
Vascular Liver Disease

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Vascular liver diseases

Arteries

Hepatic venous system

Portal venous system

Sinusoids

Lymphatics

Obstruction
Dilatation
Fistula

Vascular liver diseases causing portal hypertension

Arteries

Hepatic venous system

Portal venous system

Sinusoids

Obstruction

Dilatation

Fistula

Lymphatics

Baveno 2005, 2010, 2015

EASL CPG, J HEP 2016

Obstruction of the splanchnic venous systems

Definition

Etiology

Diagnosis

Treatment

Outcome – Prognosis

62 Baveno VI consensus statements. 1a;A to 5;D

Obstruction of the splanchnic venous systems

HVOTO	Hepatic venous outflow tract obstruction
EHPVO	Extrahepatic portal vein obstruction
IPH	Idiopathic portal hypertension

Obstruction of the splanchnic venous systems

HVOTO

EHPVO

IPH

- In the absence of malignancy
- In the absence of cirrhosis

Obstruction of the splanchnic venous systems

HVOTO

Budd-Chiari syndrome
HV thrombosis, IVC thrombosis
Membranous obstruction of IVC

EHPVO

Portal vein thrombosis
Portal cavernoma

IPH

Hepatoportal sclerosis
Non cirrhotic portal fibrosis
Idiopathic non cirrhotic portal hypertension
Obliterative portal venopathy

Obstruction of the splanchnic venous systems

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HVOTO – EHPVO – IPH

Etiology and Work-up : Baveno VI consensus

- Close collaboration with the hematologist
 - Comprehensive work-up for prothrombotic conditions, systemic diseases, local factors
 - V617F JAK2 and CALR mutations.
Bone marrow biopsy if JAK2/CALR negative
-

Risk Factors for Venous Thrombosis

	HVOTO	EHPVO	IPH
• At least one	84%	67%	20%
• Multiple	46%	18%	-
• Local factor	5%	21%	-

Darwish Murad, Ann Intern Med 2009. Plessier, Hepatology 2010.
Siramolpiwat Hepatology 2014. Cazals Hatem, J Hepatol 2011

Associated conditions

	HVOTO	EHPVO	IPH
• MPN	49%	30%	8%
• APLS	25%	10%	1%
• FV or FII Leiden	15%	16%	7%
• Other systemic	25%	5%	38%

Darwish Murad, Ann Intern Med 2009. Plessier, Hepatology 2010.
Cazals-Hatem, J Hepatol 2011. Siramolpiwat Hepatology 2014.

HVOTO – EHPVO – IPH

Etiology and Work-up : Issues

- Diagnosis of primary deficiency in PC, PS or AT
 - Diagnosis of primary antiphospholipid syndrome
 - Exclusion of myeloproliferative neoplasm
 - Other risk factors (obesity, poverty, etc.)
 - Prothrombotic combinations (chips, scores?)
 - Cause specific treatment and prognosis?
 - Site of thrombosis: related to risk factor?
-

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HVOTO – EHPVO – IPH

Diagnosis : Baveno VI consensus

Non-invasive vessel imaging
Doppler US, MRI, CT

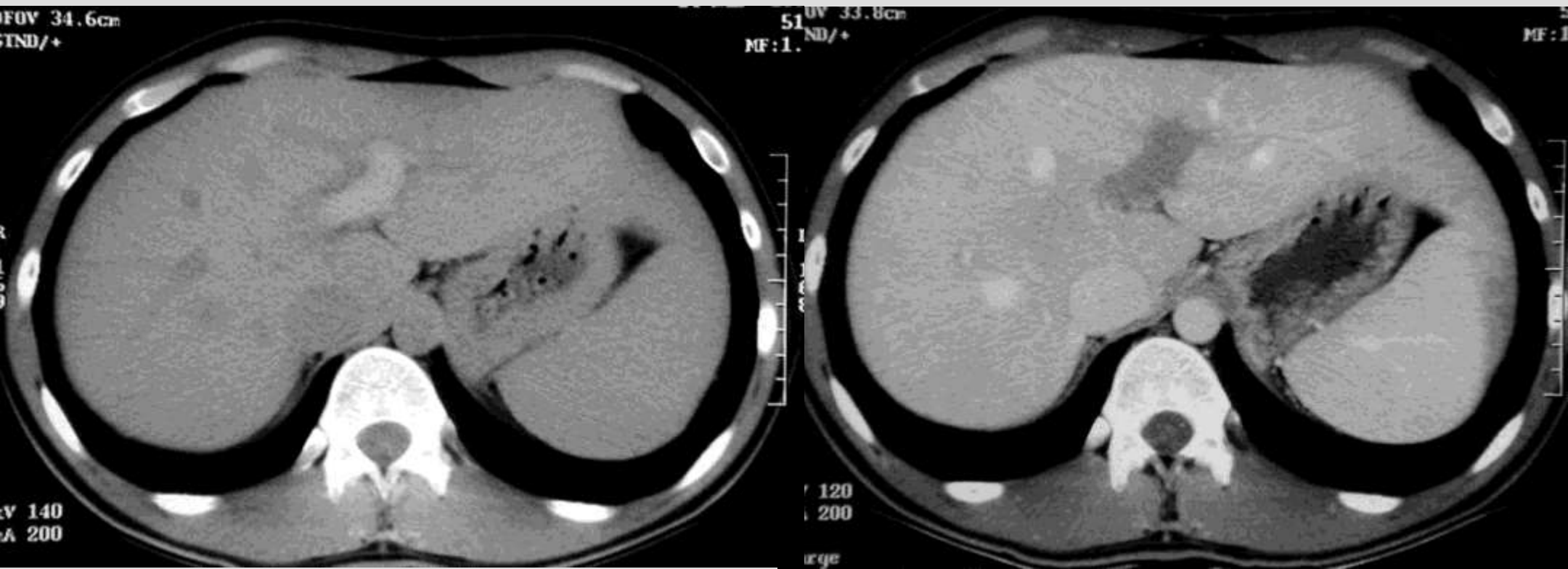
- To demonstrate HVOTO and EHPVO
 - To rule out HVOTO and EHPVO for IPH
-

