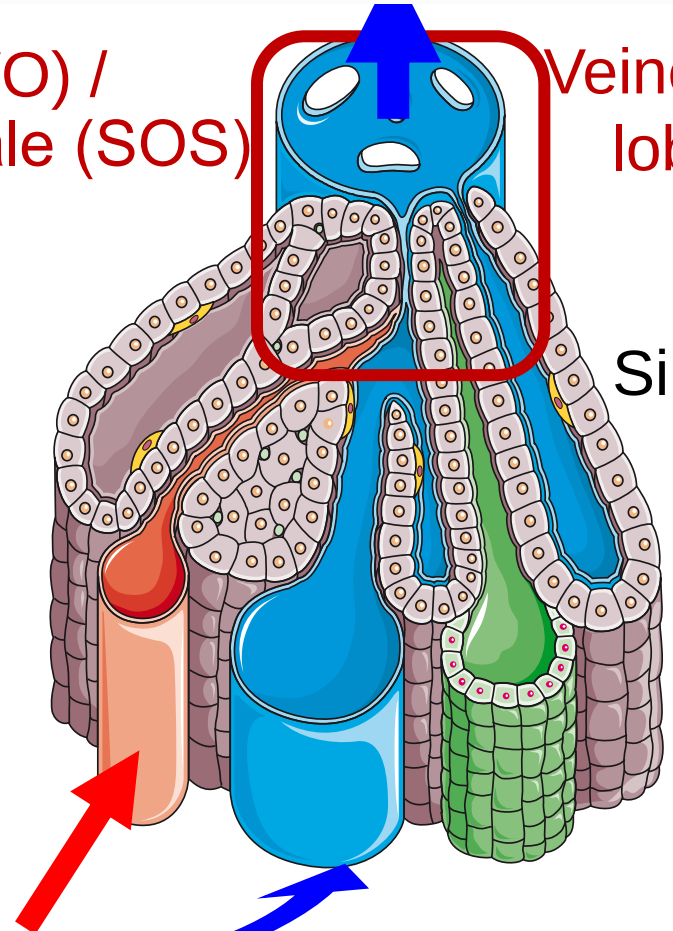
Dilatation sinusoïdale : évolution

- Régression possible si facteur aigu
- Evolution vers MVPS ?

