Porto-sinusoidal vascular disorder: case-based discussion

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Case presentation

- 26 yr-old man; **2 episodes of variceal bleeding**

- Medical history:
  - ✓ Dyslipidemia
  - ✓ Appendectomy

- Lifestyle:
  - ✓ No alcohol,
  - ✓ No drug nor substance
  - ✓ 65 kg; 1.72 m

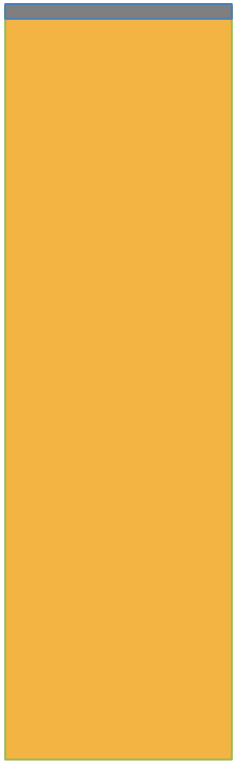
Laboratory tests:
- AST: 30 UI/L
- ALT: 36 UI/L
- ALK: 80 UI/L
- **GGT: 80 UI/L**
- **Platelet: 88 000 /mm^3**
- Serum bilirubin: 10 µmol/L
- PT: 73%
- Serum creatinine: 73 µmol/L

What would make you suspect PSVD?

Porto-sinusoidal vascular disorder

- When to suspect?
- Definition
- Associated conditions
- Outcome

PSVD among cirrhosis

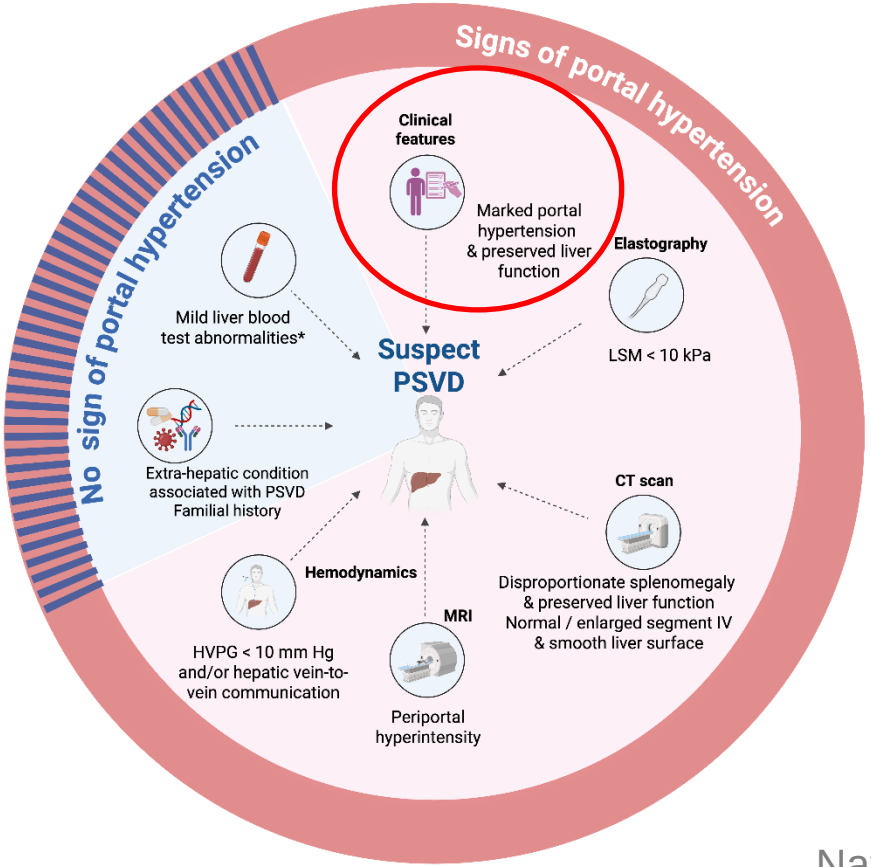


PSVD
0.4%

Cirrhosis
99.6%

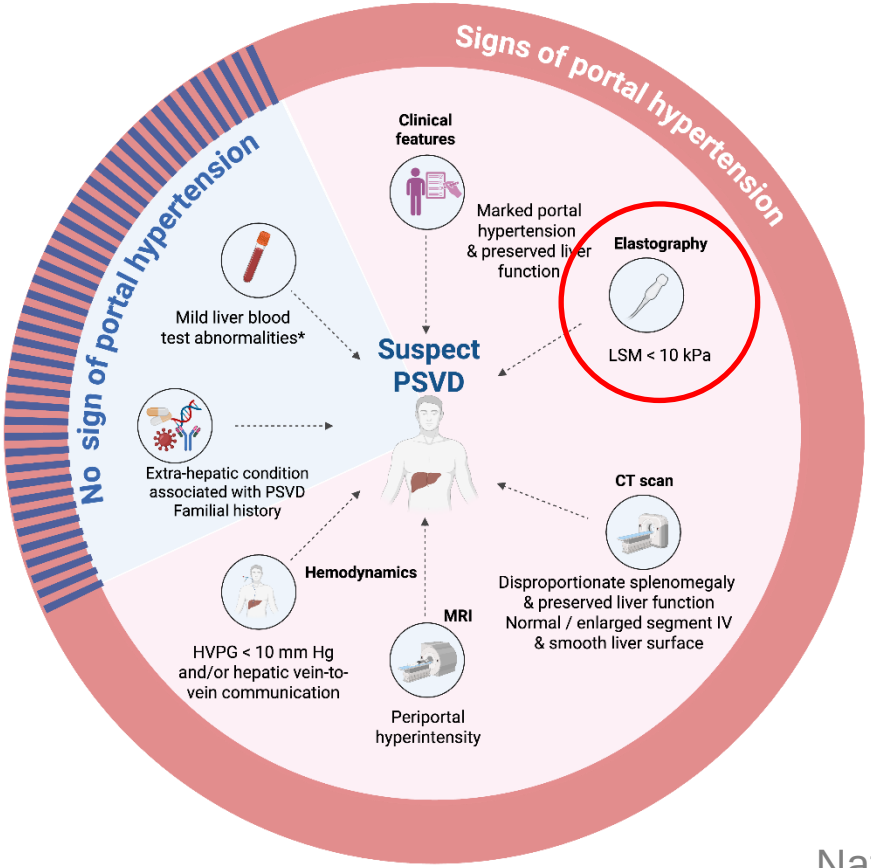
➔ Screening tools needed

PSVD: when to suspect?



Paris PSVD meeting
Nat Rev Gastro Hepato 2026

PSVD: when to suspect?



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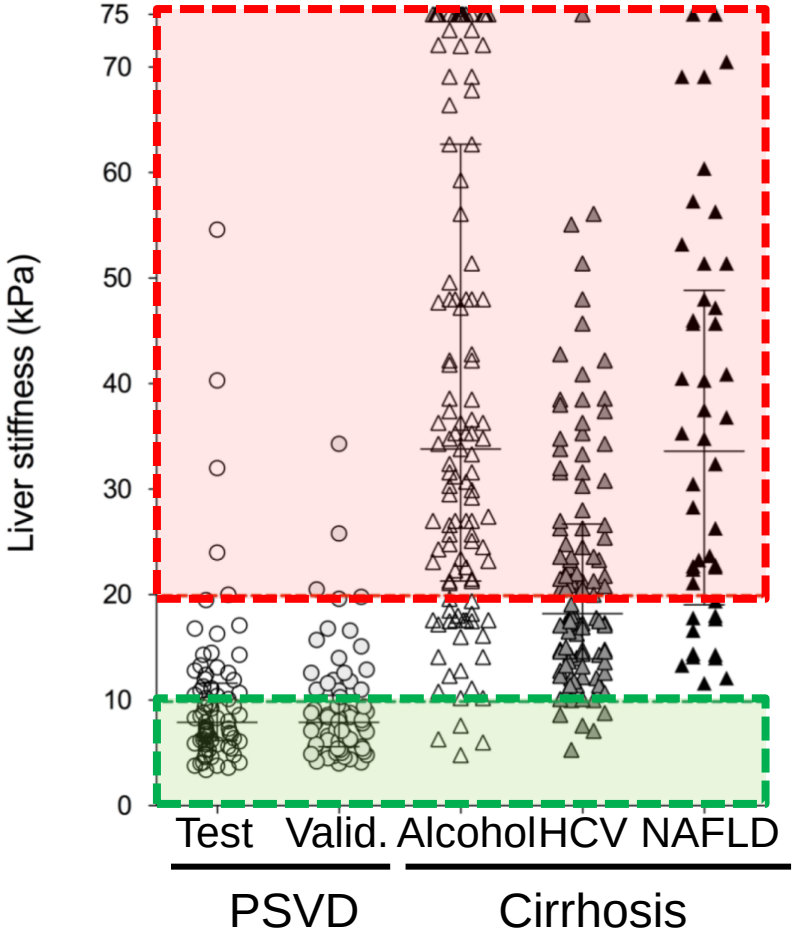
PSVD: liver stiffness measurement (TE)



