ASSISTANCE PUBLIQUE HÔPITAUX DE PARIS

Université de Paris

Vascular liver diseases

Diagnosis, management and clinico-pathological features

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MVF CENTRE DE RÉFÉRENCE MALADIES VASCULAIRES DU FOIE

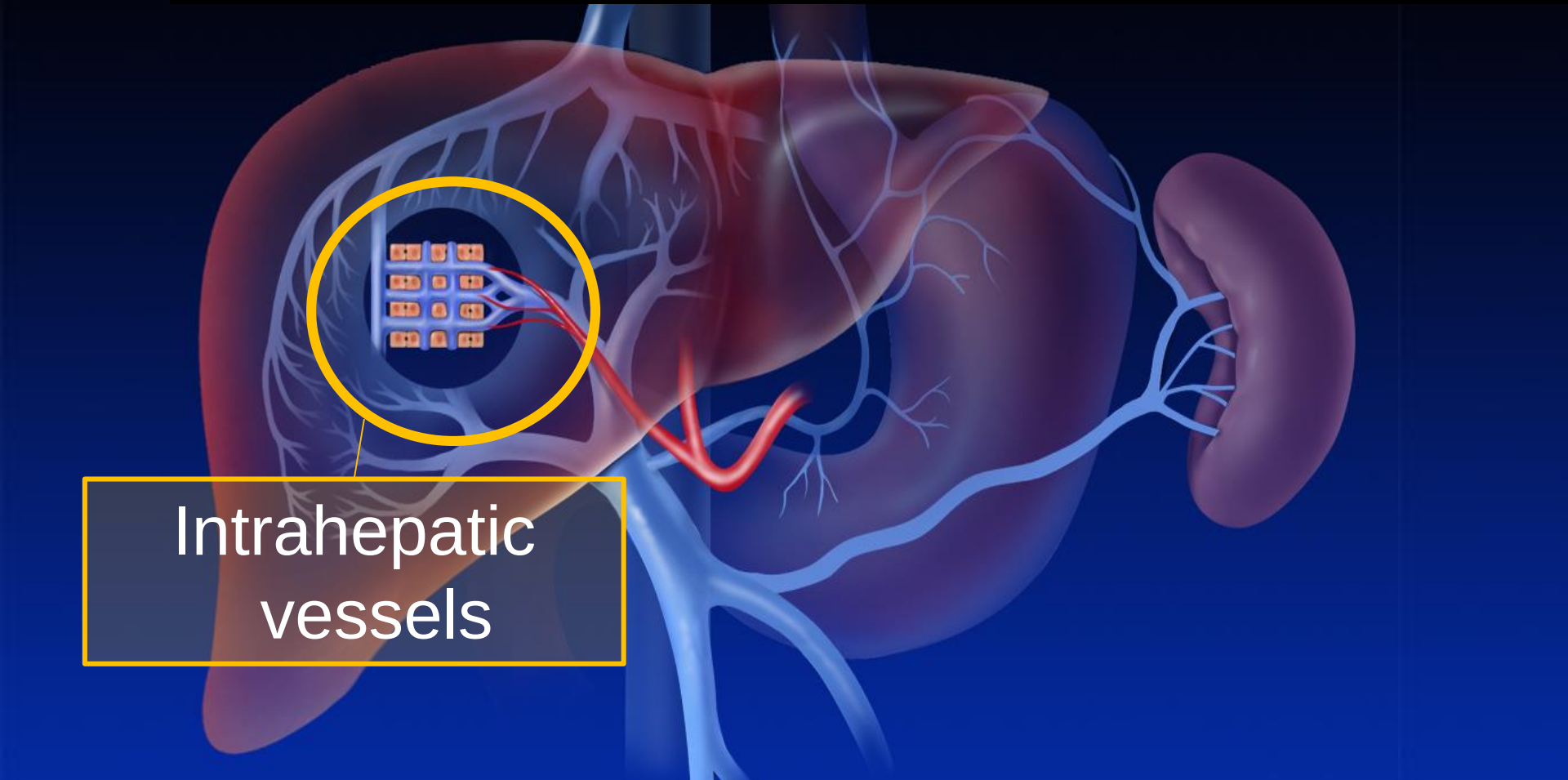
Instituts thématiques Inserm Institut national de la santé et de la recherche médicale

