



# Porto-sinusoidal vascular disorder: diagnosis and treatment

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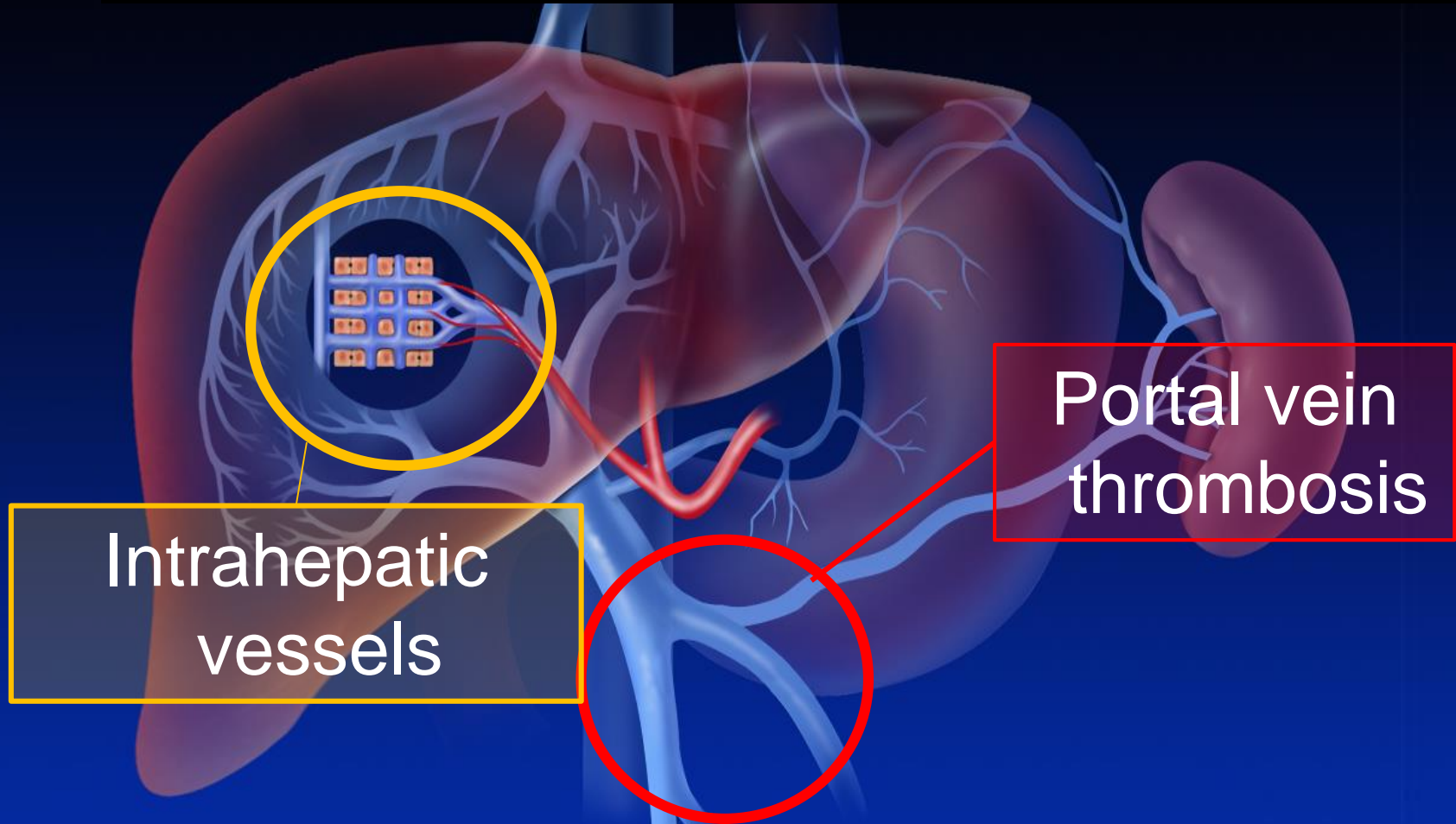
Service d'hépatologie, Hôpital Beaujon, Clichy, France

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# Porto-sinusoidal vascular disorder