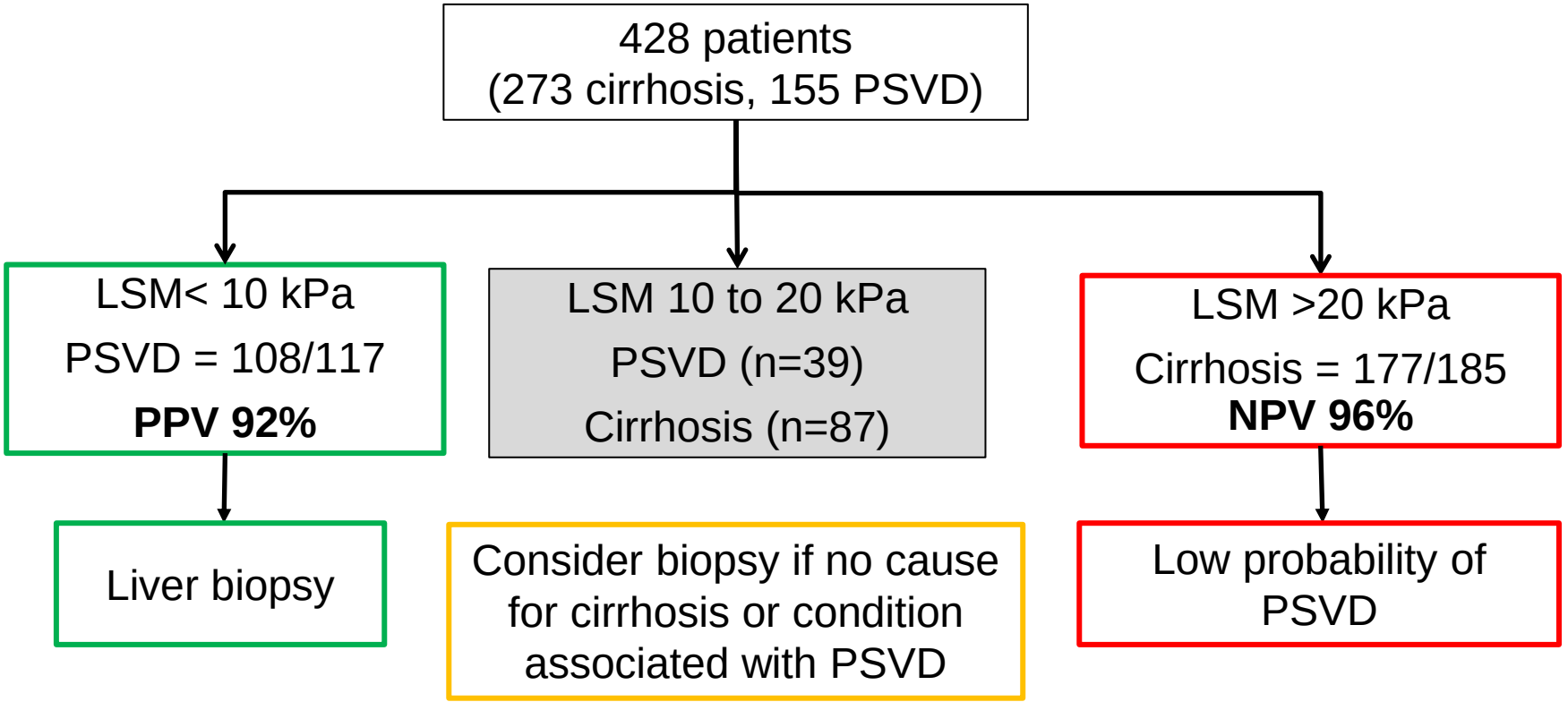
Patients with signs of portal hypertension
155 PSVD, 273 cirrhosis

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

Similar results in:

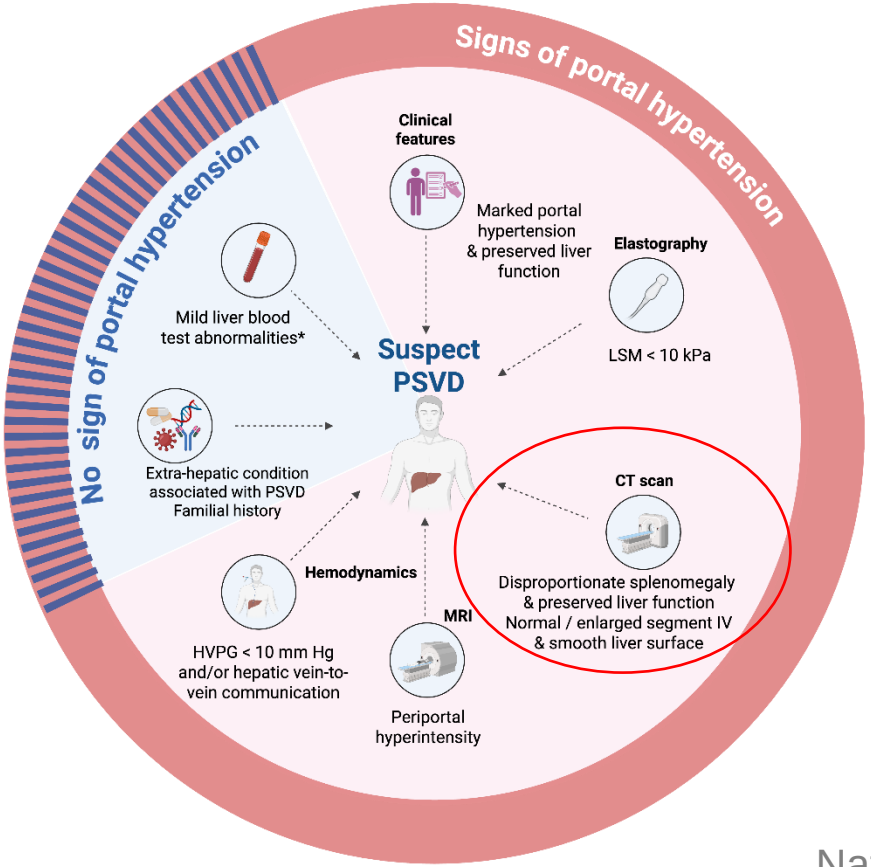
- Laharie Eur J Gastroenterol Hepatol 2010
- Seijo Dig Liver Dis 2012
- Sharma J Clin Exp Hepatol 2017
- Ferreira-Silva J, Dig Liver Dis. 2022

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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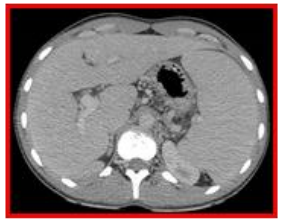
PSVD: CT-scan



50 patients with PSVD

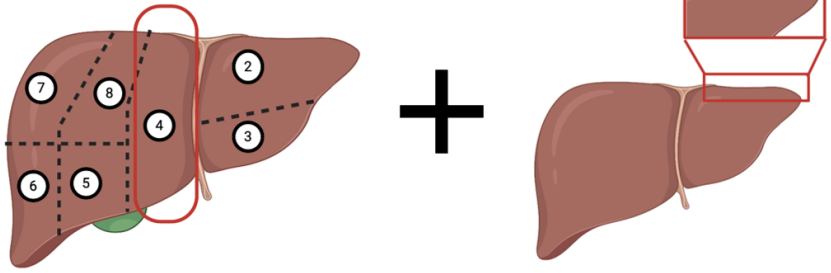
Matched on ascites

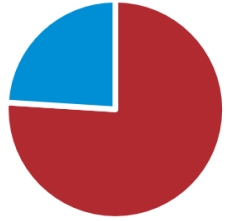
100 patients with cirrhosis

CT features for PSVD diagnosis

No atrophy of segment IV

LSN < 2.5





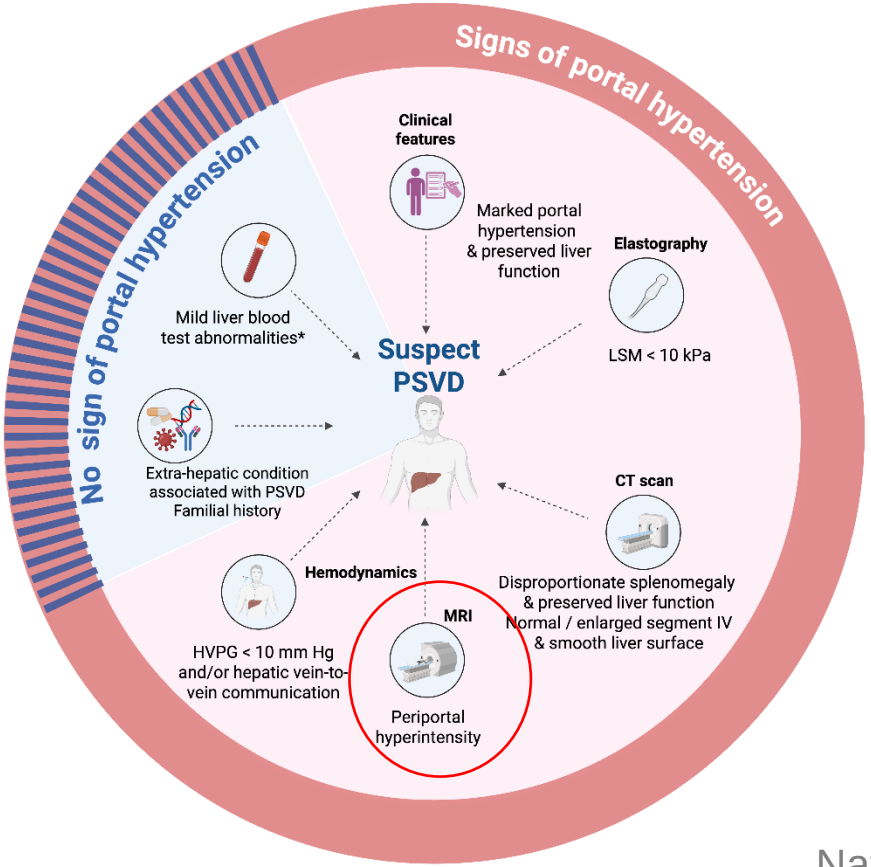
With the features

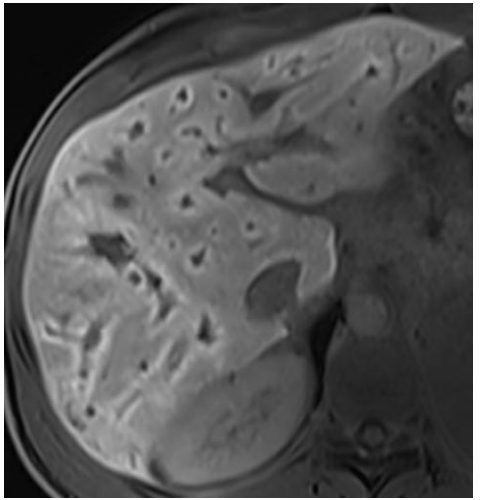
PSVD Cirrhosis

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

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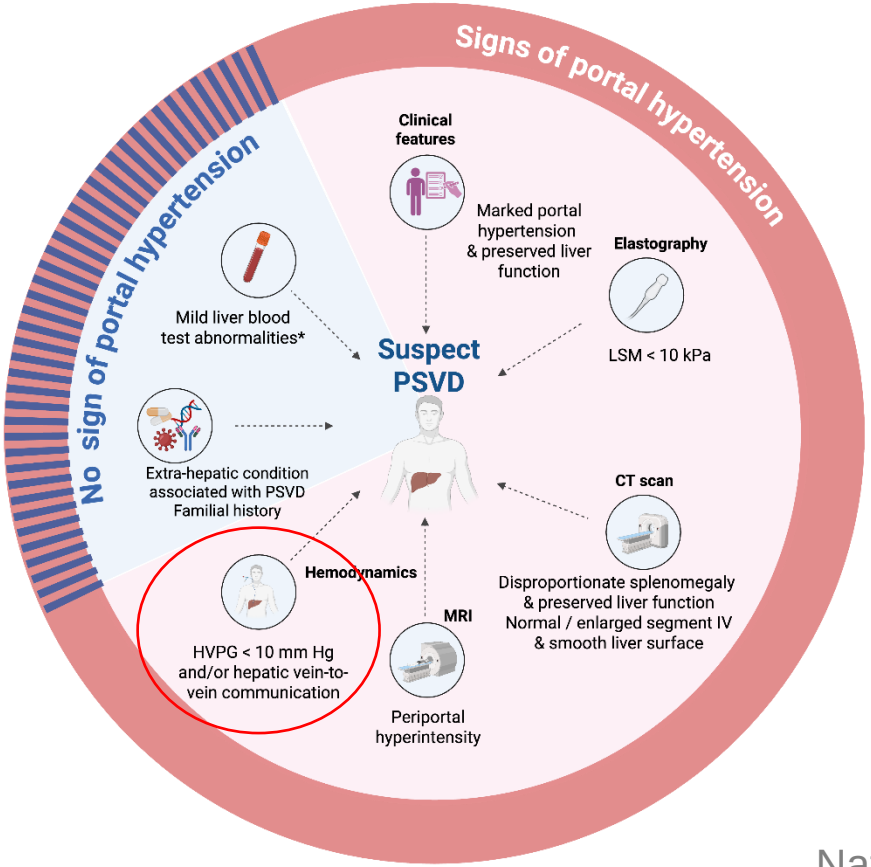
PSVD: when to suspect?