CRI CENTRE de RECHERCHE sur l'INFLAMMATION

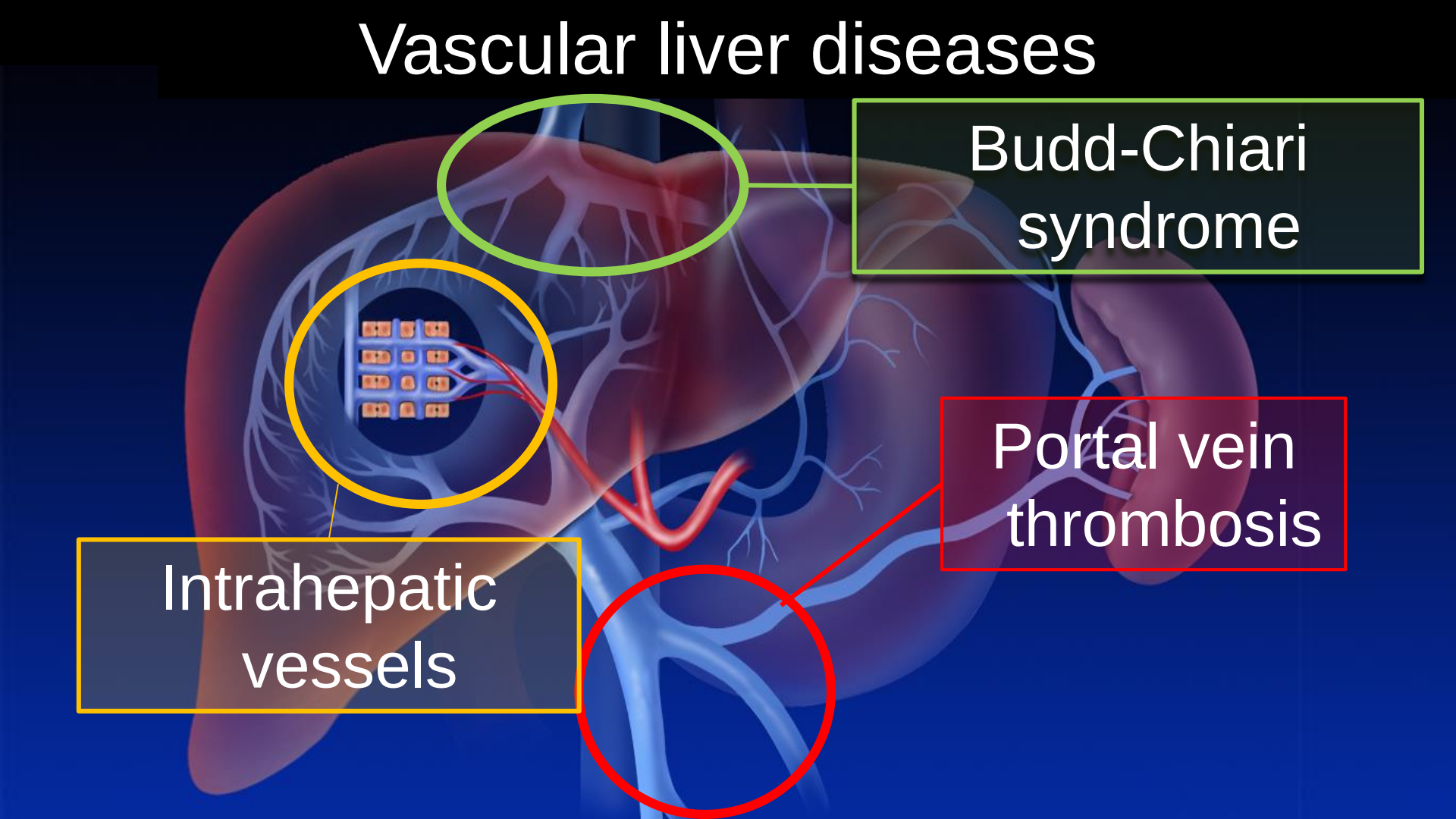
Conflict of interest

None related to the presentation

Vascular liver diseases



Vascular liver diseases

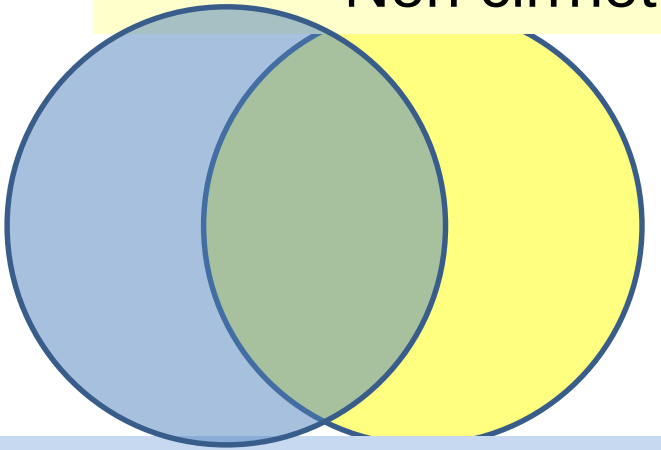


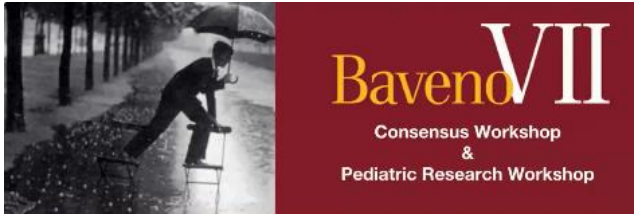
“Porto-sinusoidal vascular disorder” (PSVD)

HISTOLOGY

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

VALDIG
VASCULAR LIVER DISEASE GROUP





Idiopathic portal hypertension
Intrahepatic non cirrhotic portal hypertension

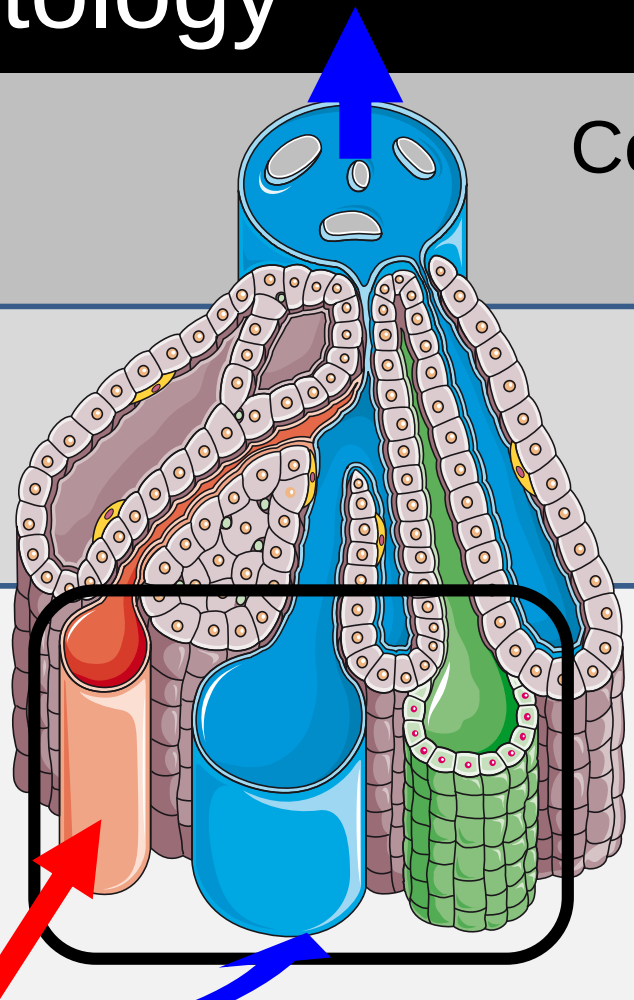
CLINIC

Liver histology

Central vein

Sinusoids

Porto-sinusoidal vascular disorder

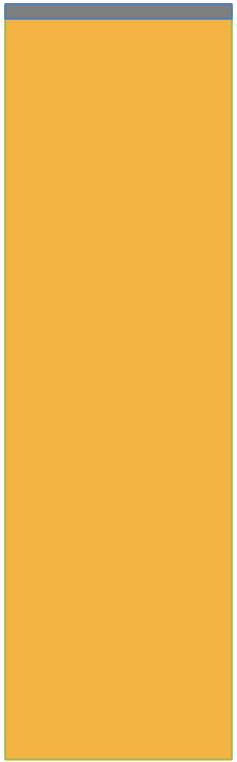


Portal tract

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?

Median age (yrs)	40 - 50
Abnormal liver blood tests	90 %
Signs of portal hypertension / complication	**70%/50%**
Portal vein thrombosis	30 - 50%
Fatigue	?
Prothrombin index < 50%	**15%**
Low liver stiffness measurement	70%
Normal or ↑ segment IV & smooth liver surface	65%
Low HVPG & hepatic veno-venous communications	60%/30%

De Gottardi et al, Lancet Gastro Hepatol 2019

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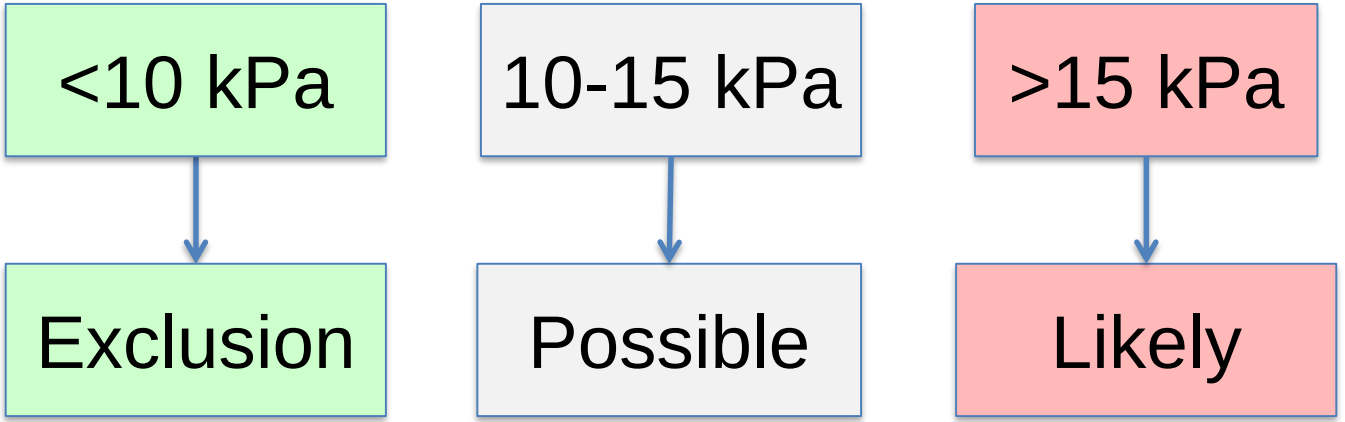
De Gottardi et al, Lancet Gastro Hepatol 2019