- Presentation
- Definition
- Associated conditions
- Outcome

# Vascular liver diseases



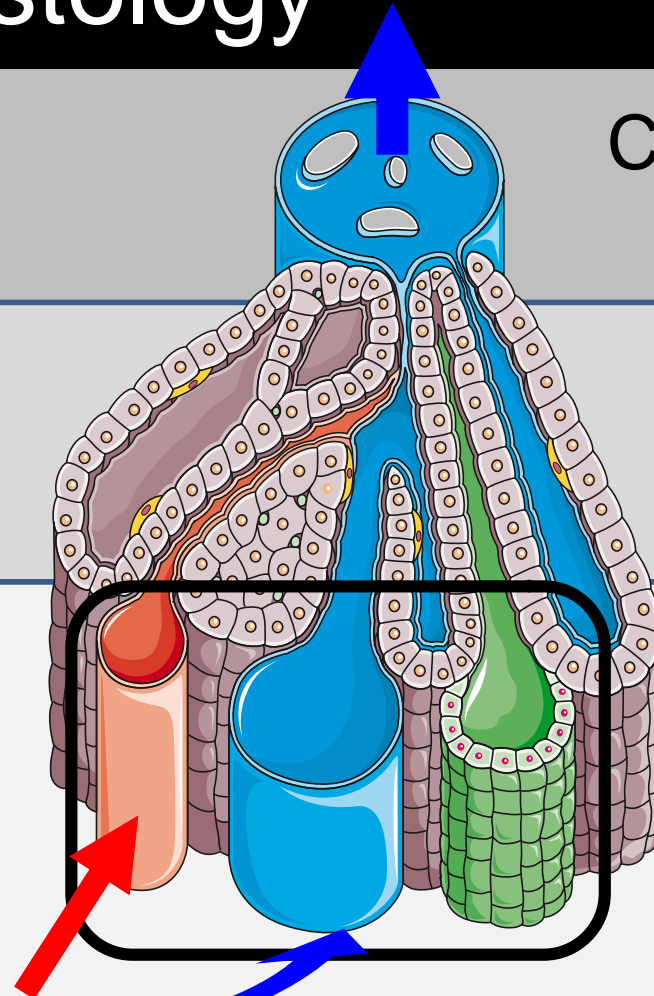
# Liver histology

Central vein

Sinusoids

Portal tract

Porto-sinusoidal  
vascular  
disorder



# PSVD: when to suspect?

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

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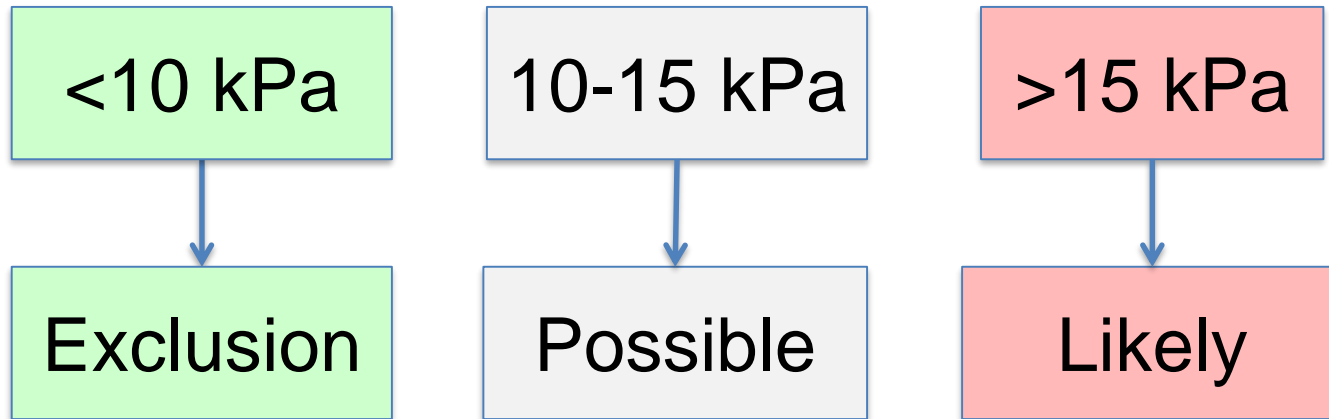
# Liver stiffness measurement (TE)

Fibroscan®

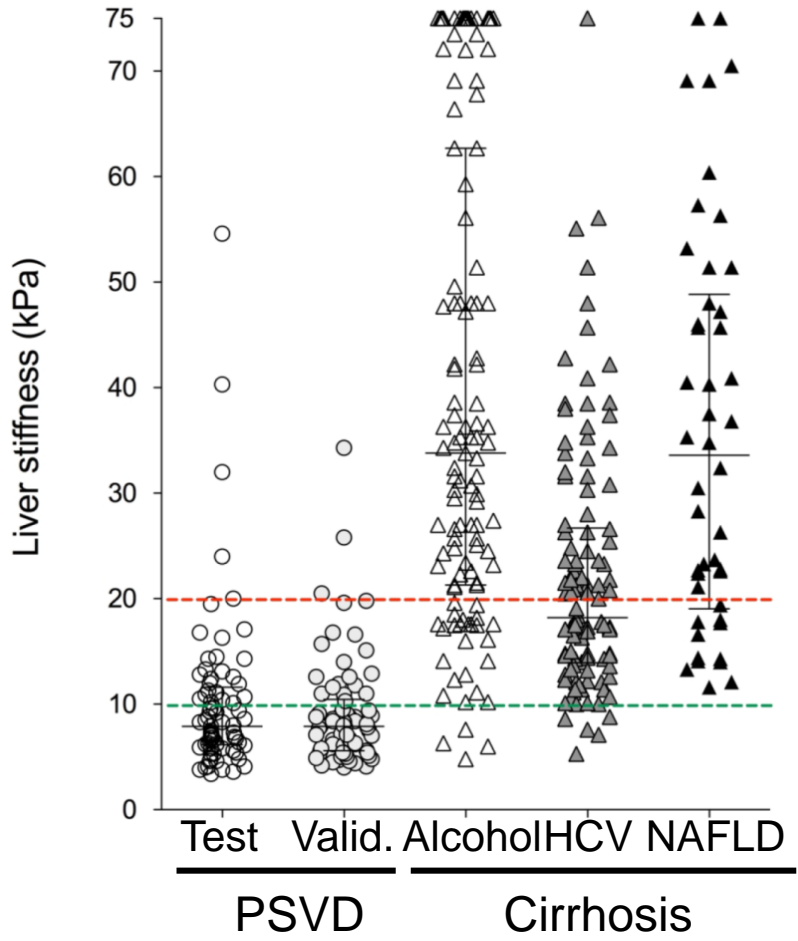


# Liver stiffness measurement (LSM-TE) for cirrhosis

2 liver stiffness measurement (Fibroscan®)