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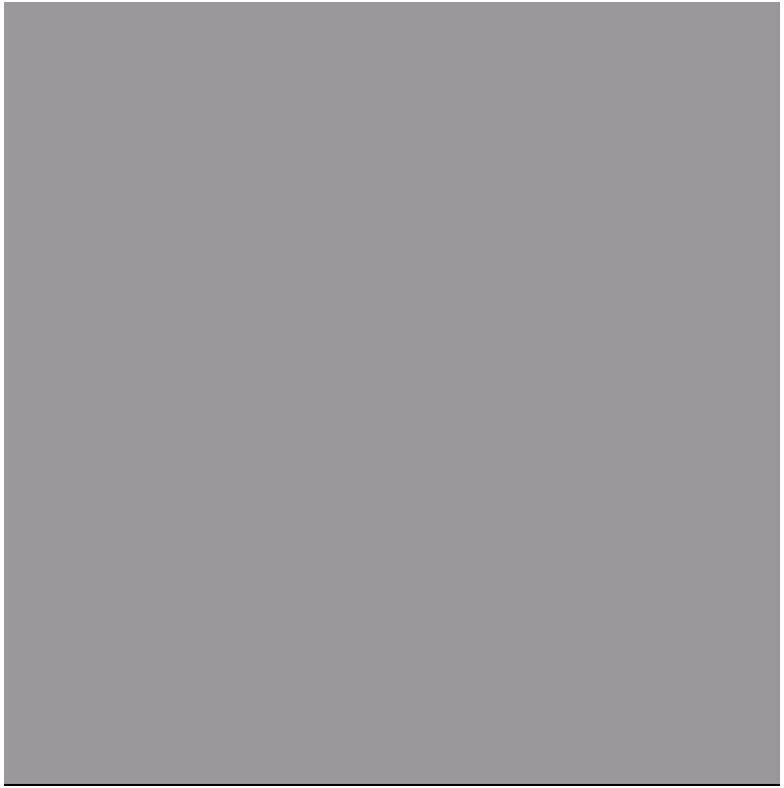
Hepatic veno-venous communications