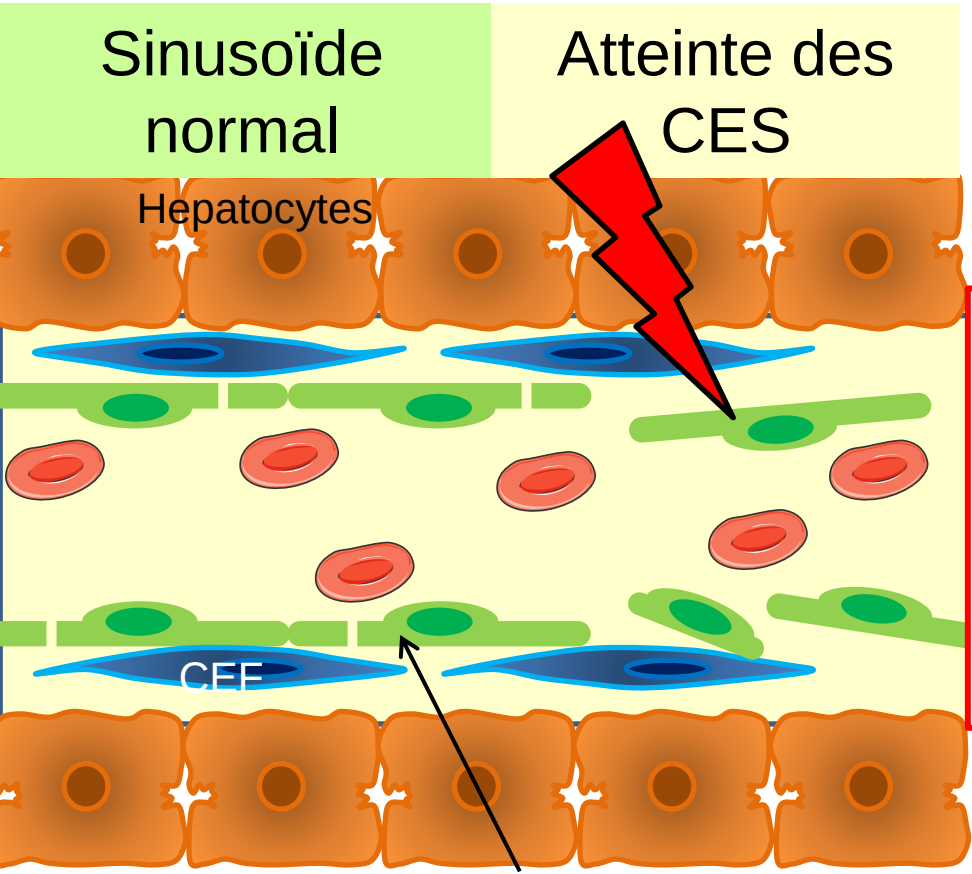
Ronot, Eur J Radiol 2016

Diseases affecting small intrahepatic vessels

Maladie veino-occlusive (MVO) /
Syndrome d'obstruction sinusoïdale (SOS)



SOS / MVO post-greffe de moelle : mécanisme



Conditionnement de greffe de moelle :

- Busulfan,
- Cyclophosphamide
- Irradiation

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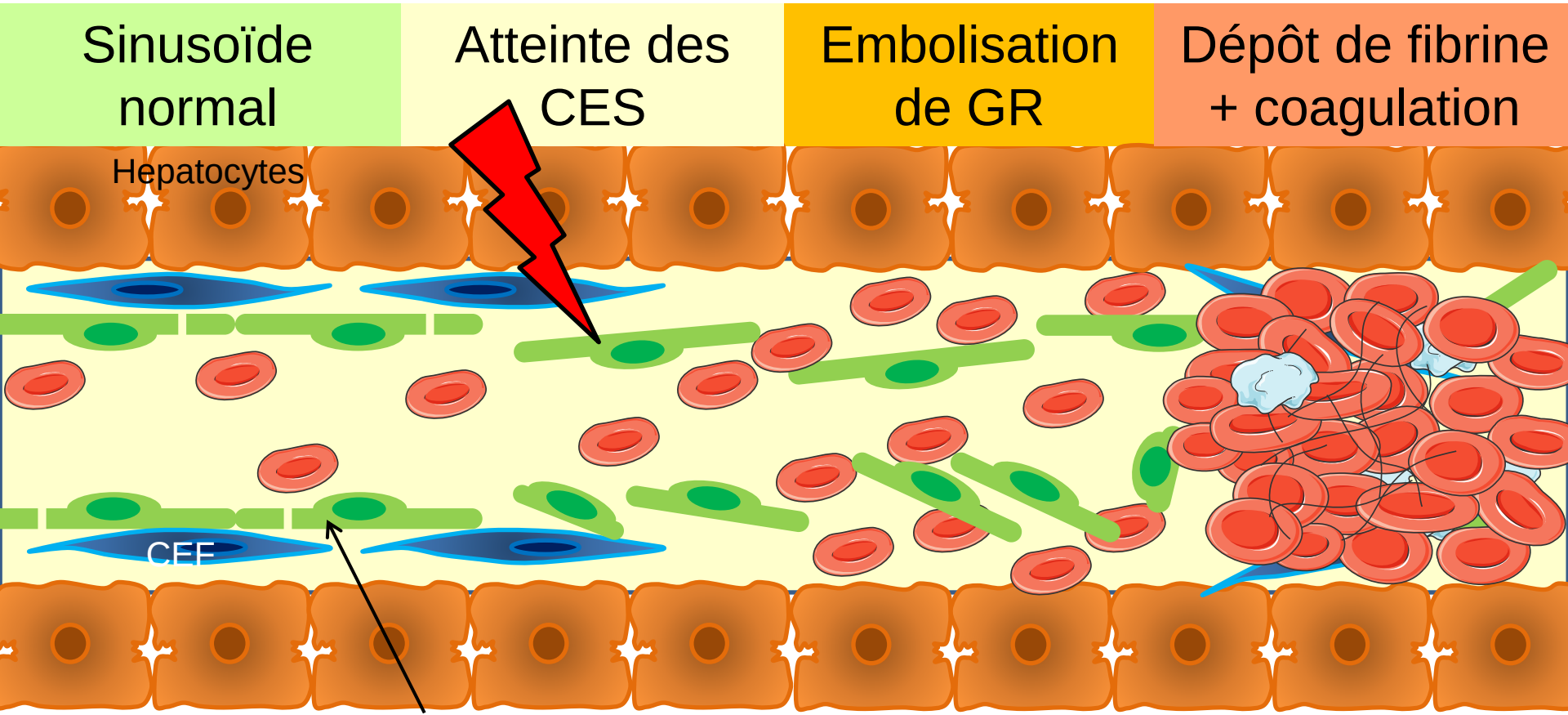
SOS / MVO post-greffe de moelle : mécanisme