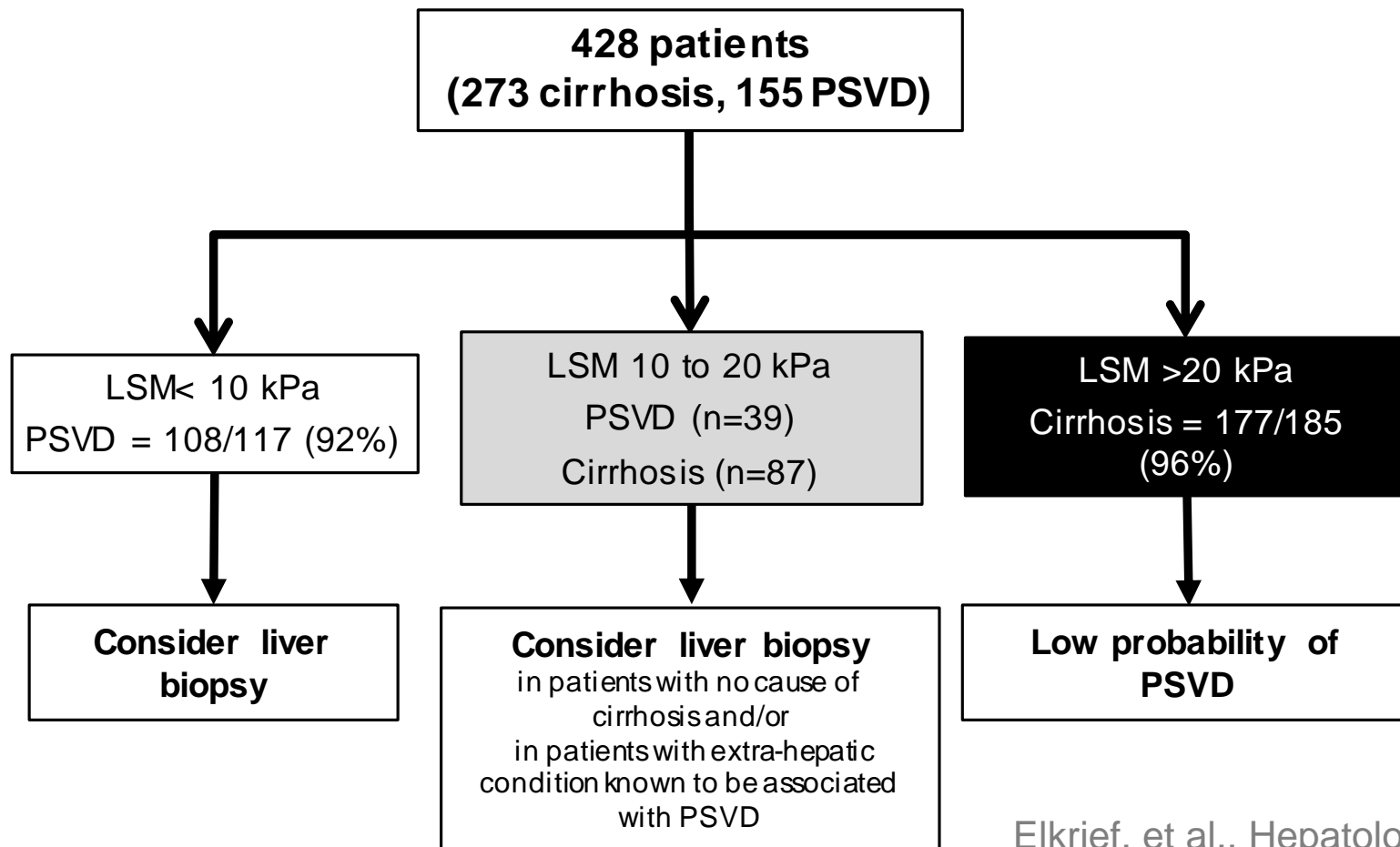


# PSVD: liver stiffness measurement (TE)



Patients with signs of portal hypertension

# PSVD: liver stiffness measurement (TE)



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# PSVD: CT-scan



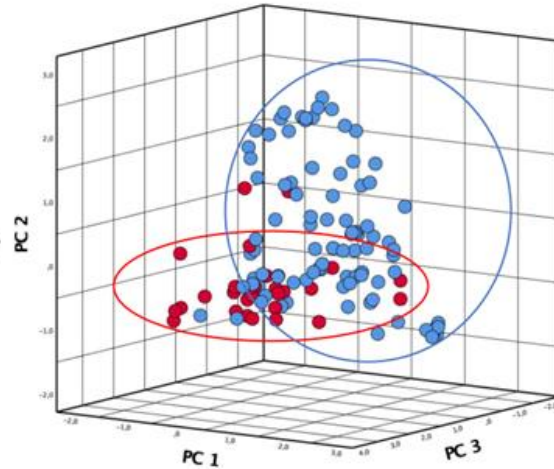
50 patients  
with PSVD

*Matched on  
ascites*

100 patients  
with cirrhosis

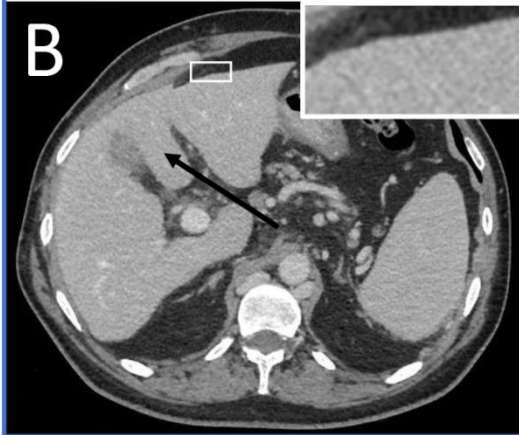
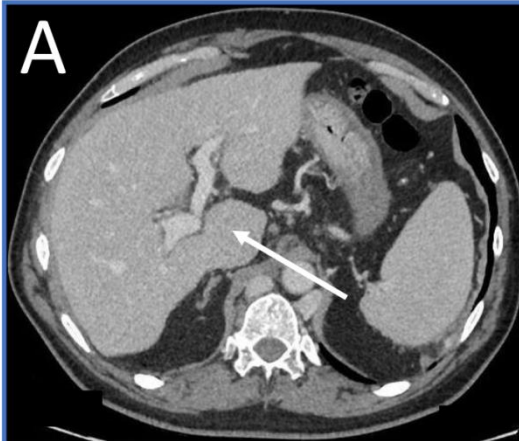


Different imaging  
pattern at CT scan



PSVD Cirrhosis

# PSVD: CT-scan



Smooth liver surface

PSVD

No segment IV atrophy

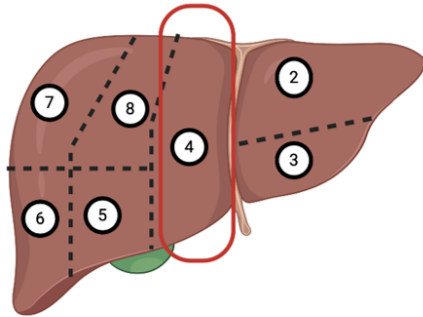
Cirrhosis

# PSVD: CT-scan

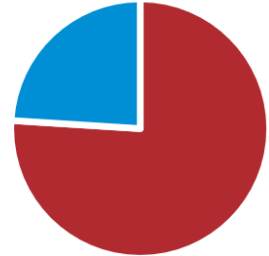
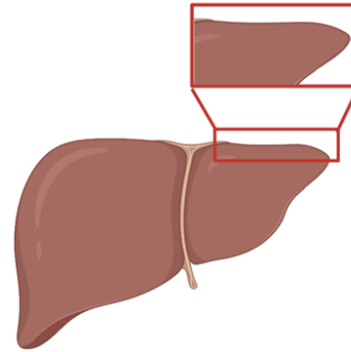
## *CT features for PSVD diagnosis*