Cirrhosis: 3%

PSVD: 30 to 50%

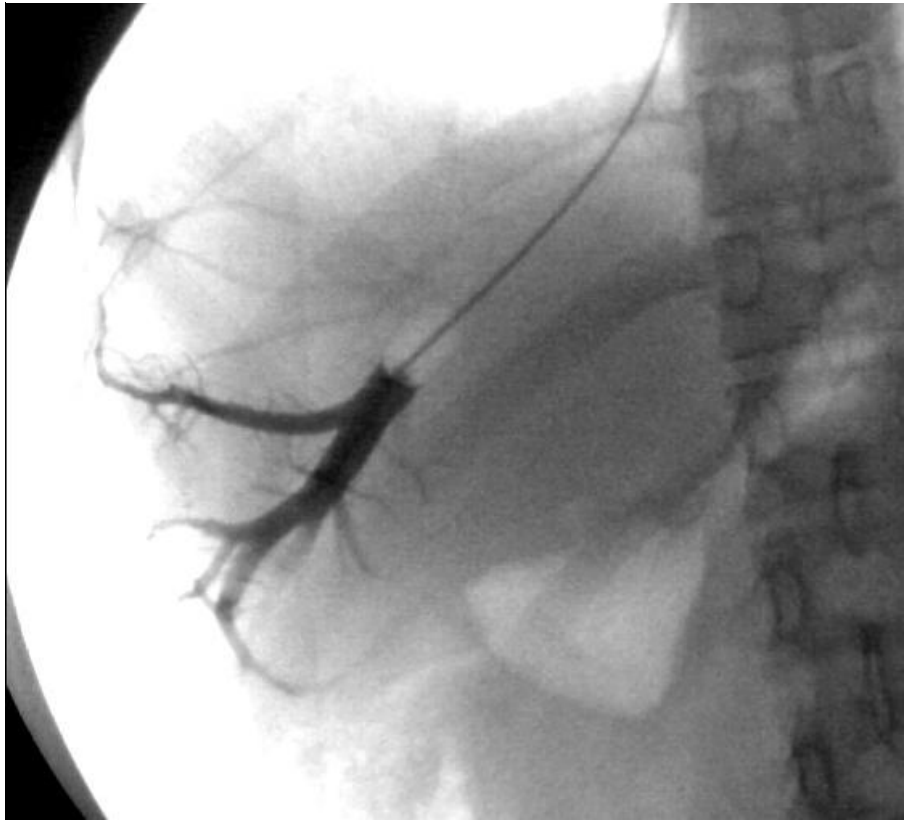


Drilhon et al; ILC 2020

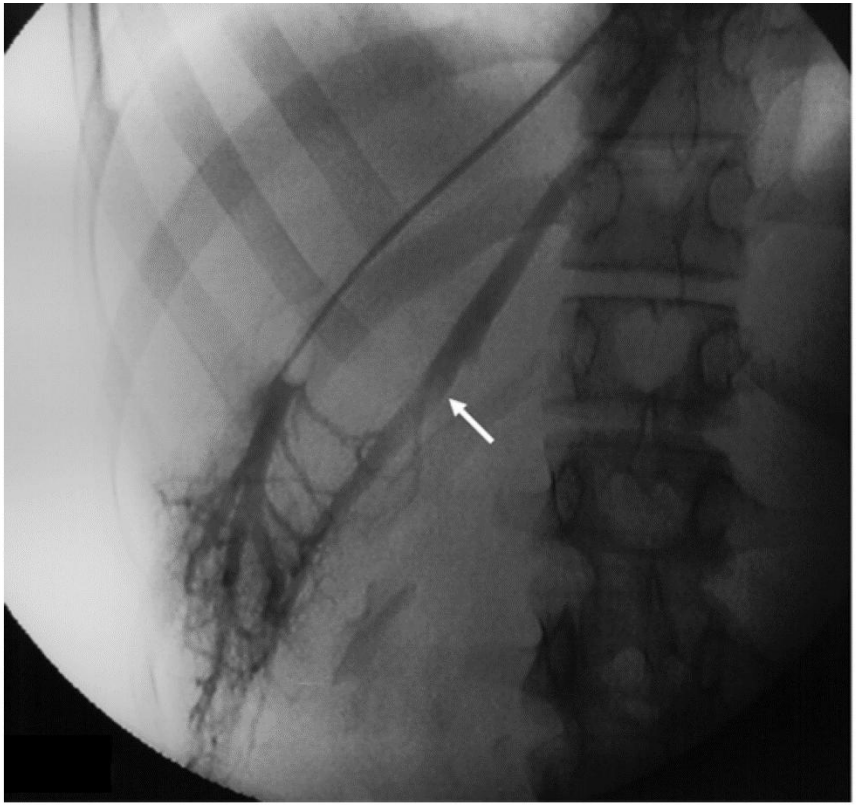
Seijo *et al.* Dig Liv Dis 2012

Hepatic veno-venous communications

Cirrhosis: 3%

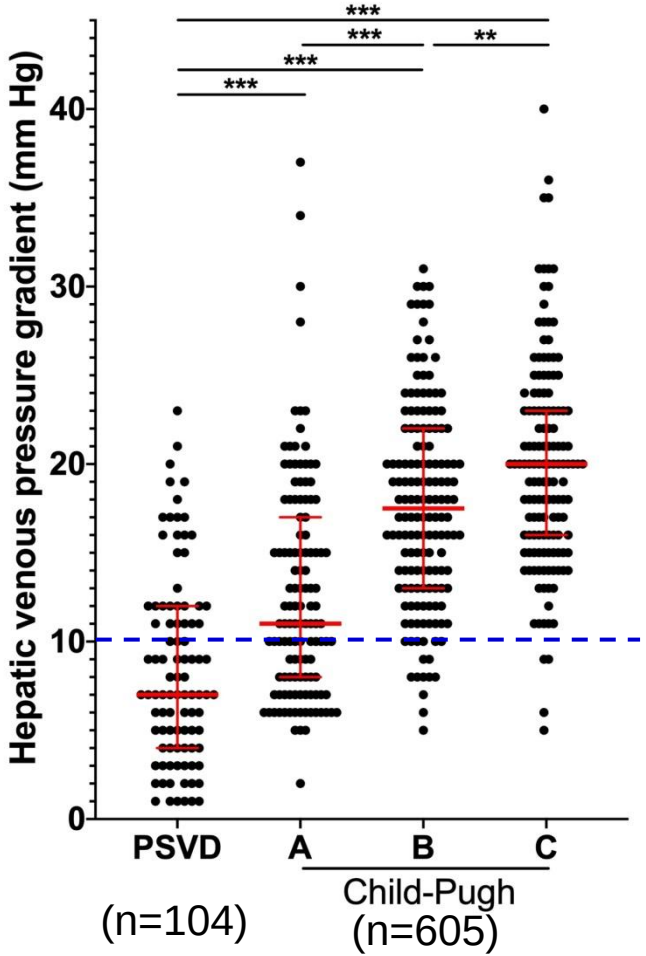


PSVD: 30 to 50%



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

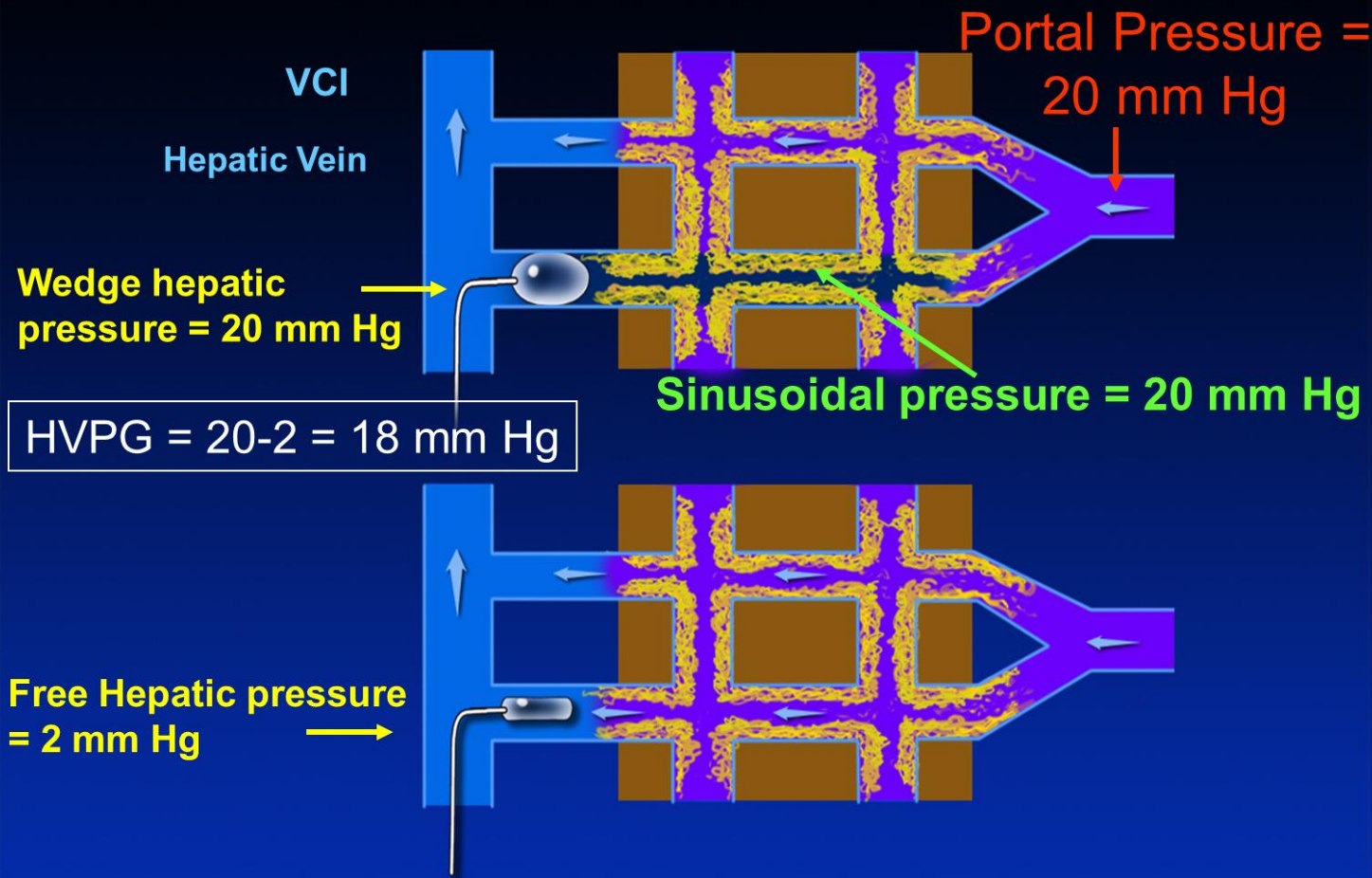
HVPG: PSVD vs. cirrhosis



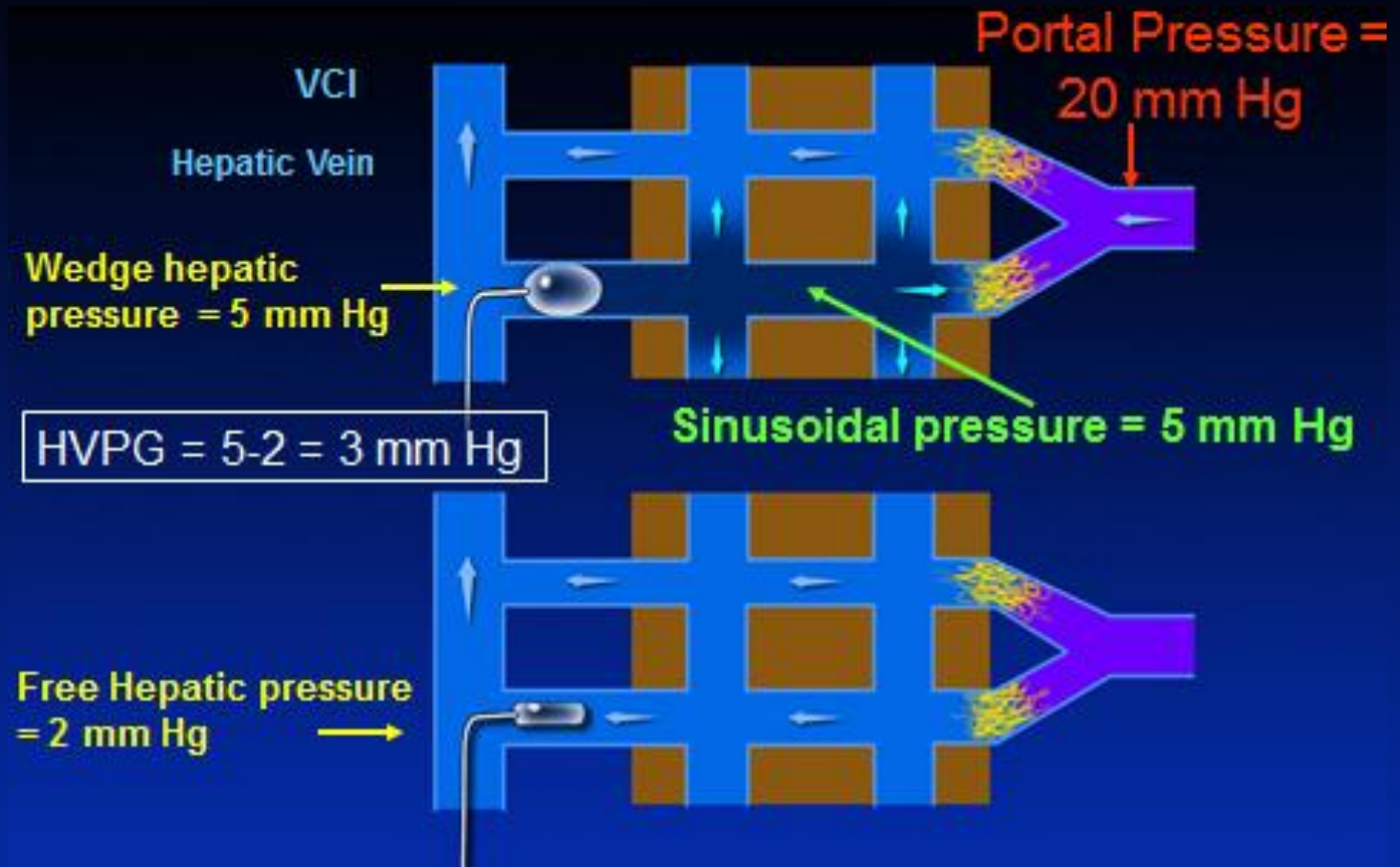
Patients with signs of portal hypertension

Drilhon *et al*, ILC 2020

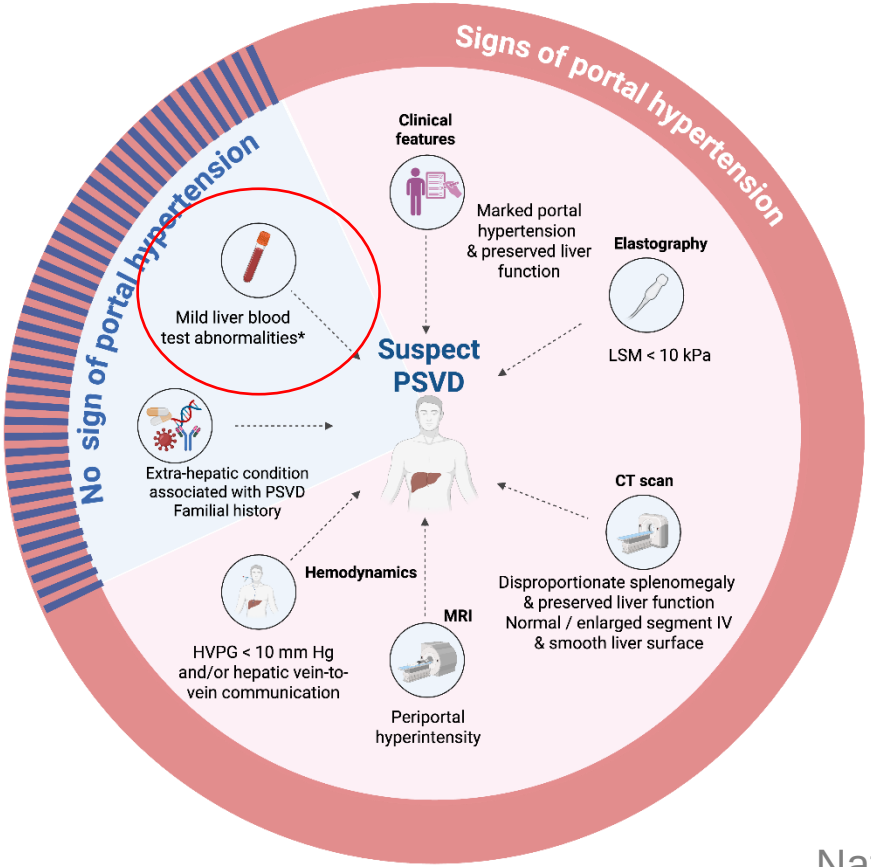
Hepatic hemodynamics: cirrhosis



Hepatic hemodynamics: PSVD



PSVD: when to suspect?