SOS / MVO post greffe de moëlle

- Dans les semaines après greffe de moelle
  - ✓ Ictère
  - ✓ Hépatomégalie ou hépatalgie
  - ✓ Prise poids

EASL guidelines on vascular liver diseases 2026

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

Apply via the COST website

Instituts thématiques

Inserm
Institut national
de la santé et de la recherche médicale

Université Paris Cité

C·R·I
CENTRE DE RECHERCHE SUR L'INFLAMMATION

Hepatology unit, Beaujon Hospital
Reference center for vascular liver diseases

Inserm U1149
www.rautoulab.com/
Team "*vessels in liver diseases*"

anr agence nationale de la recherche

EF Clif
EUROPEAN FOUNDATION FOR THE STUDY OF CHRONIC LIVER FAILURE

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AMVF

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Reconnue d'utilité publique

FRM
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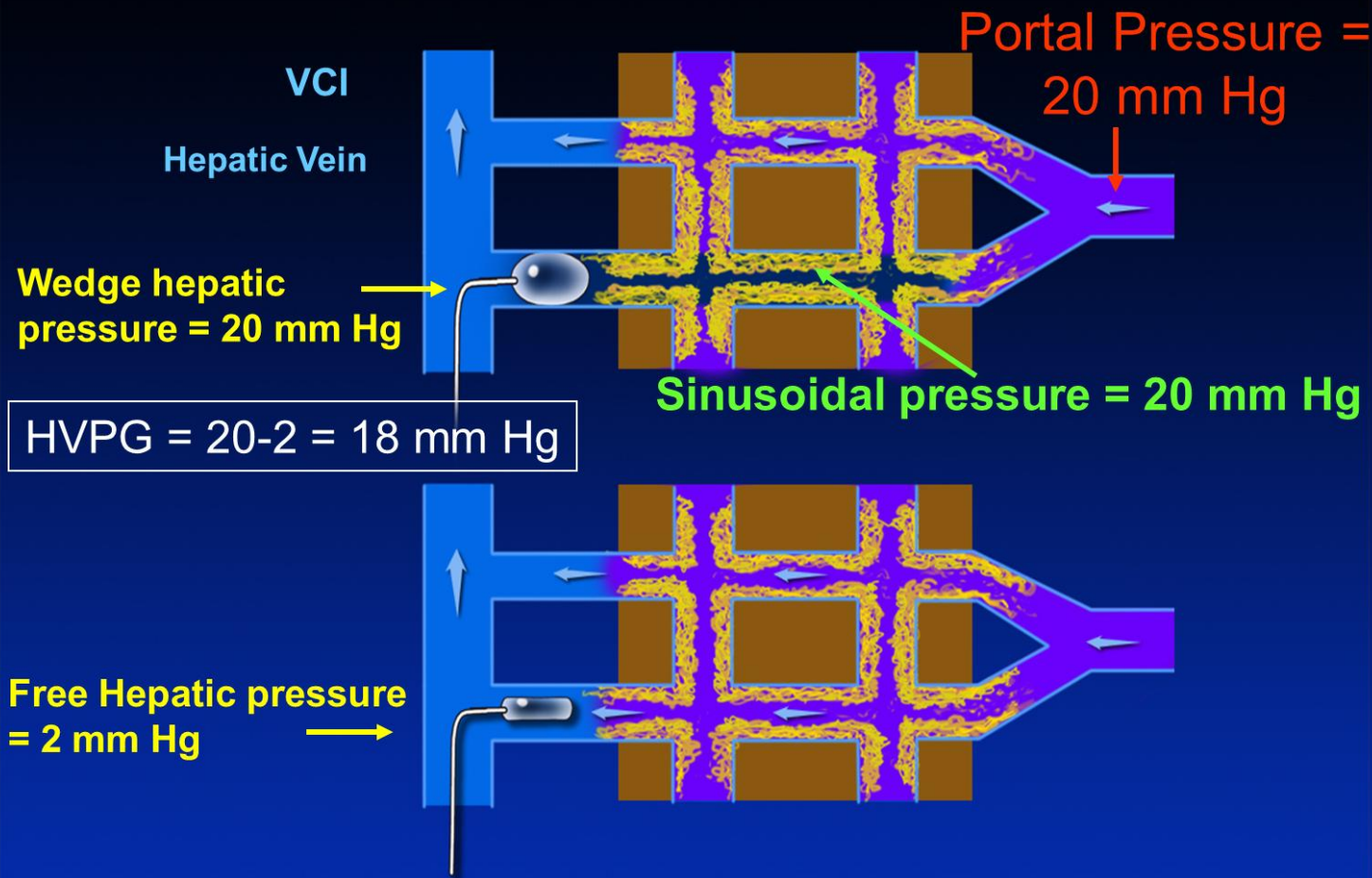
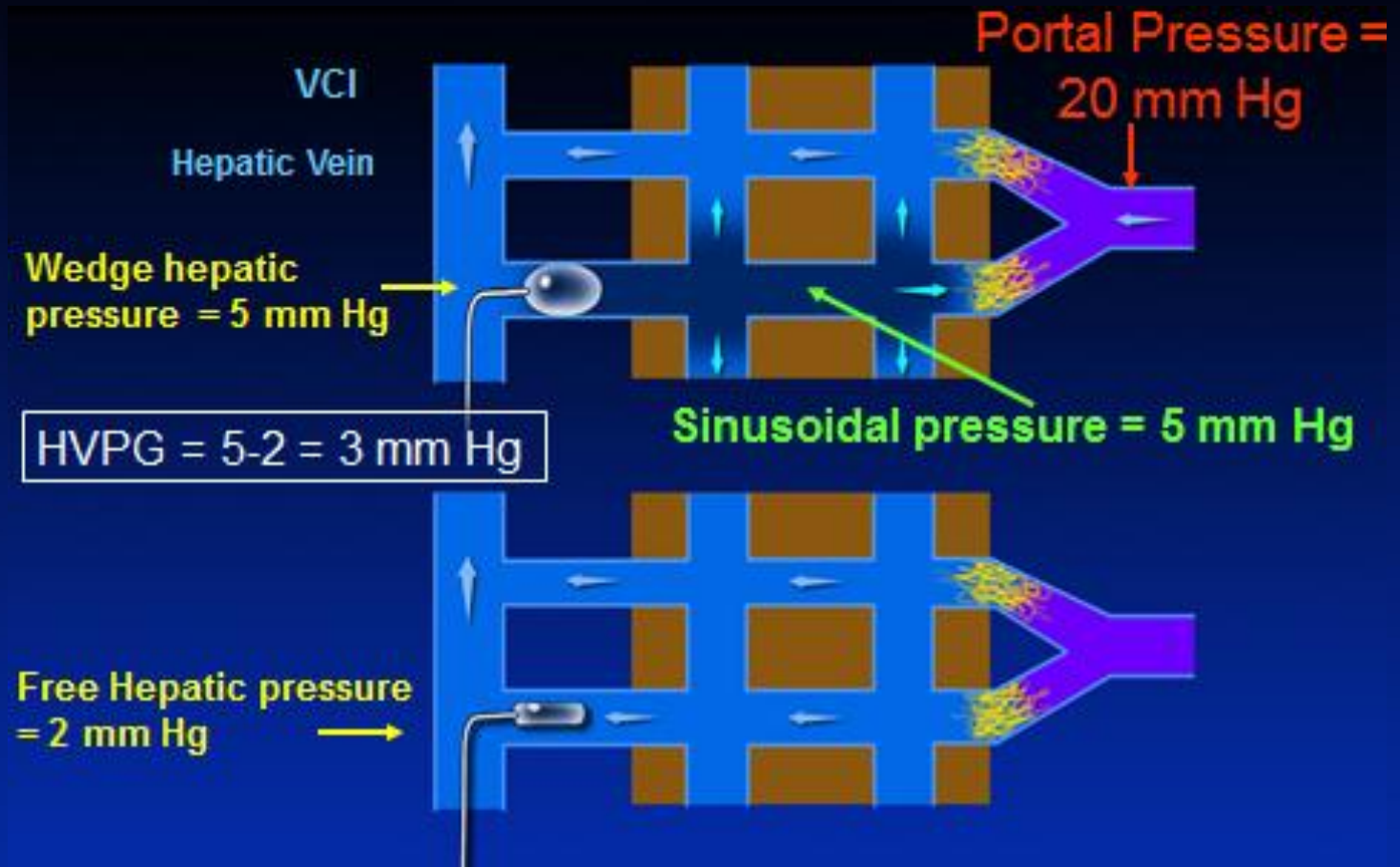
Hepatic hemodynamics: cirrhosis