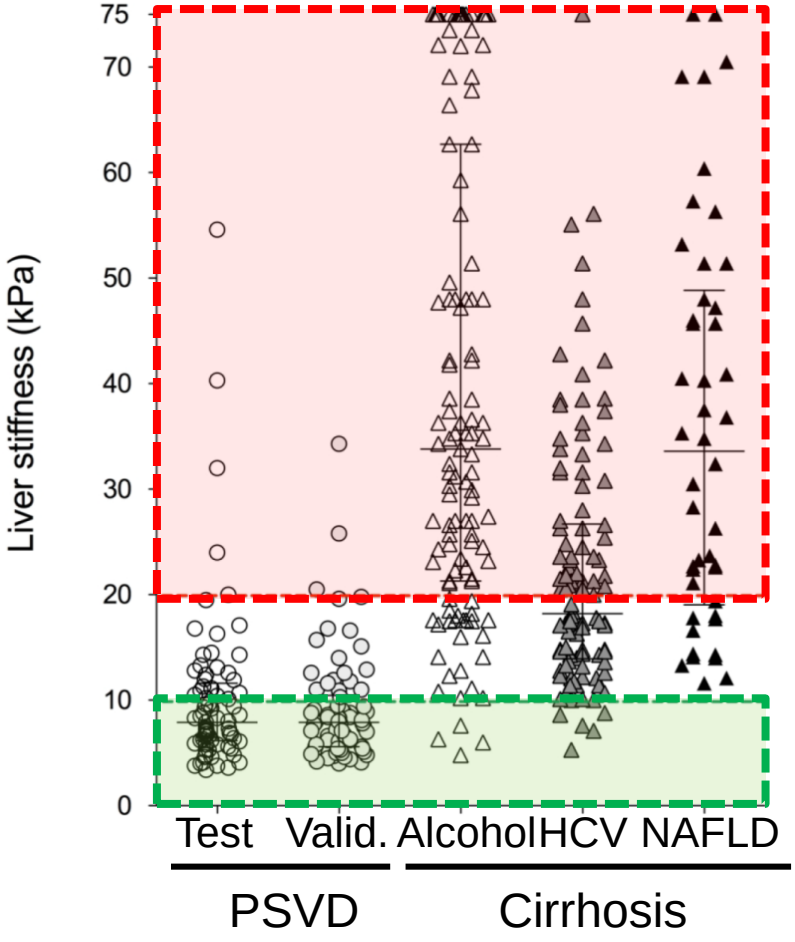
Liver stiffness measurement (LSM-TE) for cirrhosis

2 liver stiffness measurement (Fibroscan®)



De Franchis *et al.*, J Hepatol 2022

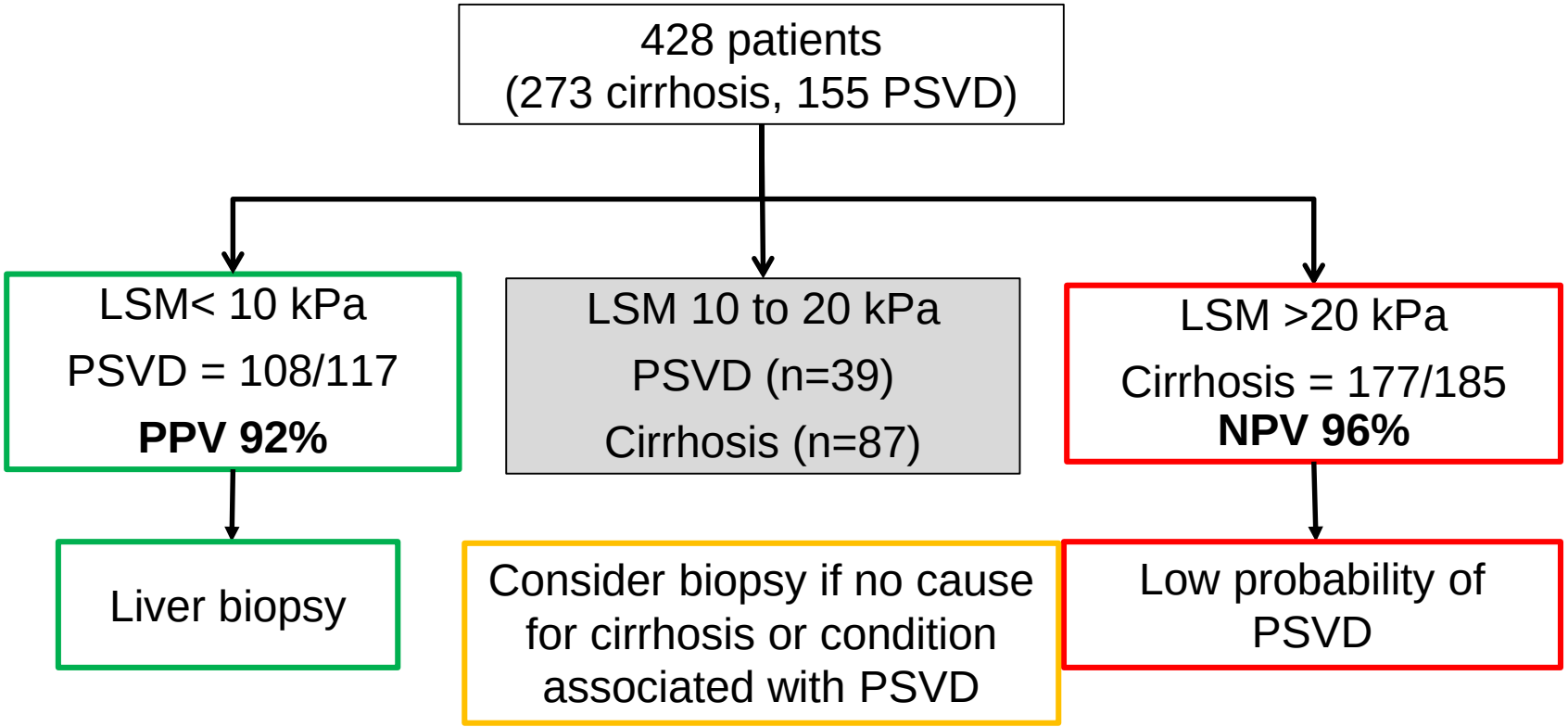
PSVD: liver stiffness measurement (TE)



Patients with signs of portal hypertension
155 PSVD, 273 cirrhosis

Elkrief, Lazareth, et al., Hepatology 2021

PSVD: liver stiffness measurement (TE)



Elkrief, Lazareth *et al.*, Hepatology 2021; De Franchis, *et al.* J Hepatol 2022

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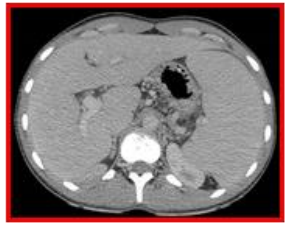
De Gottardi et al, Lancet Gastro Hepatol 2019