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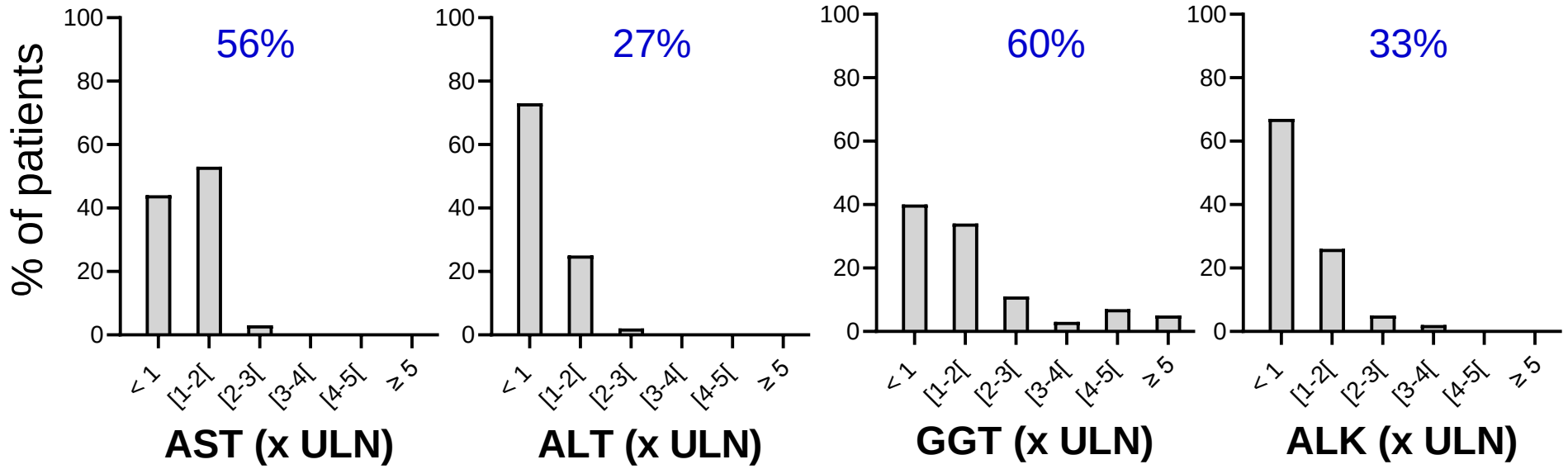
PSVD: liver blood test

Abnormal values:



Data at inclusion in APIS trial (NCT04007289)

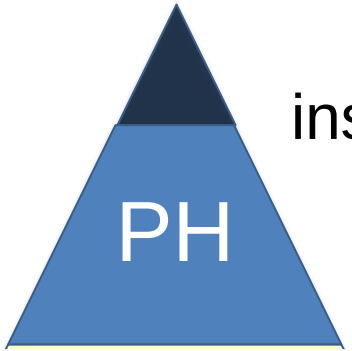
Similar in Magaz*, Giudicelli-Lett* *et al.* J Hepatol 2024

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

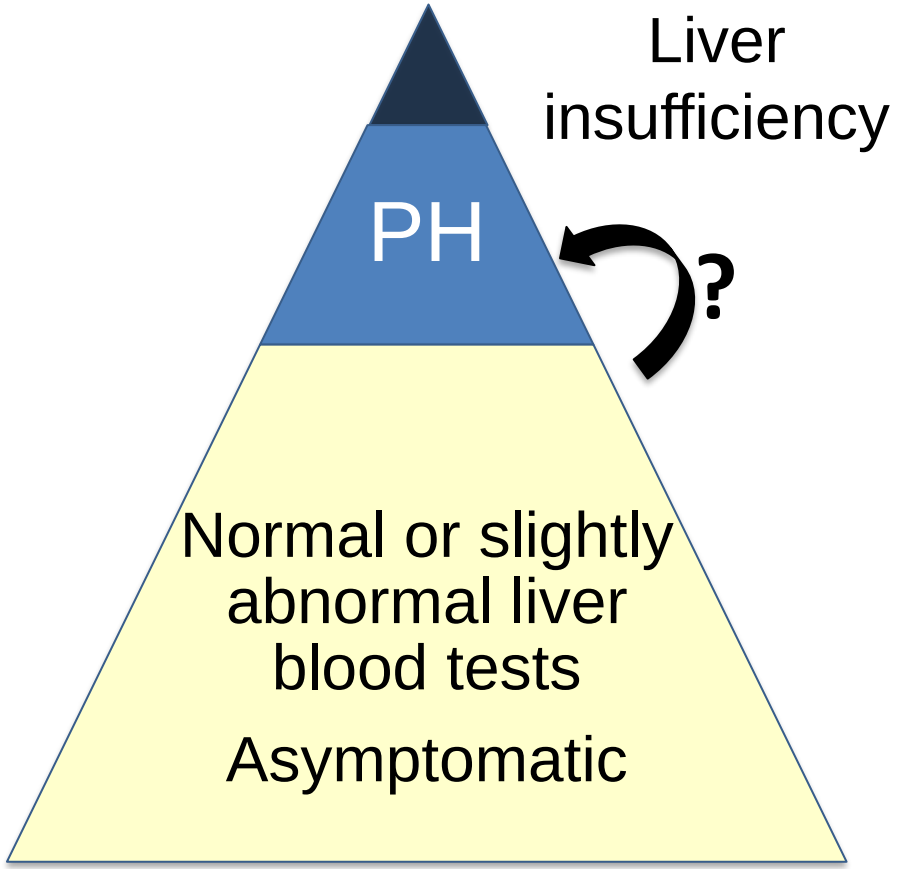
PSVD: manifestations



Liver insufficiency

Elkrief & Rautou, Liv Int 2016

PSVD: manifestations



Elkrief & Rautou, Liv Int 2016

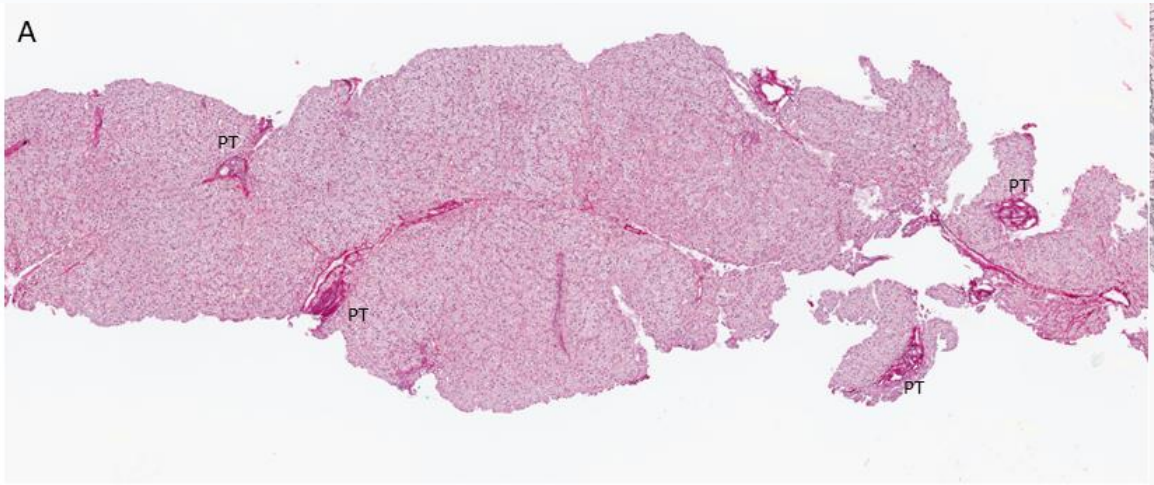
Case presentation

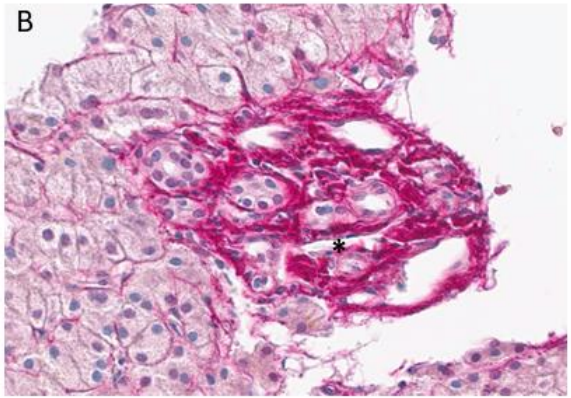
- **CT-scan:**
  - ✓ no portal vein thrombosis
  - ✓ normal segment IV; smooth liver surface
  - ✓ splenomegaly

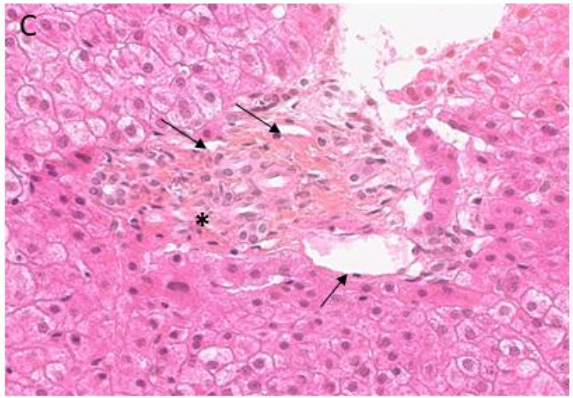
- **Work-up for causes “negative”:**
  - ✓ No hepatitis B or C virus infection
  - ✓ Normal α antitrypsin, ceruloplasmin, angiotensin conv. Enz.
  - ✓ Only dyslipidaemia
  - ✓ Only autoimmune Ab: anti–SMA (1/80); normal serum Ig

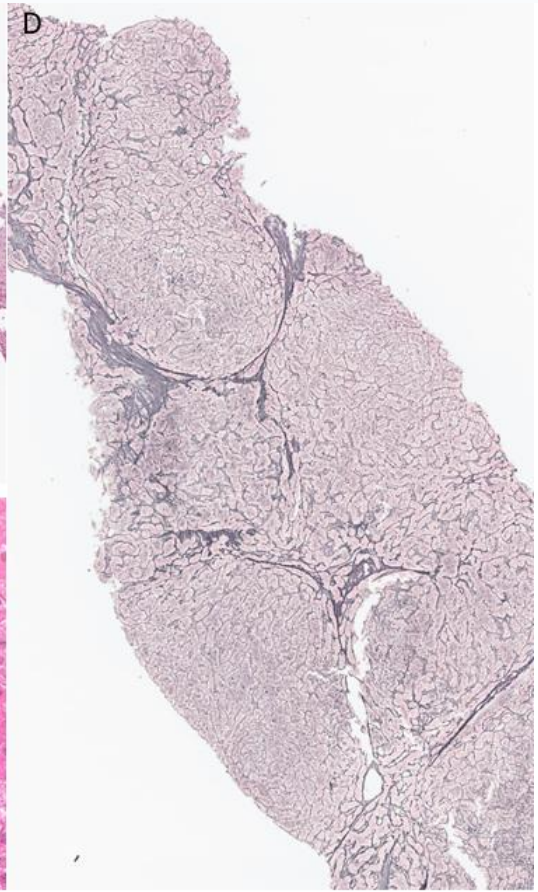
What approach do you propose?