Hepatic hemodynamics: PSVD

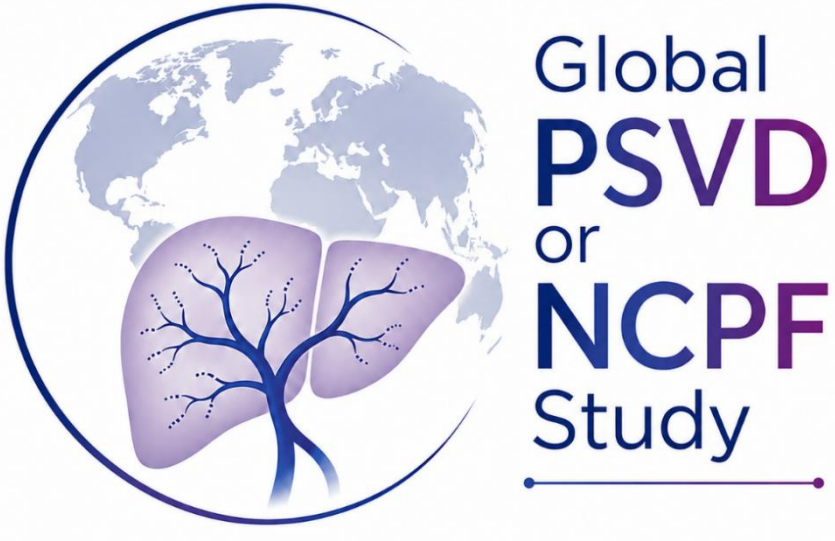


Conclusion & Next step

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