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

# PSVD: when to suspect?

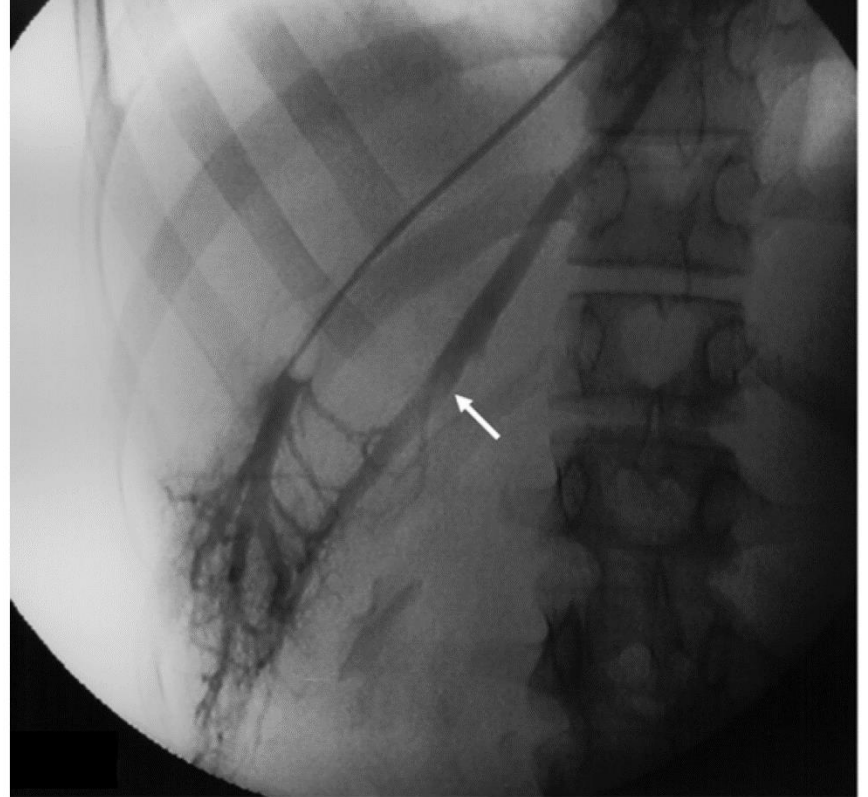
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# Hepatic veno-venous communications