Case presentation

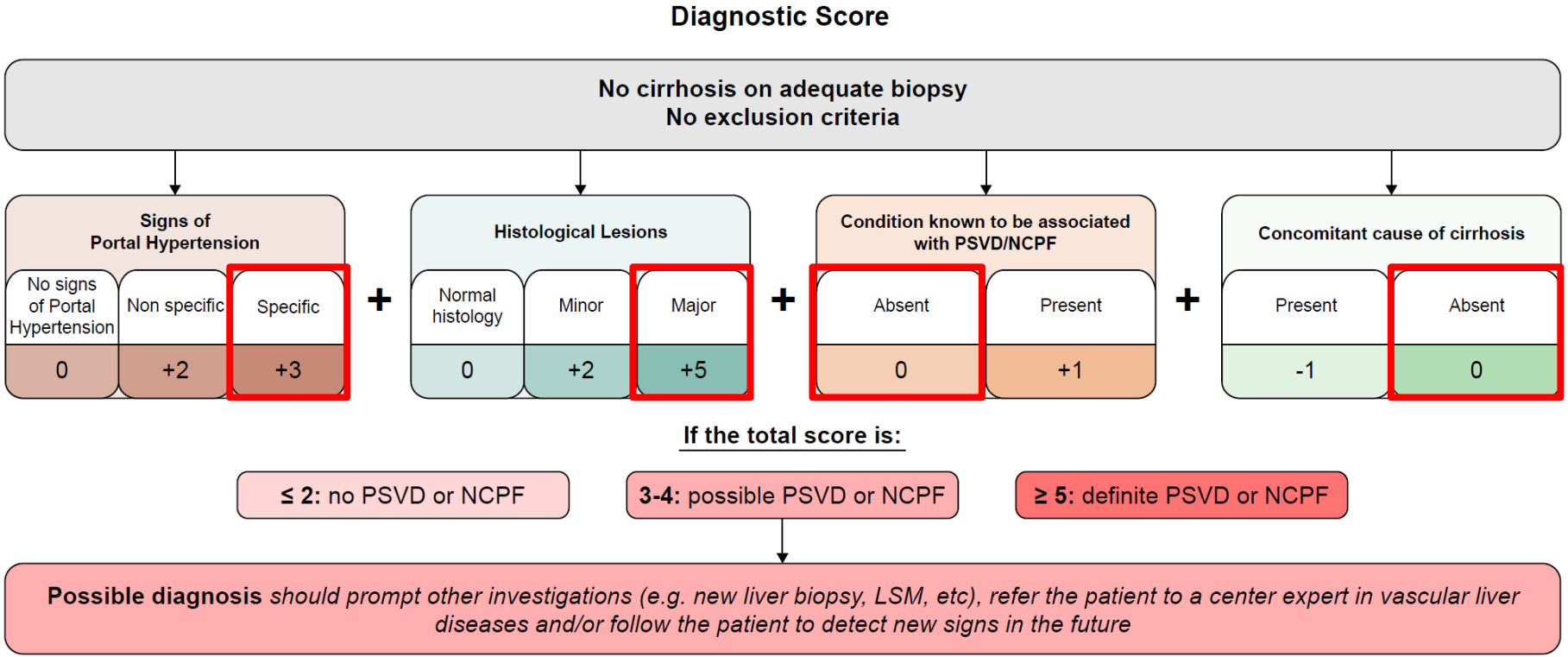








PSVD diagnostic criteria 2026



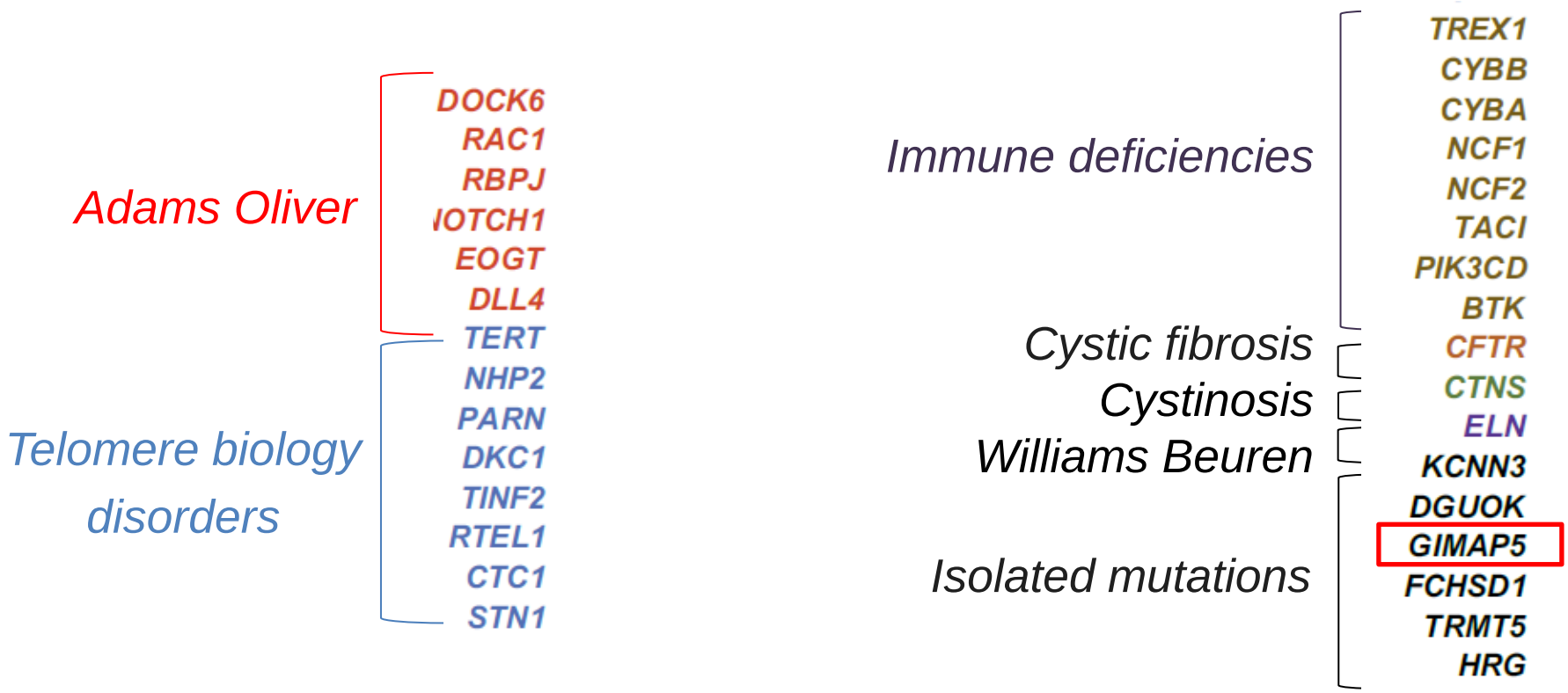
Hernandez-Gea, *et al,* J Hepatol 2026; In Press; Baveno VIII

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*

Case presentation

What explanation do you give to the patient?
What management do you propose?

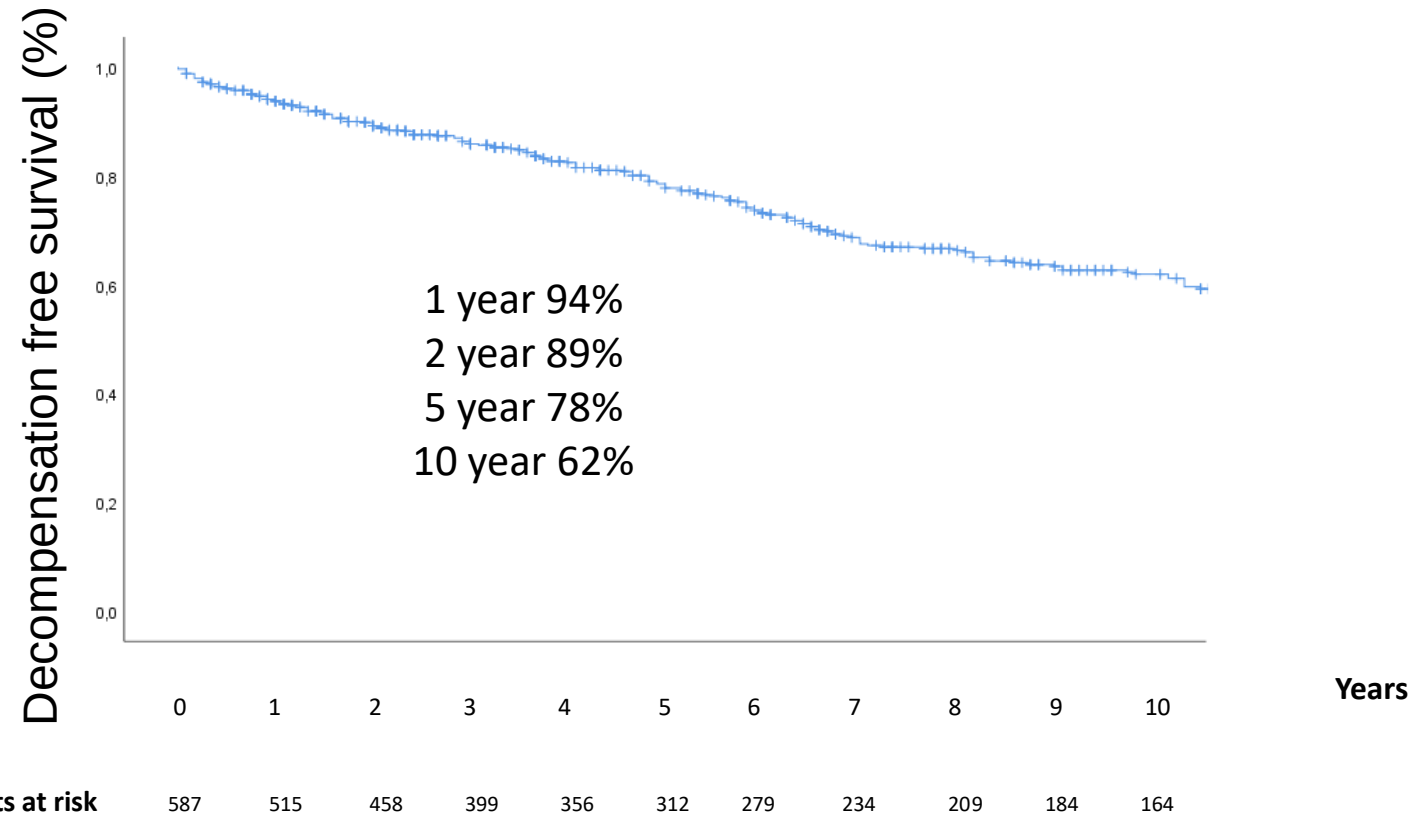
PSVD: outcome & complications

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

EASL guidelines on vascular liver diseases 2026

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

Networks for vascular liver diseases

European Reference Network | Hepatological Diseases (ERN RARE-LIVER)