PSVD: CT-scan

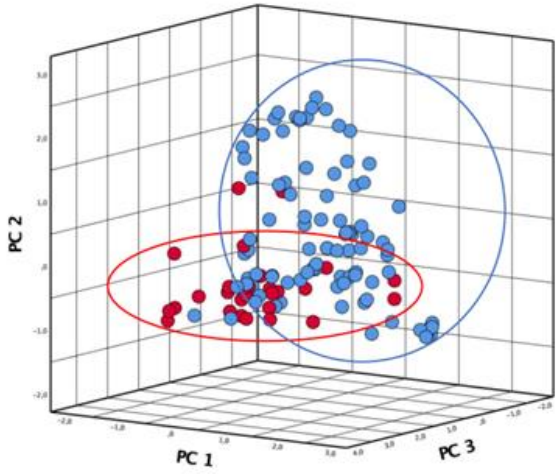


50 patients with PSVD

Matched on ascites

100 patients with cirrhosis

Different imaging pattern at CT scan

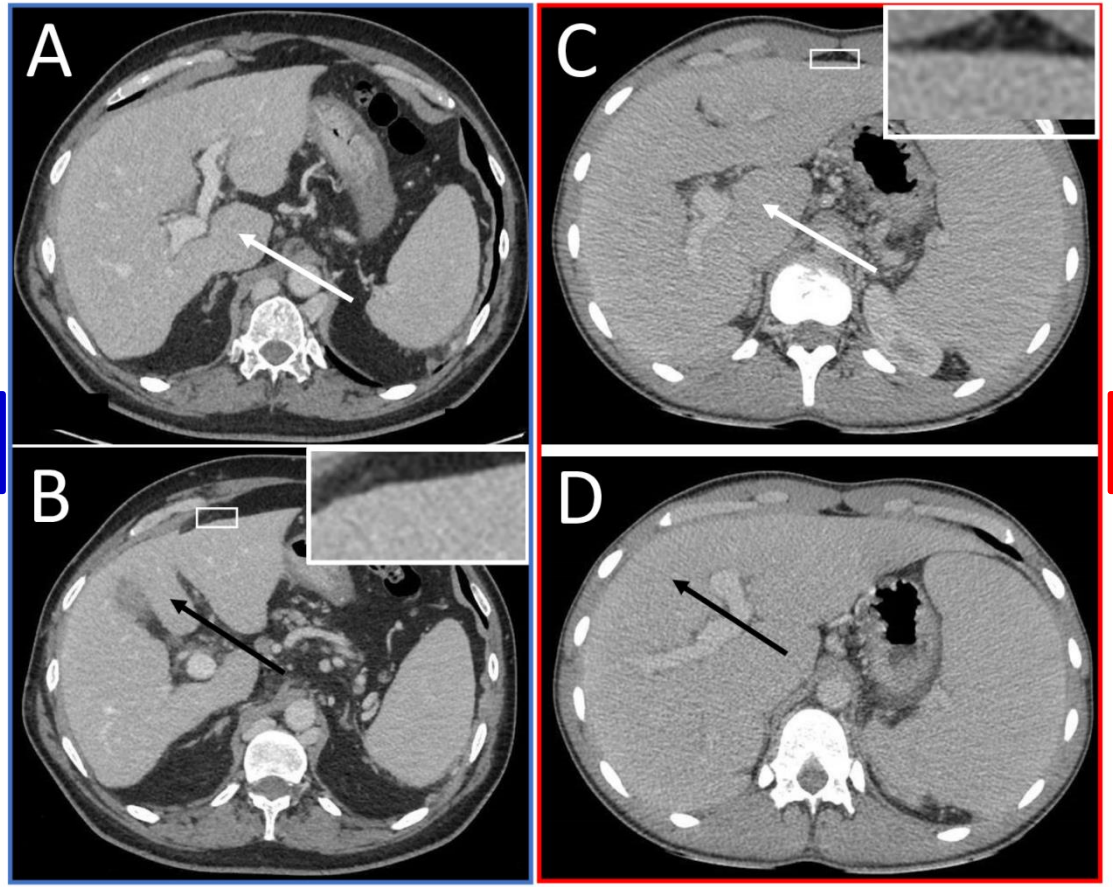


PSVD Cirrhosis

Valainathan et al. Hepatology 2022

PSVD: CT-scan

Cirrhosis



PSVD

Smooth liver surface

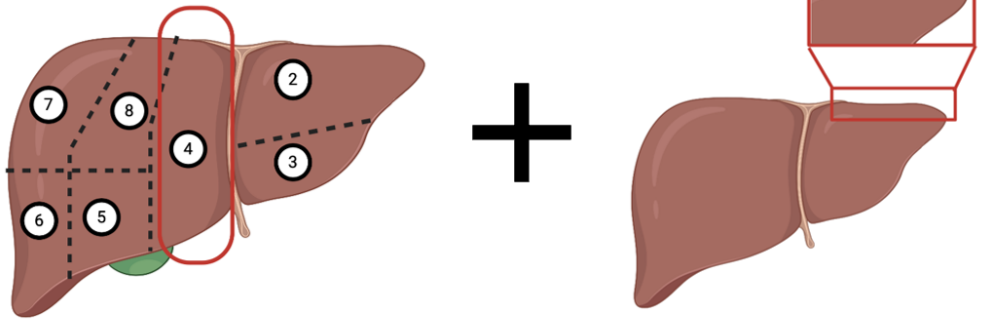
No segment IV atrophy

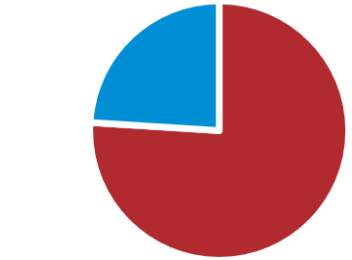
PSVD: CT-scan

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

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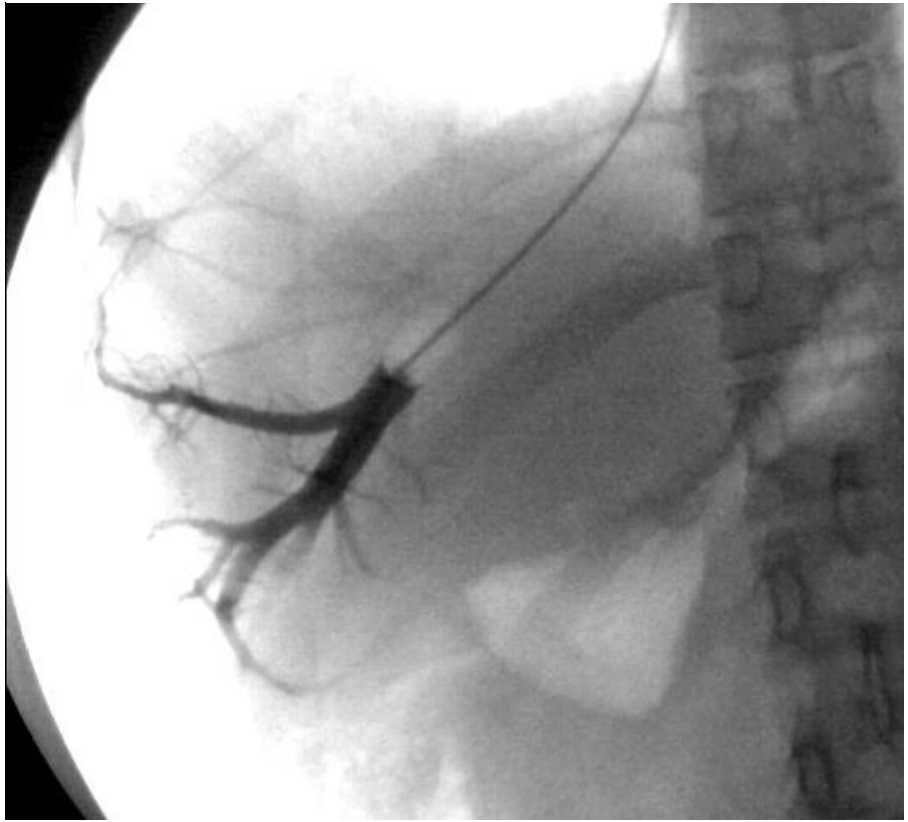
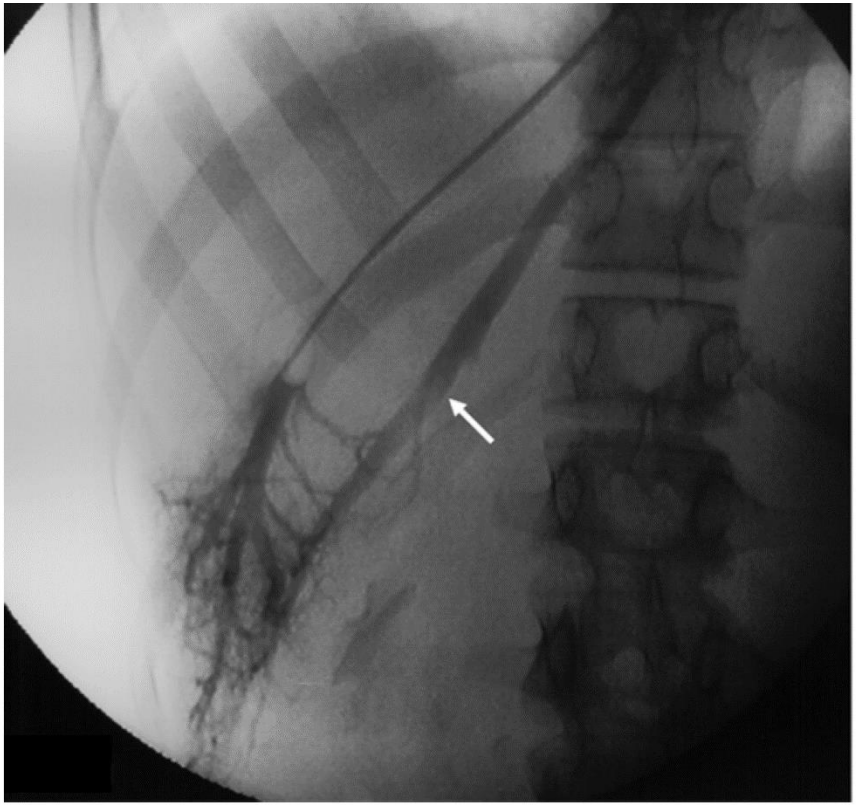
De Gottardi et al, Lancet Gastro Hepatol 2019