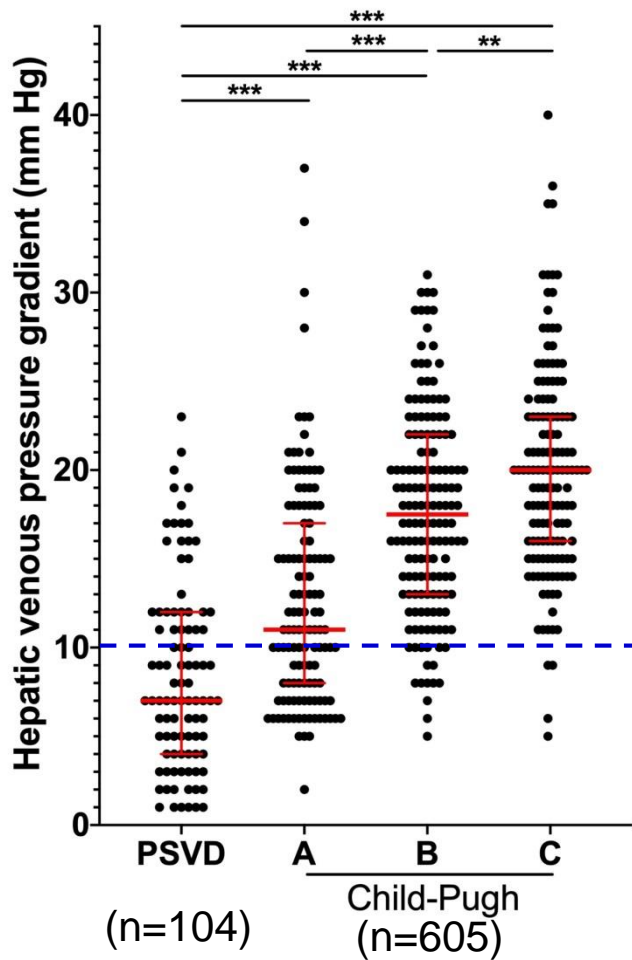
Cirrhosis: 3%



PSVD: 30 to 50%

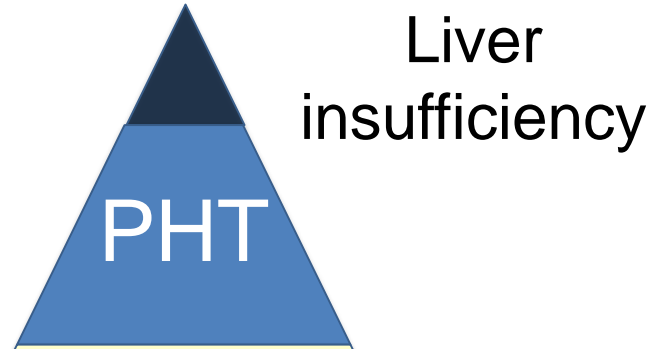


# HVPG: PSVD vs. cirrhosis

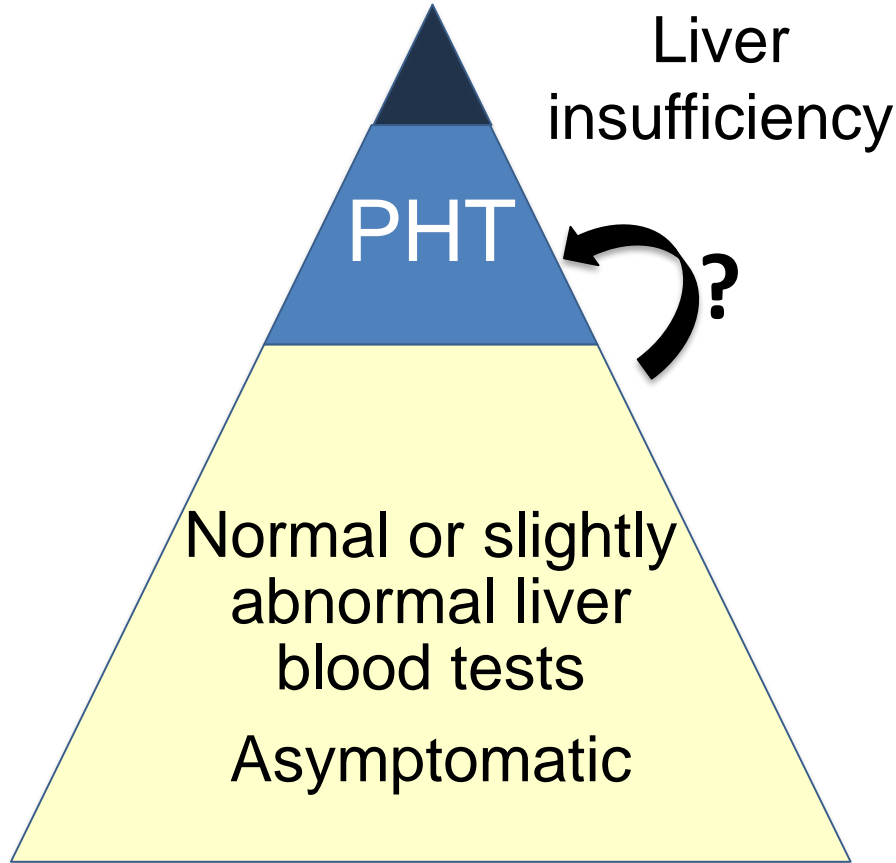


Patients with signs of portal hypertension

# PSVD: manifestations



# PSVD: manifestations



# PSVD: when to suspect?

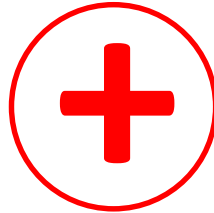
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# Porto-sinusoidal vascular disorder: definition

Biopsy  
> 20 mm  
without cirrhosis



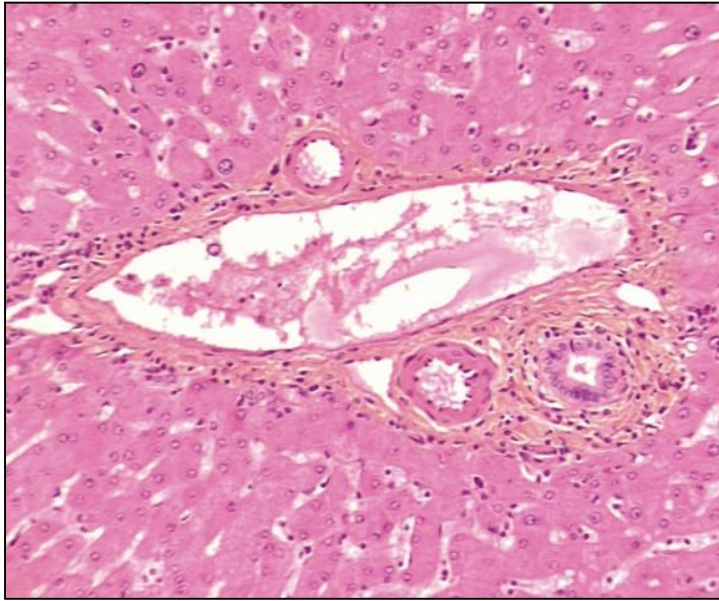
1 specific  
sign

# Signs specific for portal hypertension

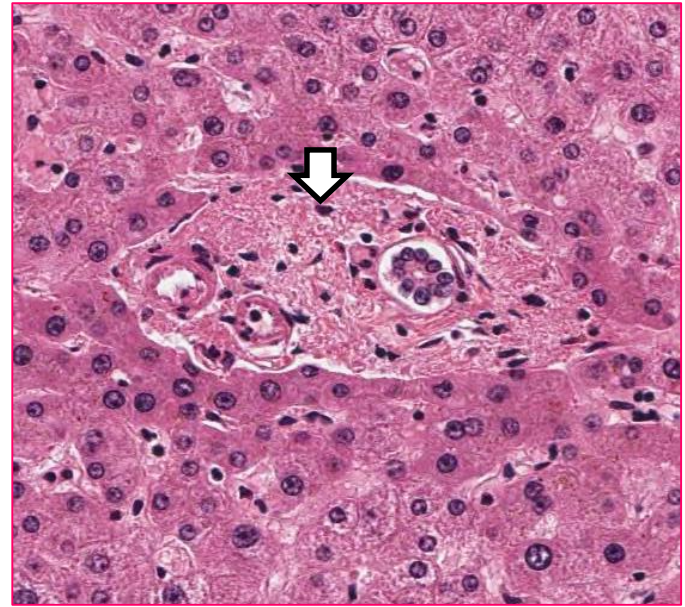
- ✓ Esophageal, gastric or ectopic varices
- ✓ Portal hypertension related bleeding
- ✓ Porto-systemic collaterals at imaging