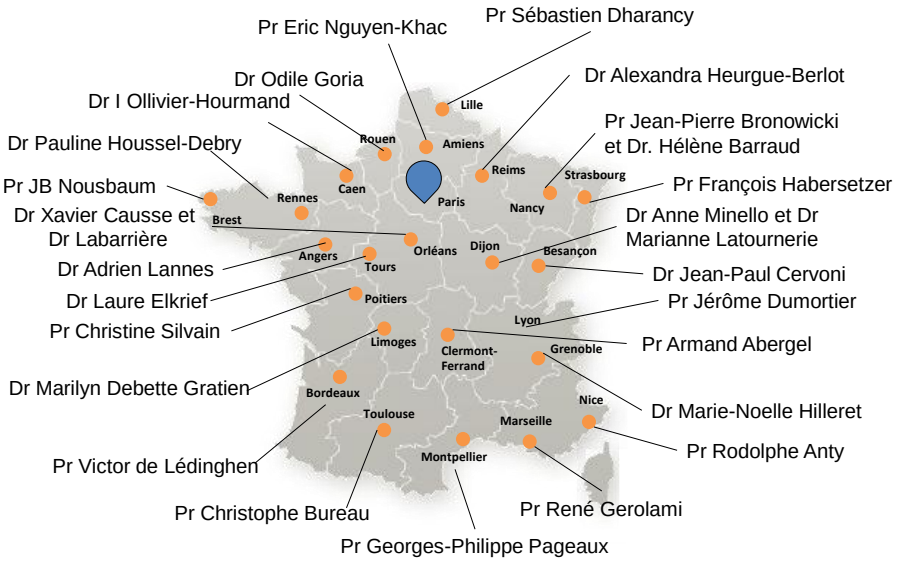
VALDIG

VASCULAR LIVER DISEASE GROUP

French network for vascular liver diseases

MVF

CENTRE DE RÉFÉRENCE MALADIES VASCULAIRES DU FOIE



Instituts thématiques

Inserm
Institut national
de la santé et de la recherche médicale

Université Paris Cité

CRI
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

HORIZON 2020

Les Programmes Hospitaliers de Recherche Clinique (PHRC)

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AMVF

FONDATION ARC POUR LA RECHERCHE SUR LE CANCER

FRM FONDATION RECHERCHE MÉDICALE

TIPSS for GI bleeding

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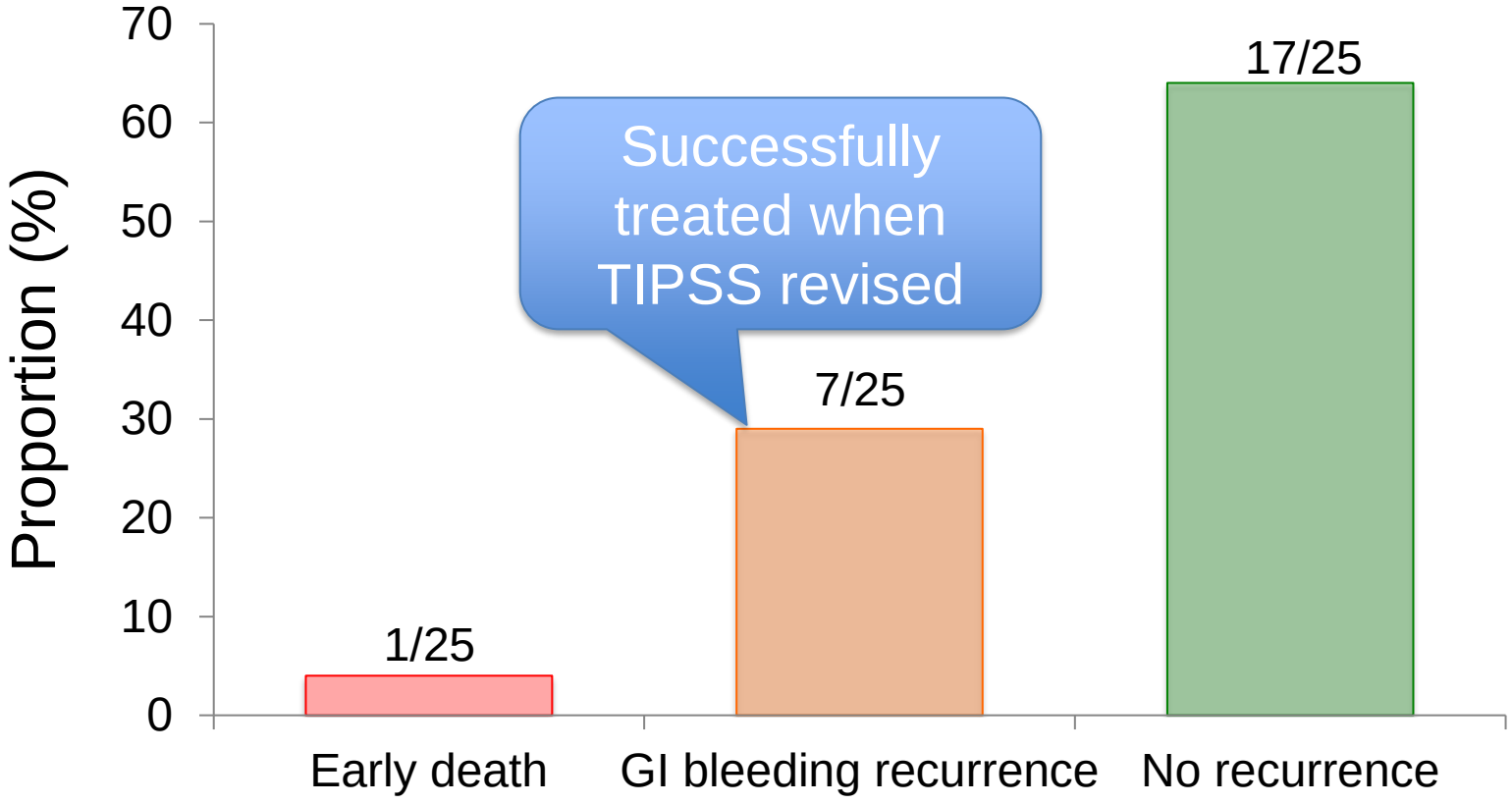
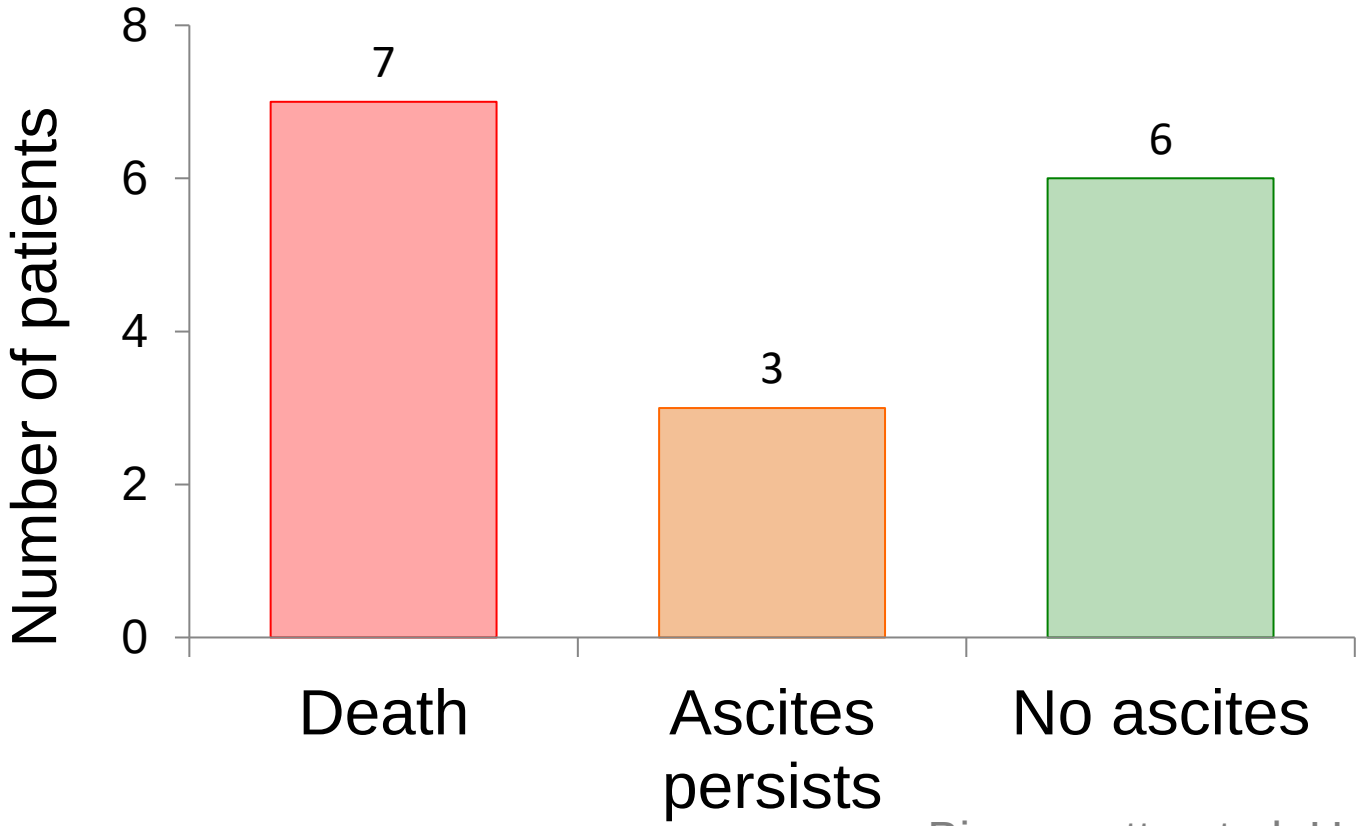