Hepatic veno-venous communications

Cirrhosis: 3%

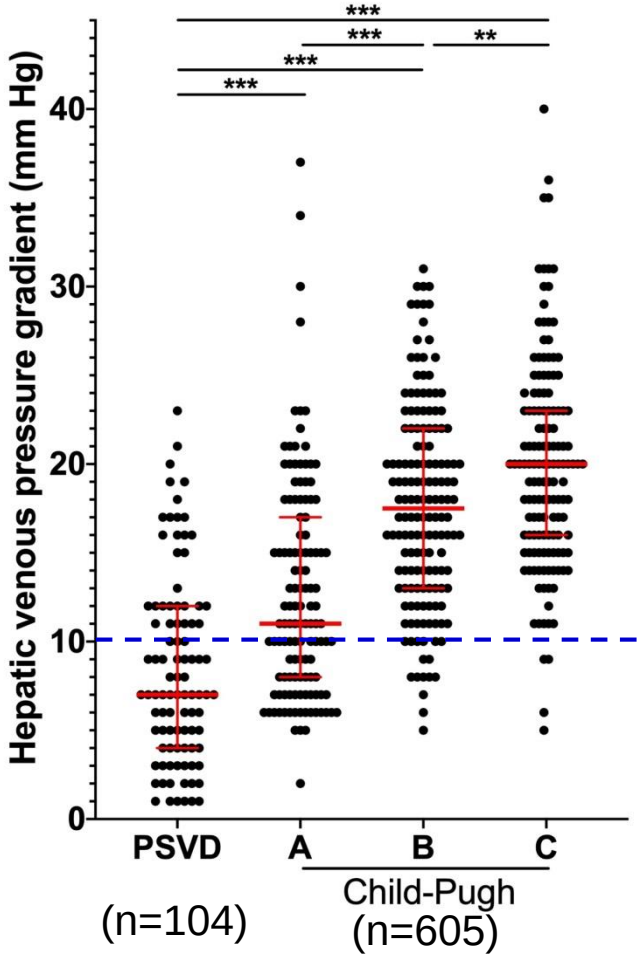


PSVD: 30 to 50%



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

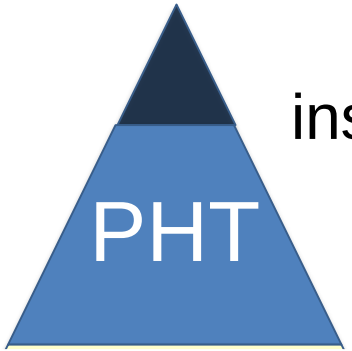
HVPG: PSVD *vs.* cirrhosis



Patients with signs of portal hypertension

Drilhon et al; ILC 2020

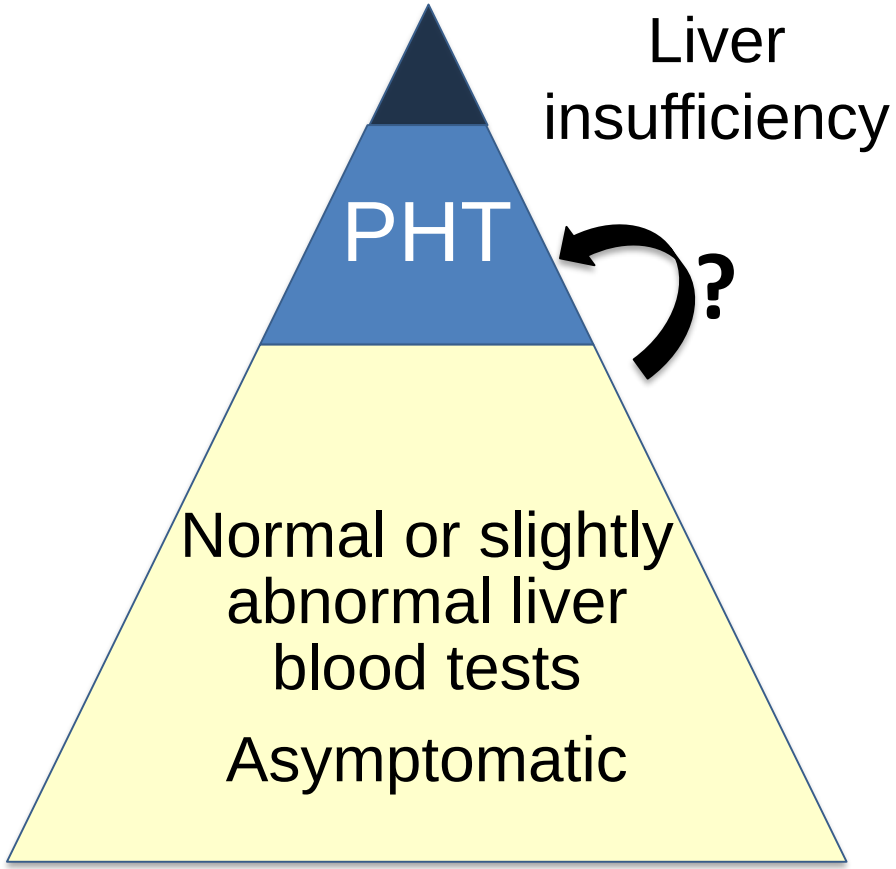
PSVD: manifestations



Liver insufficiency

Elkrief & Rautou, Liv Int 2016

PSVD: manifestations



Elkrief & Rautou, Liv Int 2016

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Guido, Liv Int 2016; Barge, Liv Int 2016

Porto-sinusoidal vascular disorder

- Presentation
- Definition
- Associated conditions
- Outcome

Porto-sinusoidal vascular disorder: definition

Biopsy
> 20 mm
without cirrhosis

1 specific sign

De Gottardi *et al,* Lancet Gastro Hepat 2019

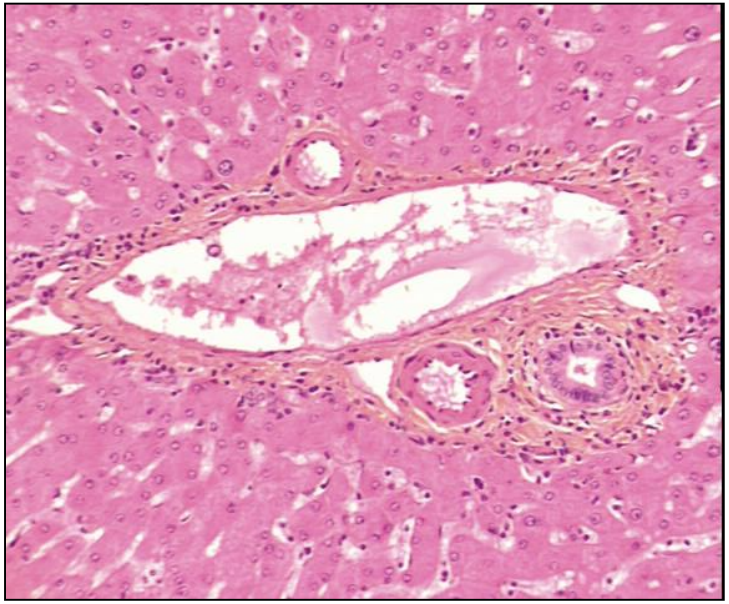
Signs specific for portal hypertension

- ✓ Esophageal, gastric or ectopic varices
- ✓ Portal hypertension related bleeding
- ✓ Porto-systemic collaterals at imaging

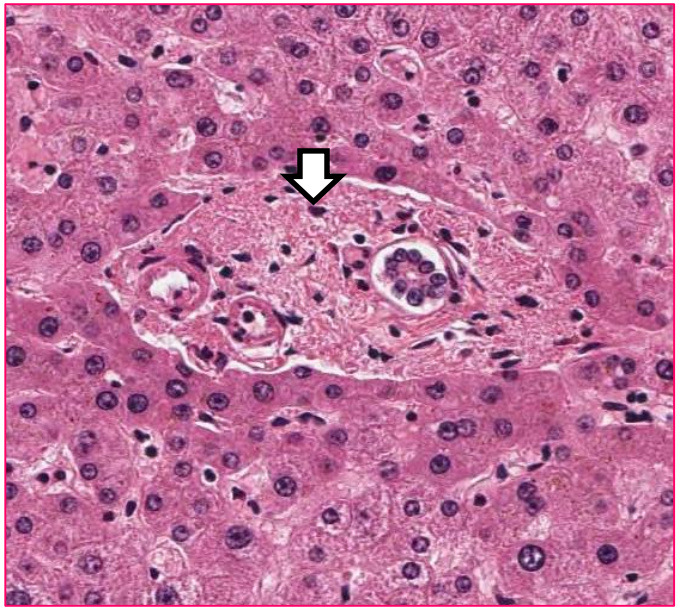
De Franchis, J Hepatol 2022
De Gottardi *et al,* Lancet Gastro Hepat 2019