# Histological lesions specific for PSVD

- Obliterative portal venopathy



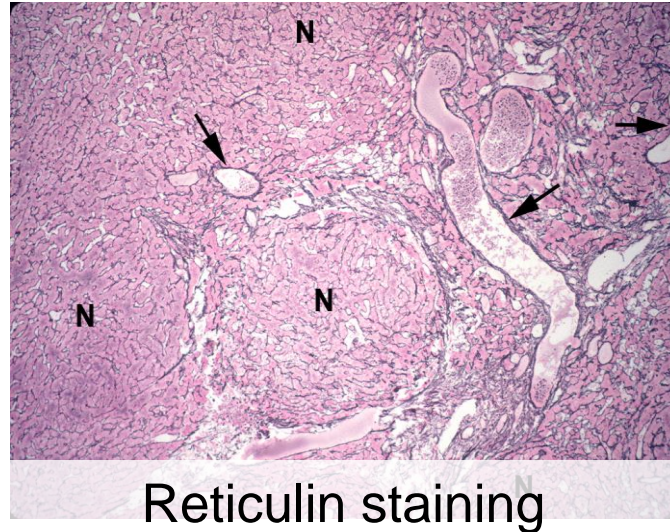
Normal portal tract



Obliterative portal venopathy

# Histological lesions specific for PSVD

- Obliterative portal venopathy
- Nodular regenerative hyperplasia



# 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  $< 150'000/\text{mm}^3$
- Spleen height  $> 13 \text{ cm}$

# Histological lesions non specific for PSVD

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

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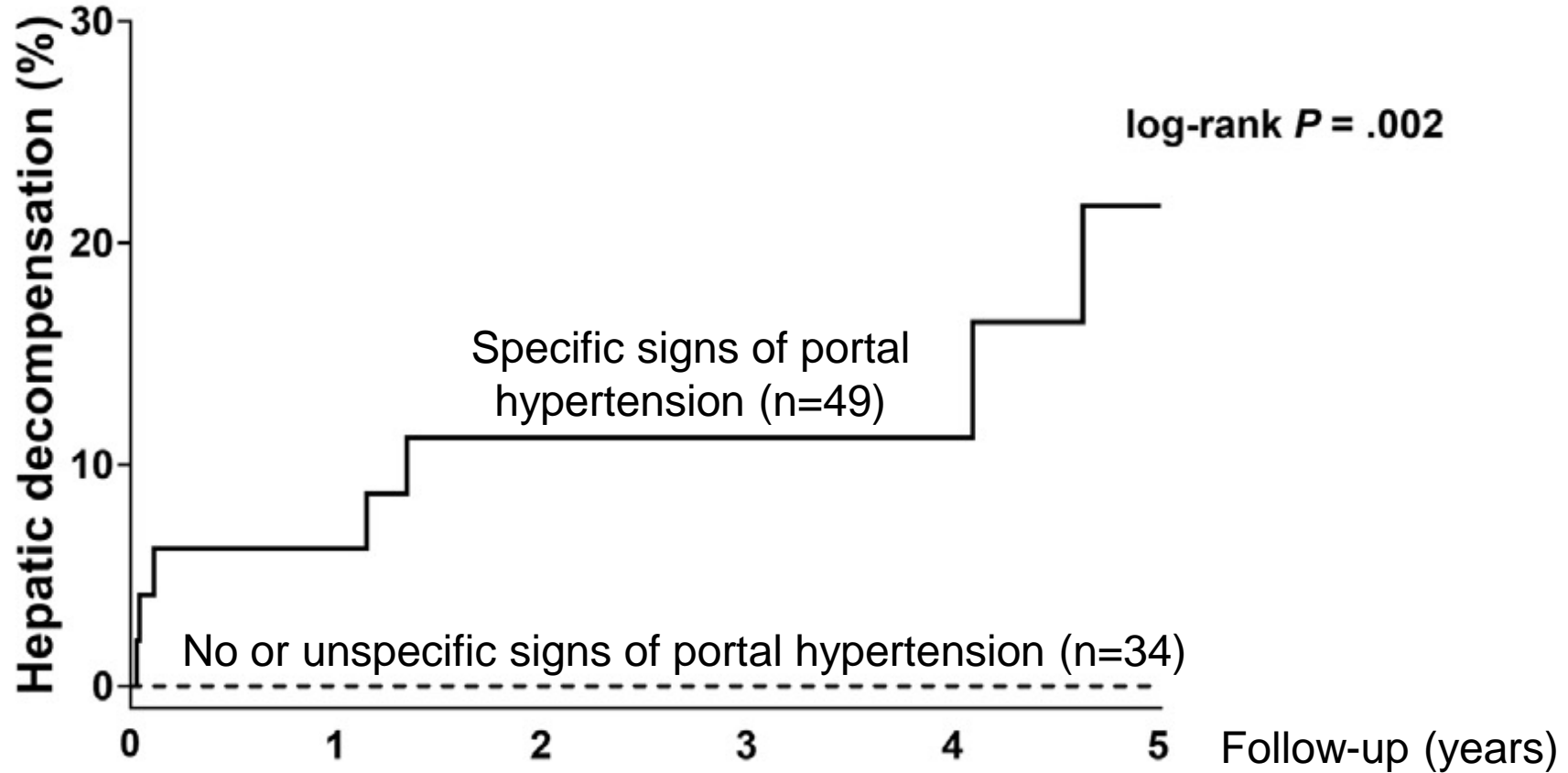
# PSVD: associated conditions

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

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# Outcome of PSVD: portal hypertension