Bissonnette et al. Hepatology 2016; Lv APT 2019

TIPSS for refractory ascites



Bissonnette et al. Hepatology 2016

Survival after TIPSS

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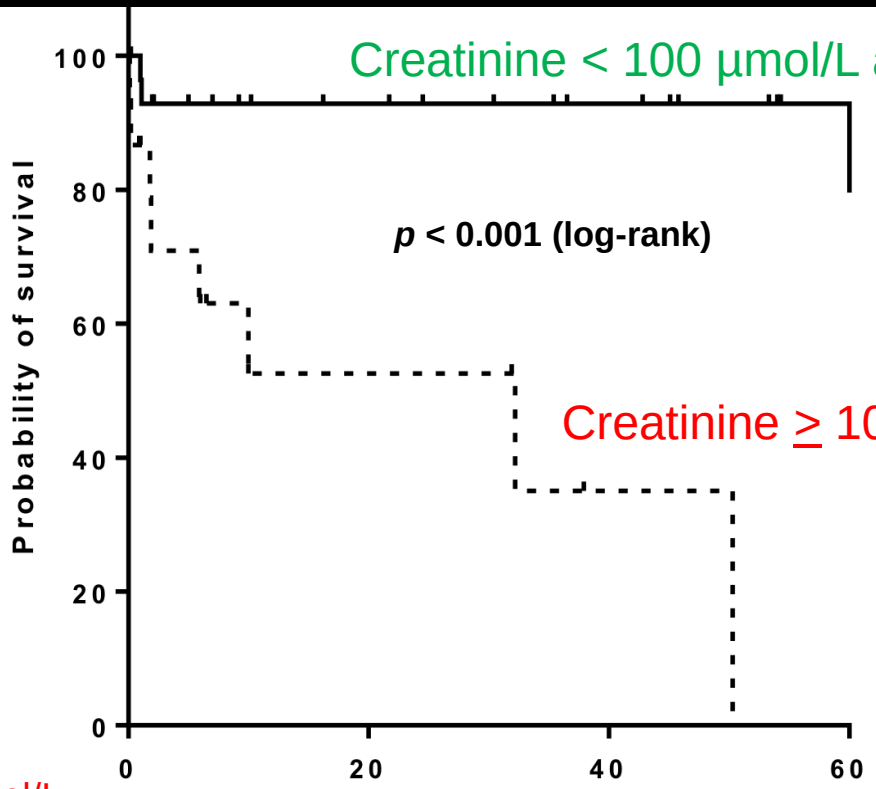

Creatinine ≥ 100 µmol/L AND comorbidities	7	2	1	0
Others	34	18	12	7

Bissonnette et al. Hepatology 2016

Liver transplantation PSVD

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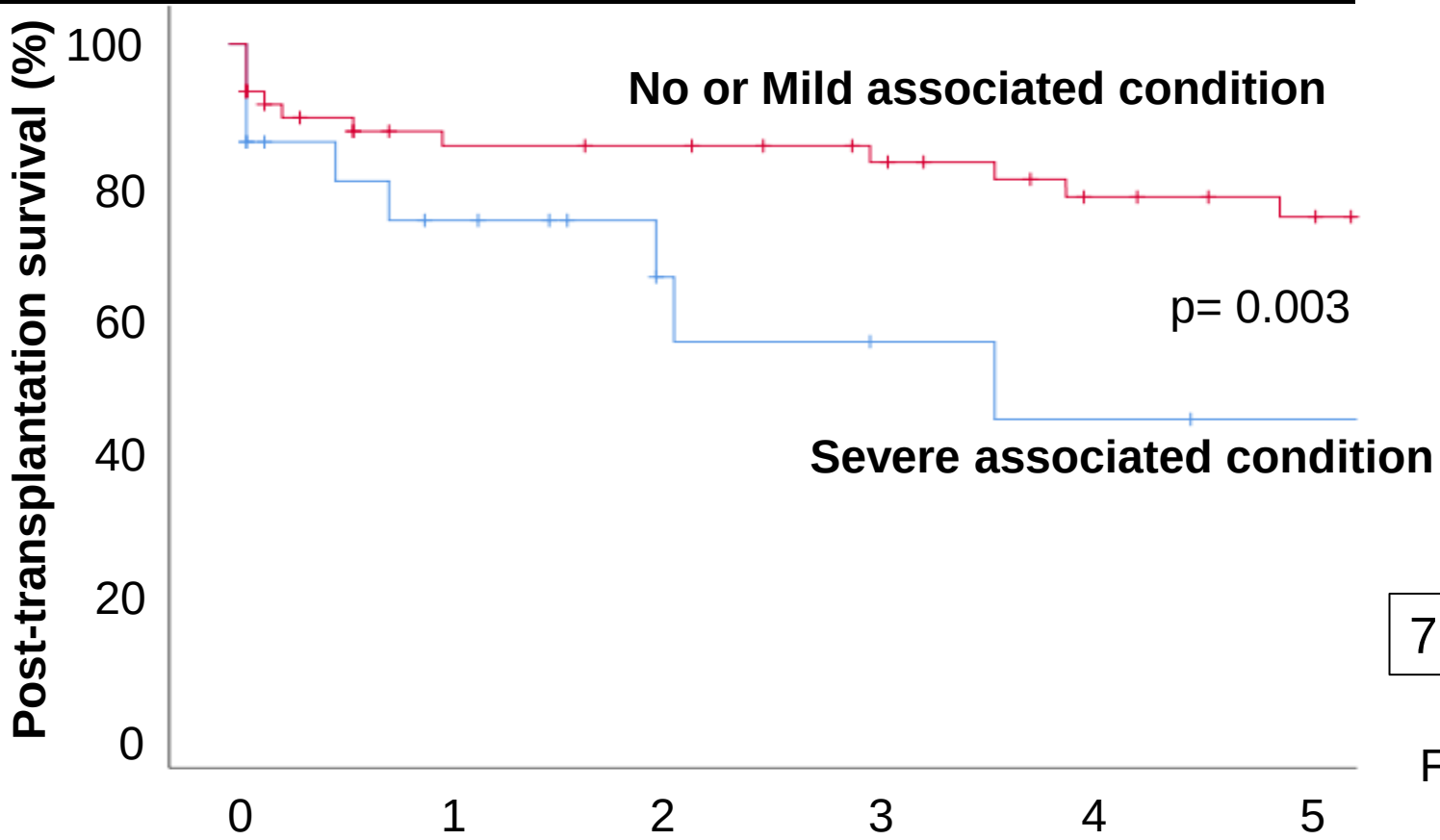

79 patients

Magaz*, Giudicelli*, *et al,* Transplantation 2023

Liver transplantation PSVD

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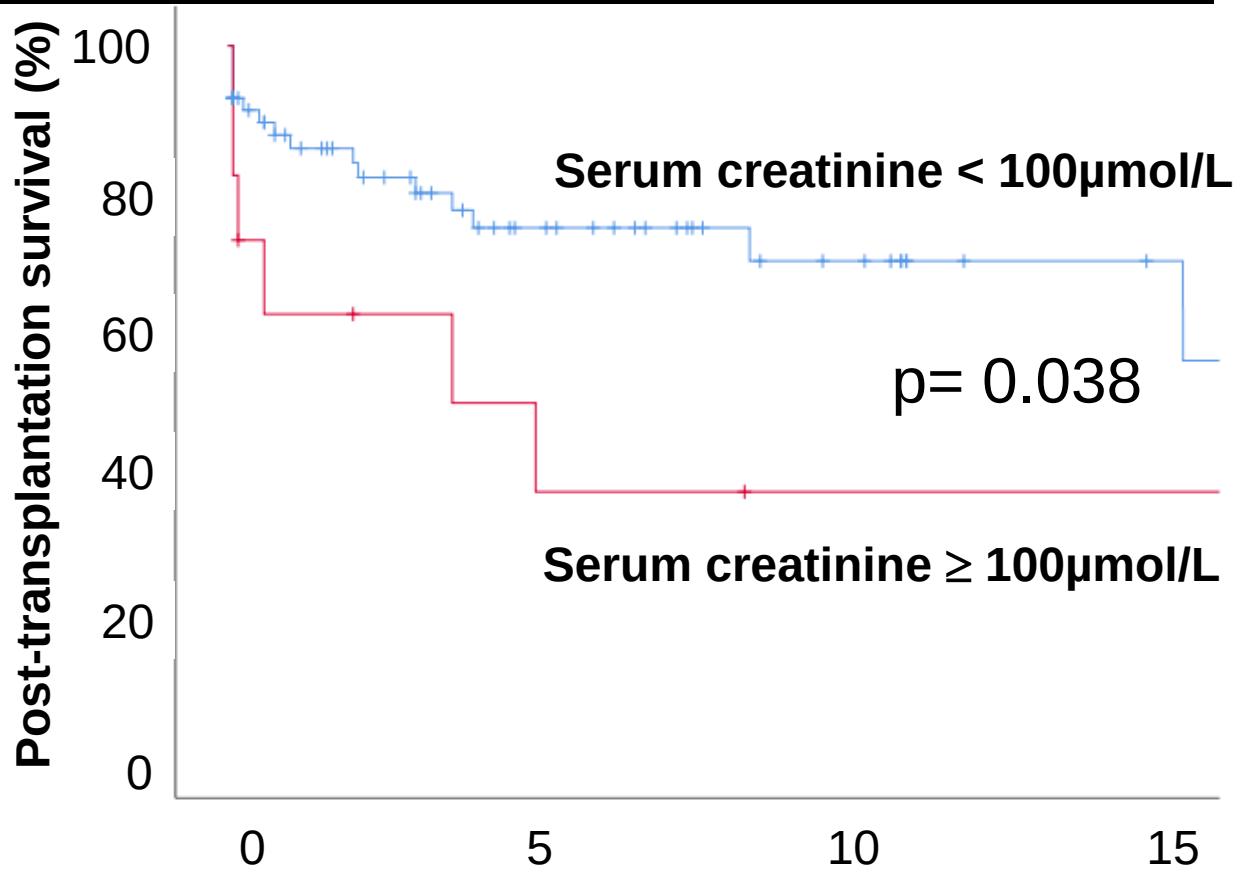

Magaz*, Giudicelli*, *et al,* Transplantation 2023