Histological lesions specific for PSVD

- Obliterative portal venopathy



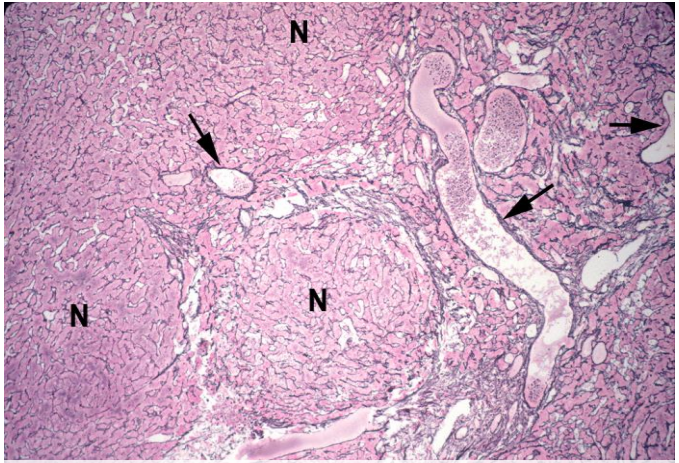
Normal portal tract



Obliterative portal venopathy

Histological lesions specific for PSVD

- Obliterative portal venopathy
- Nodular regenerative hyperplasia



Reticulin staining

Histological lesions specific for PSVD

- Obliterative portal venopathy
- Nodular regenerative hyperplasia
- Incomplete septal fibrosis/cirrhosis

Porto-sinusoidal vascular disorder: definition

Biopsy
> 20 mm
without cirrhosis

+

1 specific sign

Biopsy
> 20 mm
w/o cirrhosis

+

1 non specific sign of PHT and
1 non specific histological lesion

Non specific signs of portal hypertension

- Ascites
- Platelet count < 150’000/mm^3
- Spleen height > 13 cm

Histological lesions non specific for PSVD

- Portal tract abnormalities
- Architectural changes
- Sinusoidal dilatation
- Peri-sinusoidal fibrosis

Porto-sinusoidal vascular disorder

- Presentation
- Definition
- Associated conditions
- Outcome

PSVD: associated conditions

Prothrombotic conditions	++
Systemic or blood diseases	++
Infections (HIV, schistosomiasis)	++
Toxic / Drugs	+
Congenital diseases	+
Liver hyperarterialization	+
Aging	2%
No cause	+++

De Franchis, J Hepatol 2022; De Gottardi *et al,* Lancet Gastro Hepat 2019

Porto-sinusoidal vascular disorder

- Presentation
- Definition
- Associated conditions
- Outcome

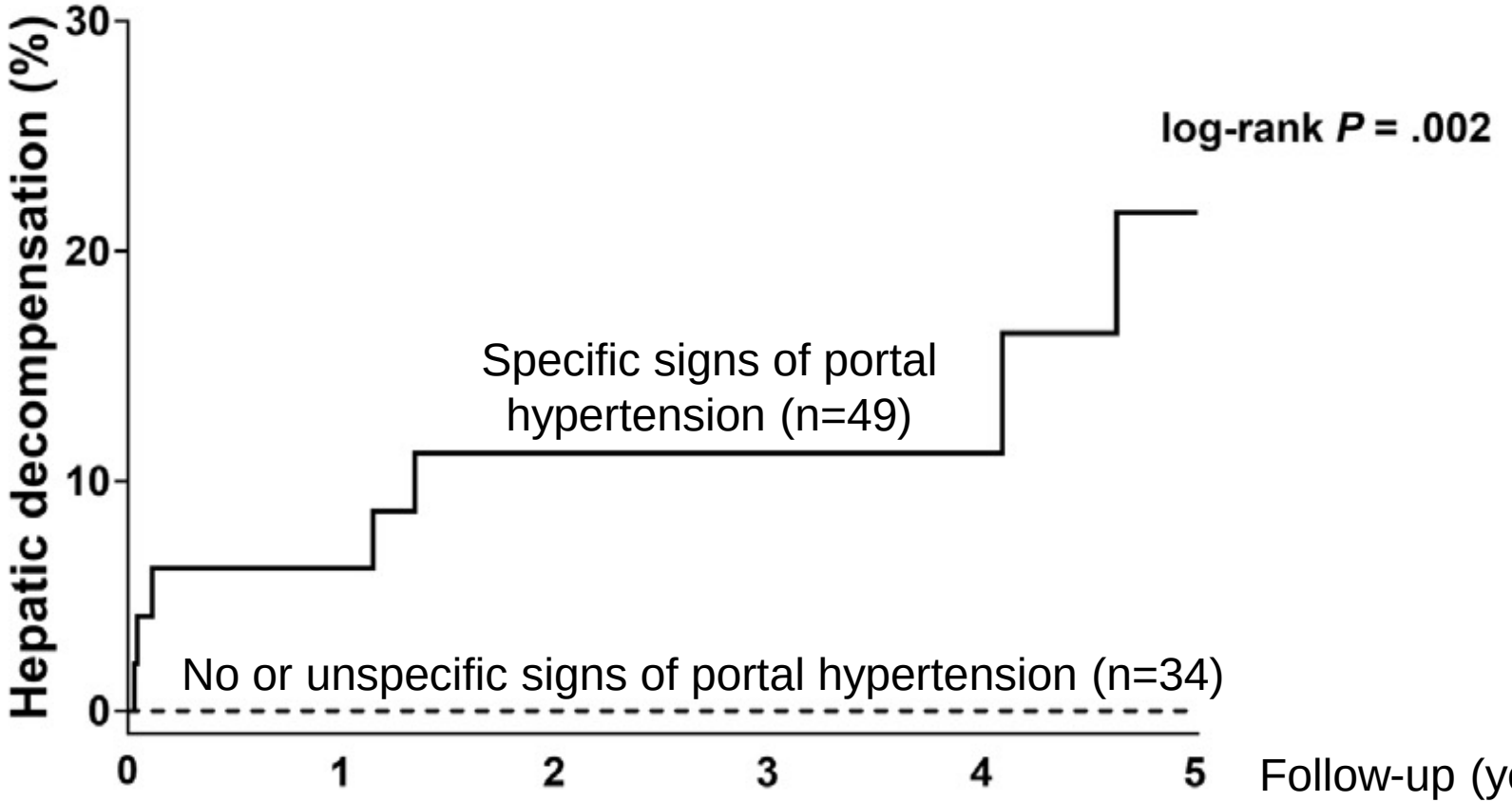
PSVD: outcome and complications

Gastro-intestinal bleeding	**23% at 5 yrs** **(despite prophylaxis)**
Portal vein thrombosis	**33% at 5 yrs**
Ascites	26% (at the time of GI bleeding; easily controlled)
Hepatic encephalopathy	7%
Hepatocellular carcinoma	0%

69 patients followed for 7 yrs

Siramolpiwat et al. Hepatology 2014
Similar data in: Gioia, et al. Dig Liver Dis. 2018

Outcome of PSVD: portal hypertension



Wöran, Clinical Gastroenterol Hepatol, 2020

PSVD: treatment

- Prophylaxis for variceal bleeding: like in cirrhosis
- Anticoagulation?
- Diuretics if ascites
- Rarely: TIPSS or liver transplantation

De Franchis, J Hepatol 2022

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)