# PSVD: outcome and complications

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

69 patients followed for 7 yrs

# PSVD: treatment

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

# 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

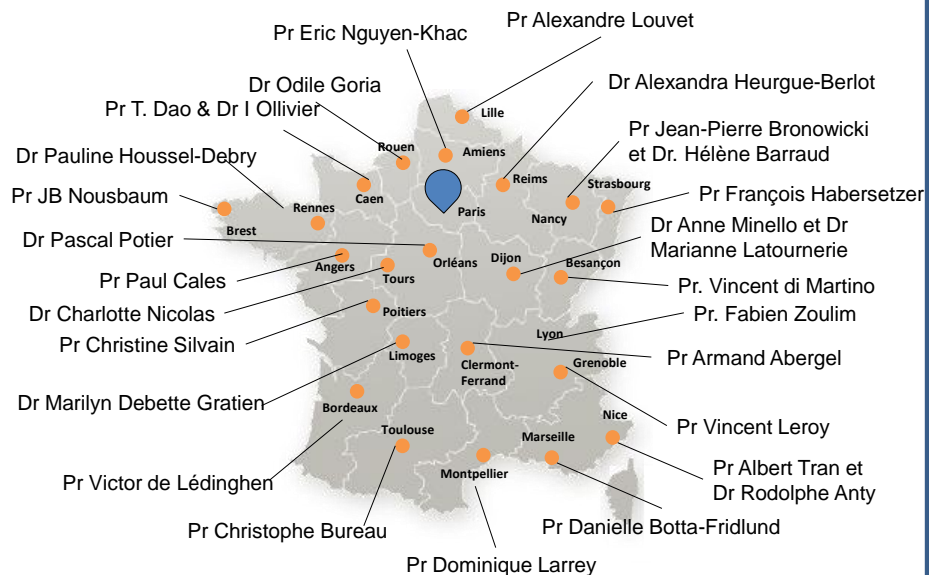
VASCULAR LIVER DISEASE GROUP



## French network for vascular liver diseases



CENTRE DE RÉFÉRENCE  
MALADIES VASCULAIRES  
DU FOIE





Instituts  
thématiques

**Inserm**  
Institut national  
de la santé et de la recherche médicale

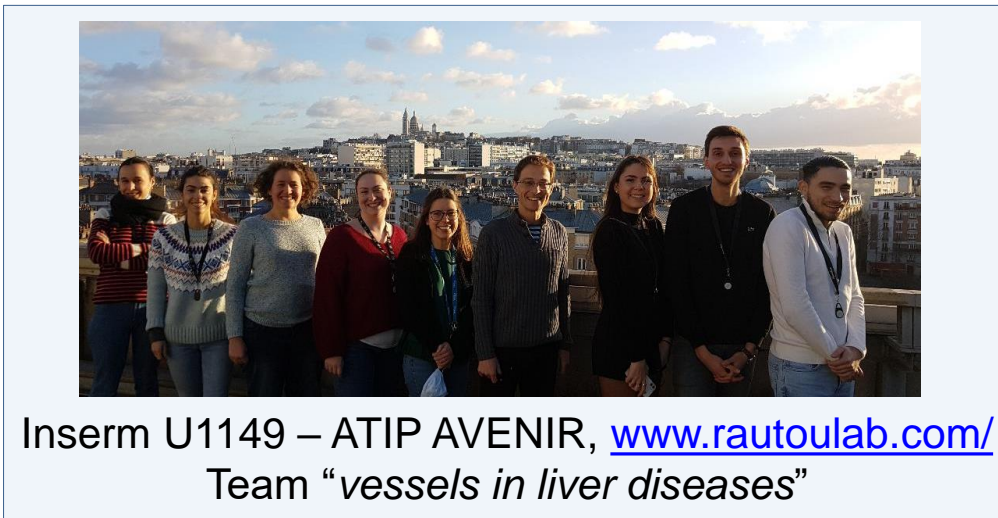


**Université  
de Paris**

**CRi**  
CENTRE de RECHERCHE  
sur l'INFLAMMATION



Hepatology unit, Beaujon Hospital  
Reference center for vascular liver diseases



Inserm U1149 – ATIP AVENIR, [www.rautoulab.com/](http://www.rautoulab.com/)  
Team “vessels in liver diseases”



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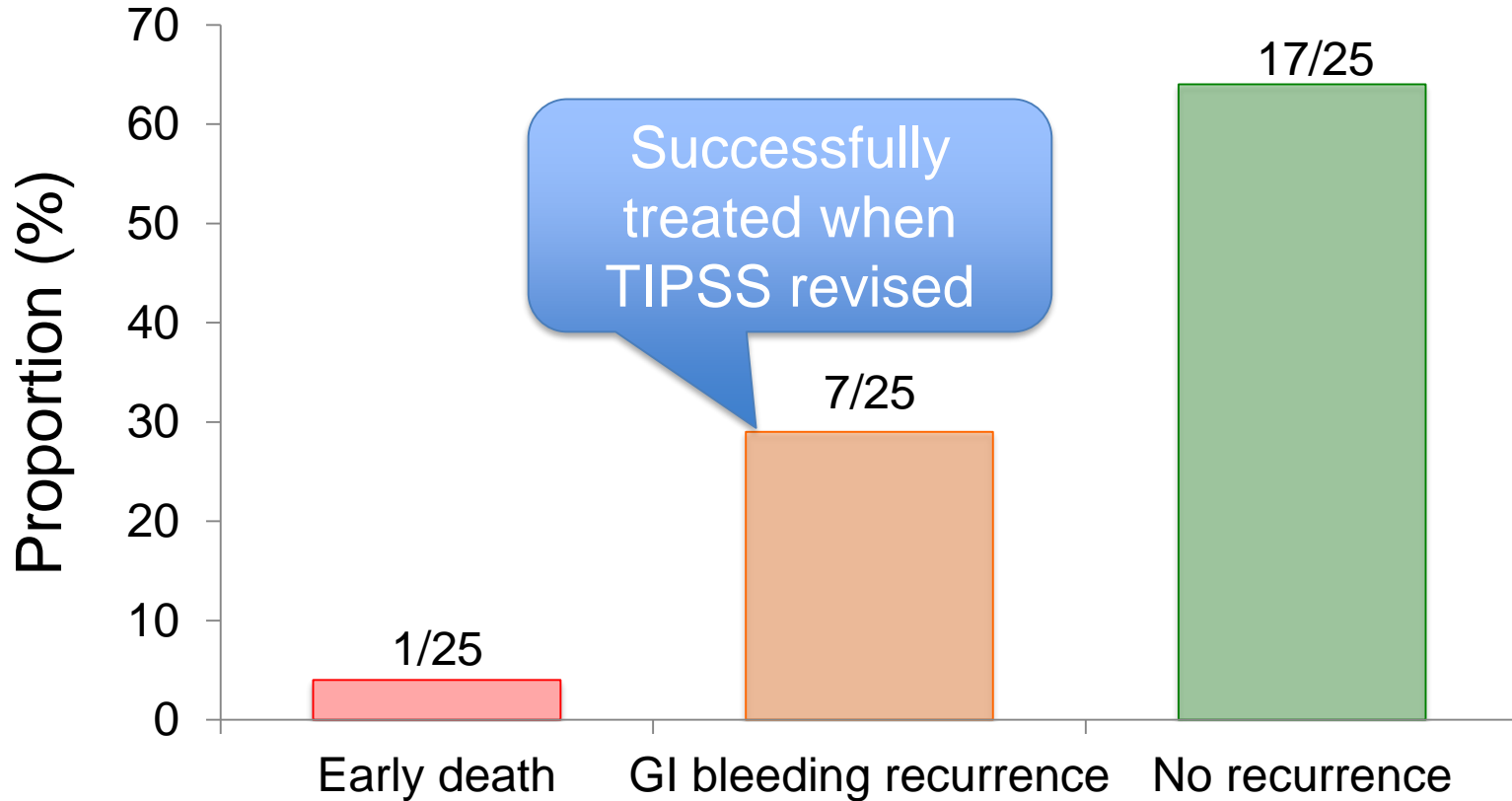
Les Programmes Hospitaliers  
de Recherche Clinique  
(PHRC)