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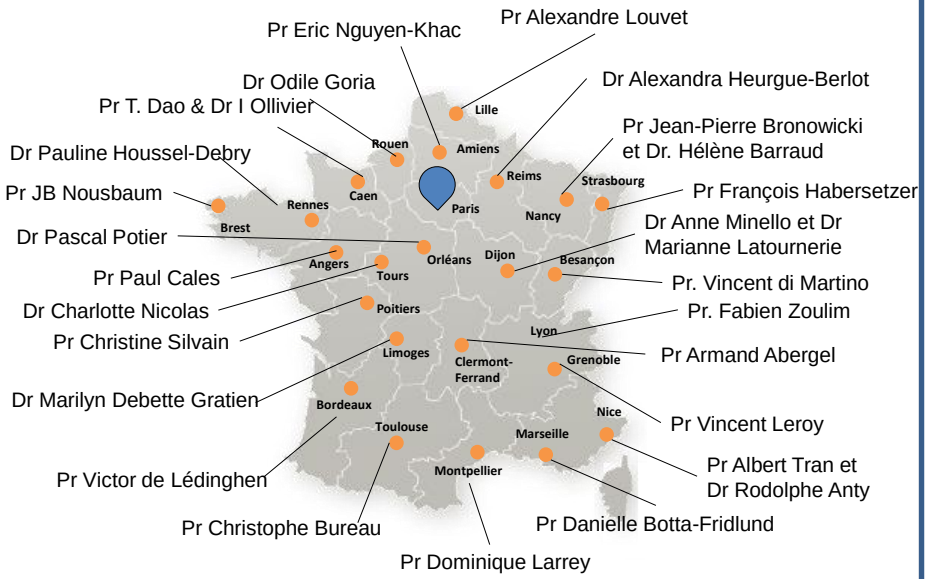
VASCULAR LIVER DISEASE GROUP

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 de Paris

CRI
CENTRE de RECHERCHE sur l'INFLAMMATION

Hepatology unit, Beaujon Hospital
Reference center for vascular liver diseases

Inserm U1149 – ATIP AVENIR, www.rautoulab.com/
Team "*vessels in liver diseases*"

AGENCE NATIONALE DE LA RECHERCHE ANR

EF Clif
EUROPEAN FOUNDATION FOR THE STUDY OF CHRONIC LIVER FAILURE

HORIZON 2020

AFEF
ASSOCIATION FRANÇAISE POUR L'ETUDE DU FOIE
(Association reconnue d'utilité publique)

Société Nationale Française de Gastro-Entérologie
SNFGE

Association des Malades des Vaisseaux du Foie
AMVF

Les Programmes Hospitaliers de Recherche Clinique (PHRC)

Get these slides there:

TIPSS for GI bleeding

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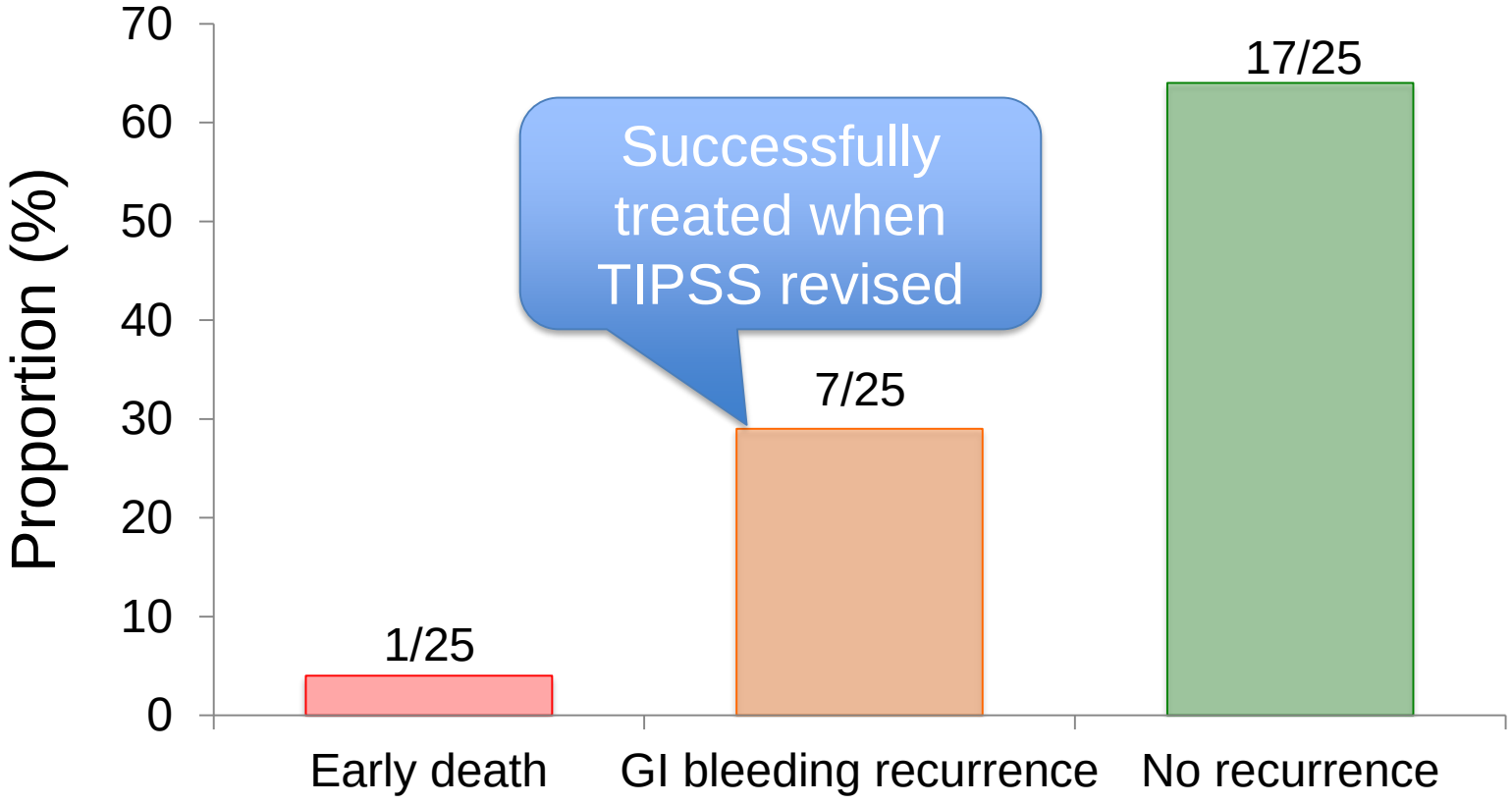
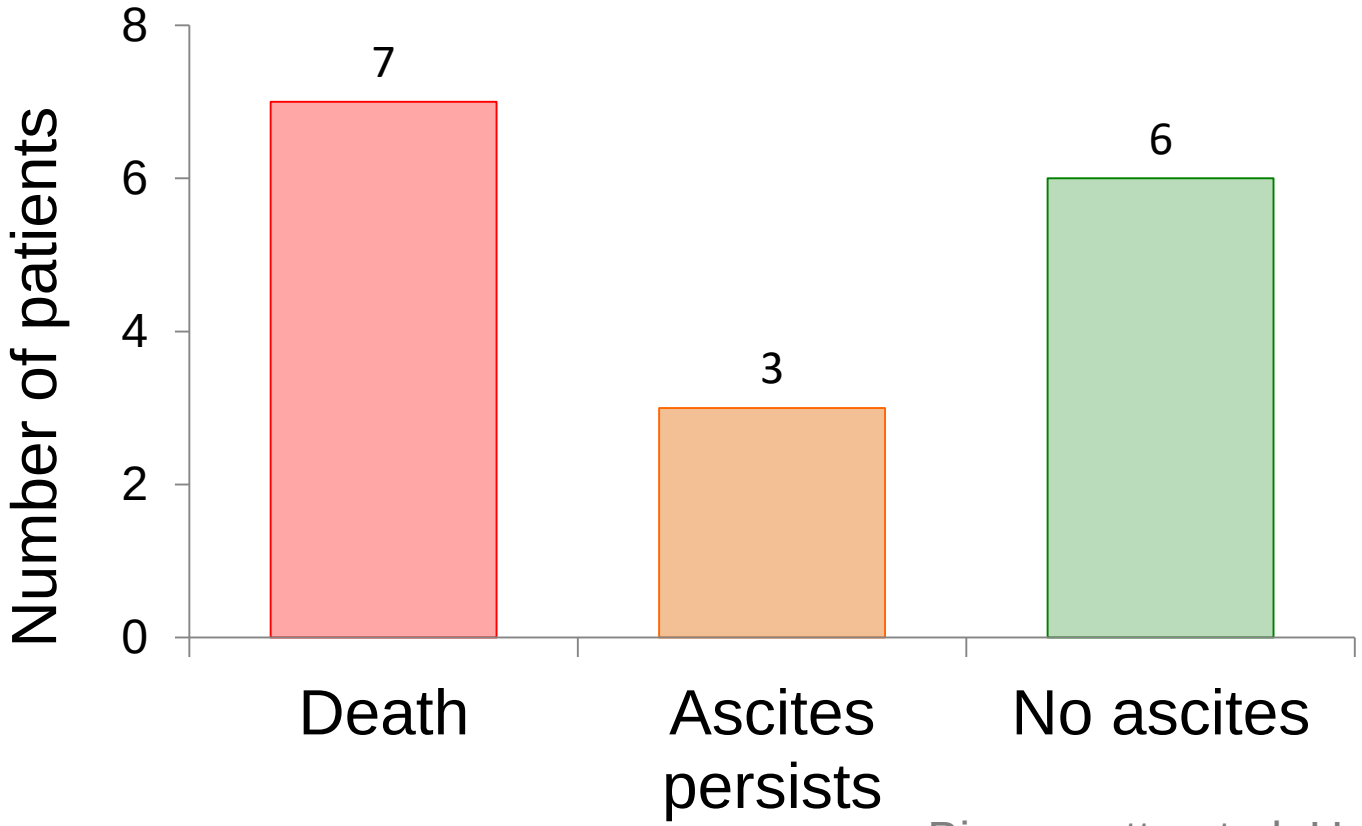