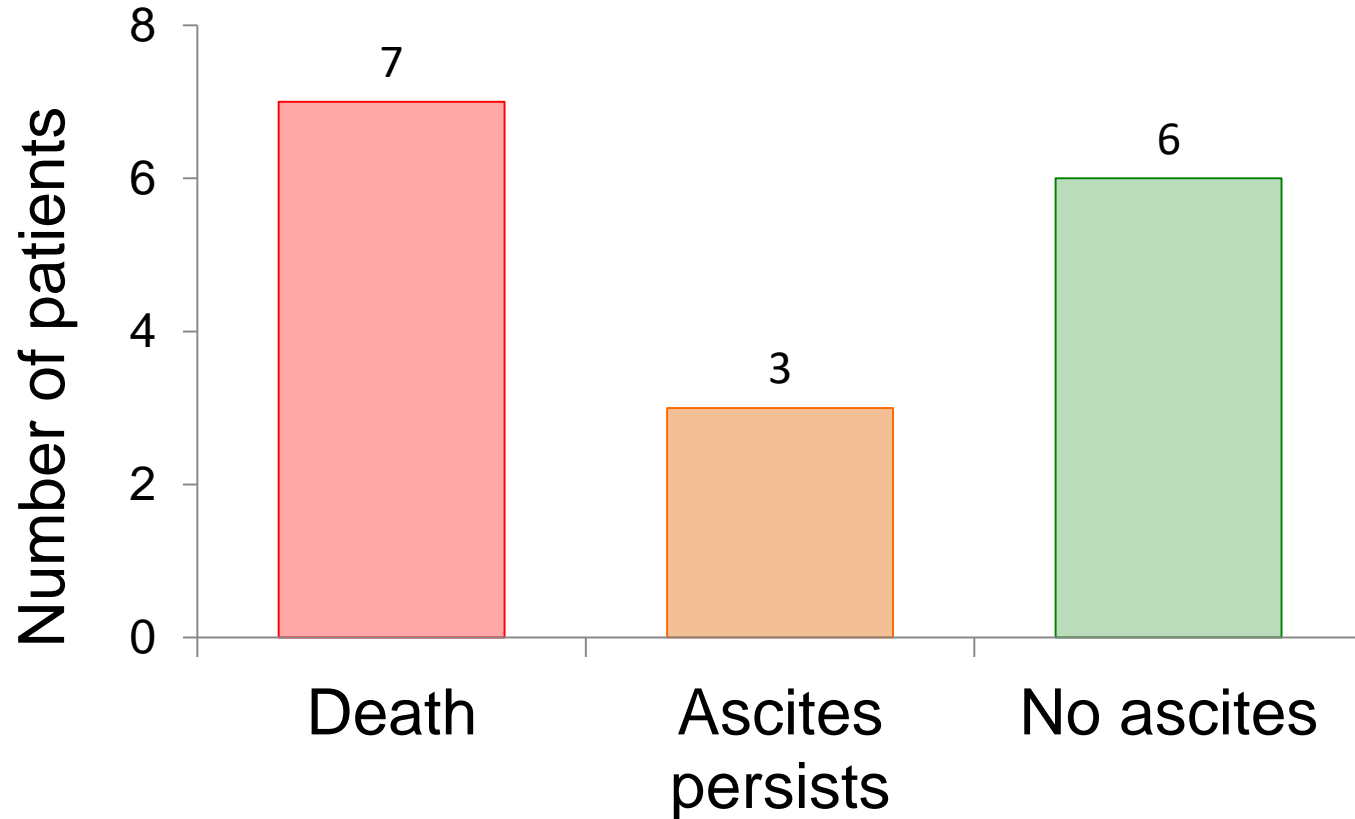




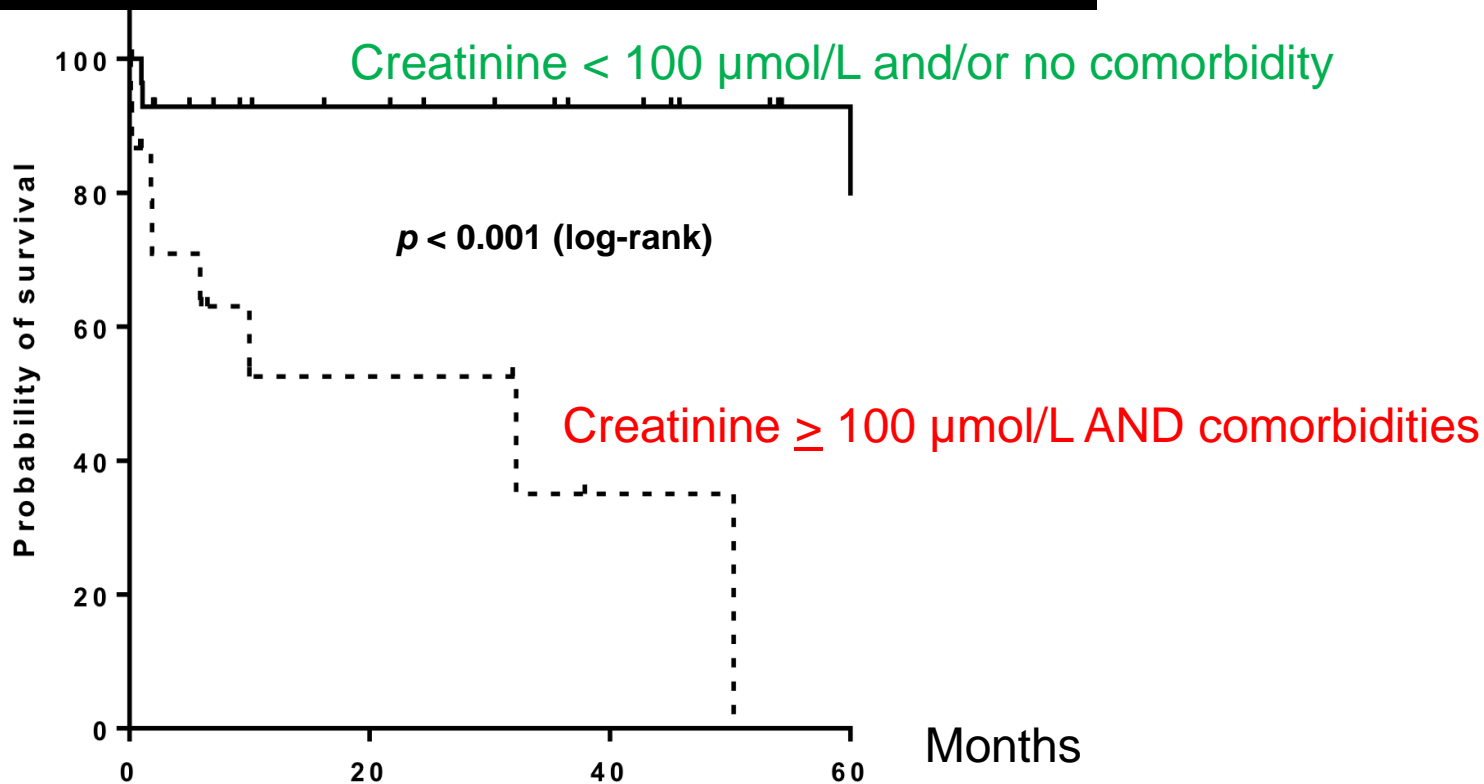
# TIPSS for GI bleeding



# TIPSS for refractory ascites



# Survival after TIPSS



Creatinine  $\geq 100 \mu\text{mol/L}$   
AND comorbidities

Others

7	2	1	0
34	18	12	7

# Liver transplantation PSVD