Bissonnette et al. Hepatology 2016; Lv APT 2019

TIPSS for refractory ascites



Bissonnette et al. Hepatology 2016

Survival after TIPSS

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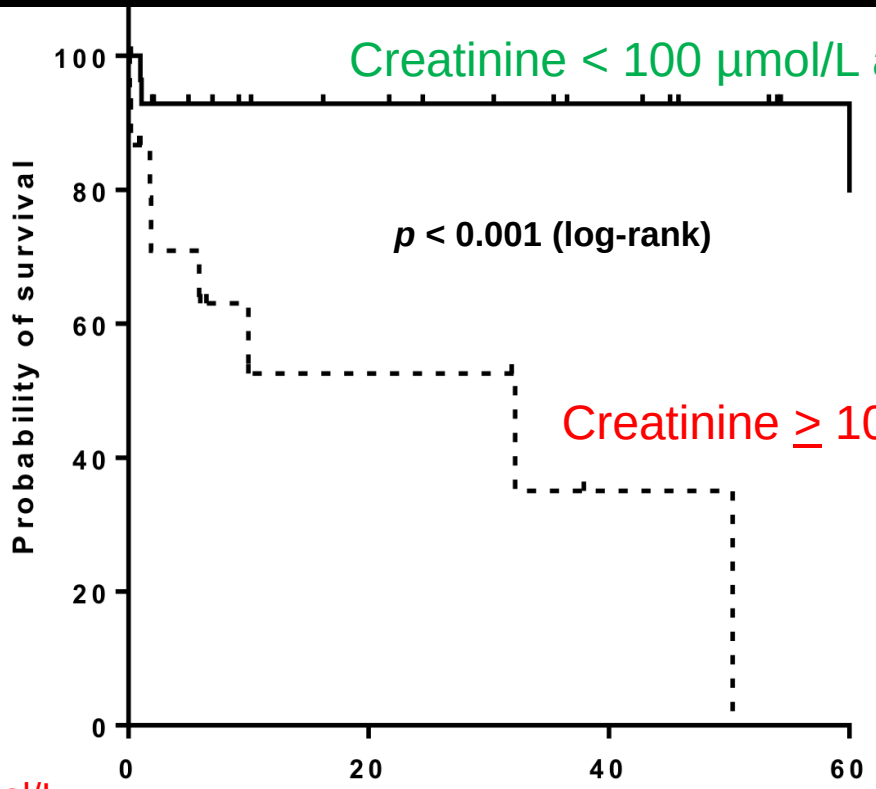

Creatinine ≥ 100 µmol/L AND comorbidities	7	2	1	0
Others	34	18	12	7

Bissonnette et al. Hepatology 2016

Liver transplantation PSVD

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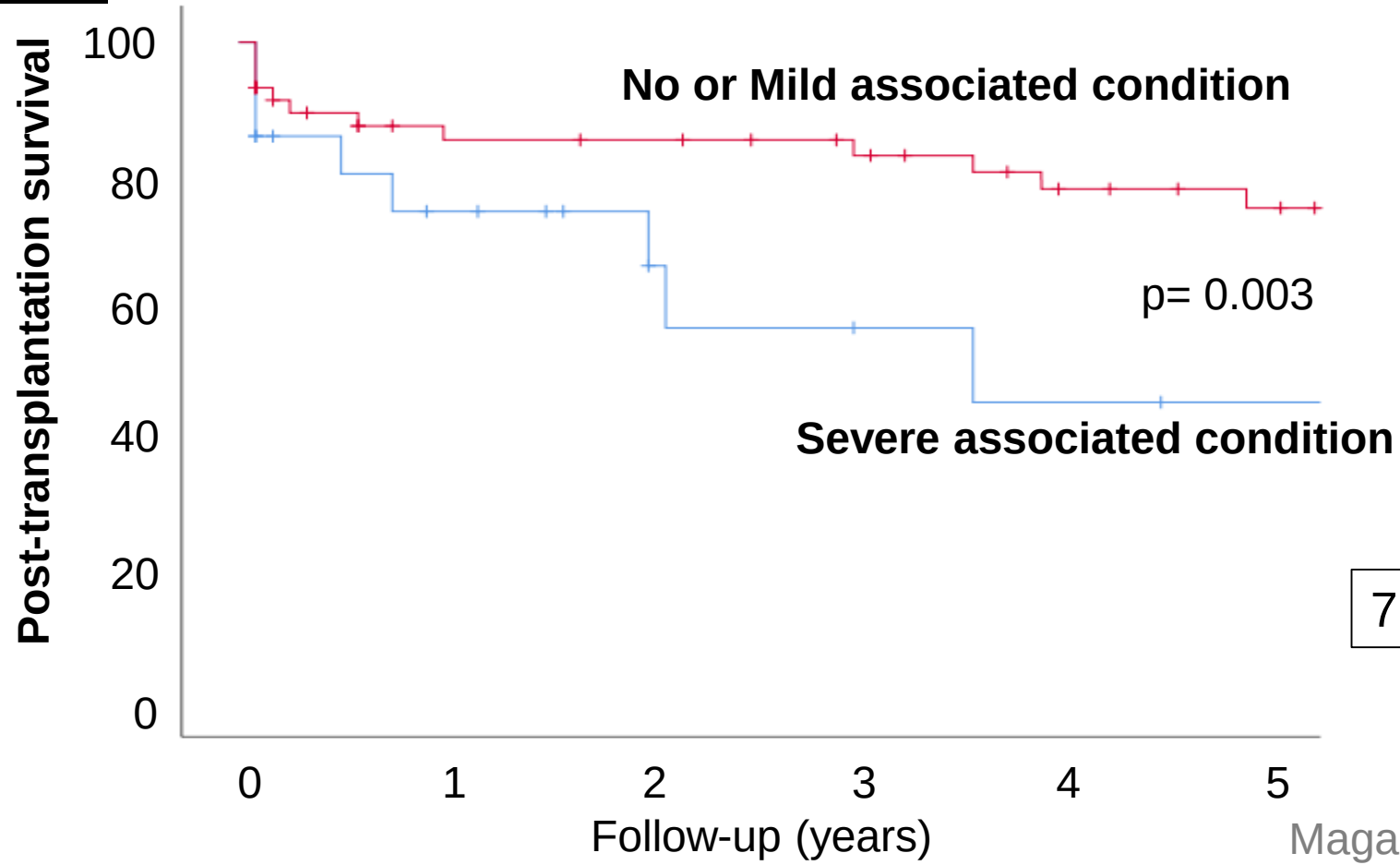

79 patients

Magaz, et al, submitted