Budd-Chiari syndrome

Collateral circulation

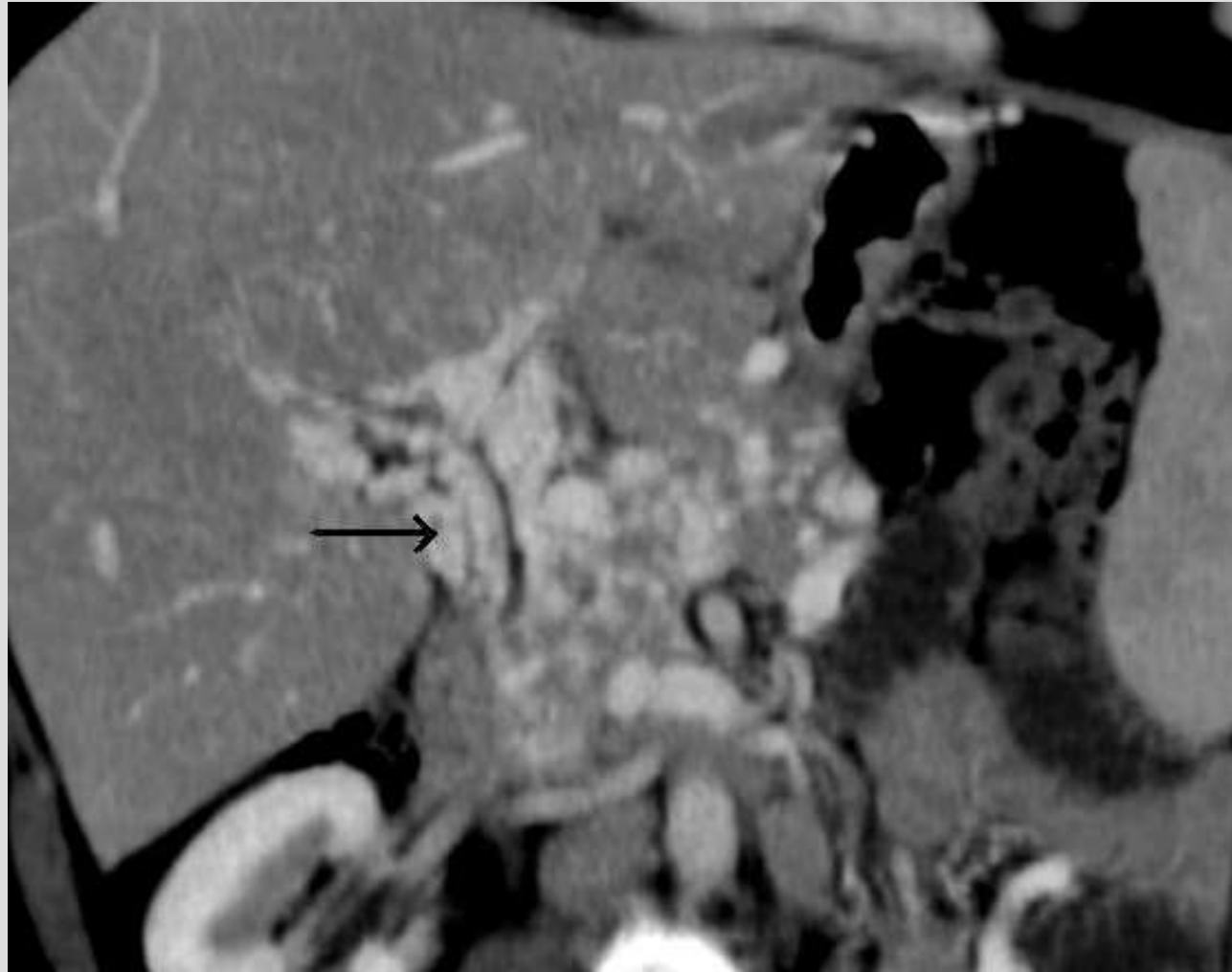


Recent* Portal Vein Thrombosis



* “Acute”

Portal cavernoma



HVOTO – EHPVO – IPH

Diagnosis : Baveno VI consensus

Liver biopsy

- HVOTO : Unnecessary
 - EHPVO : Helpful
 - IPH : Crucial to rule out cirrhosis
-

IPH – Diagnosis : Issues

- From a vague syndrome to discrete entities
 - Non-invasive tools for diagnosis
 - Idiopathic portal hypertension without PHT but typical vascular changes at biopsy?
-

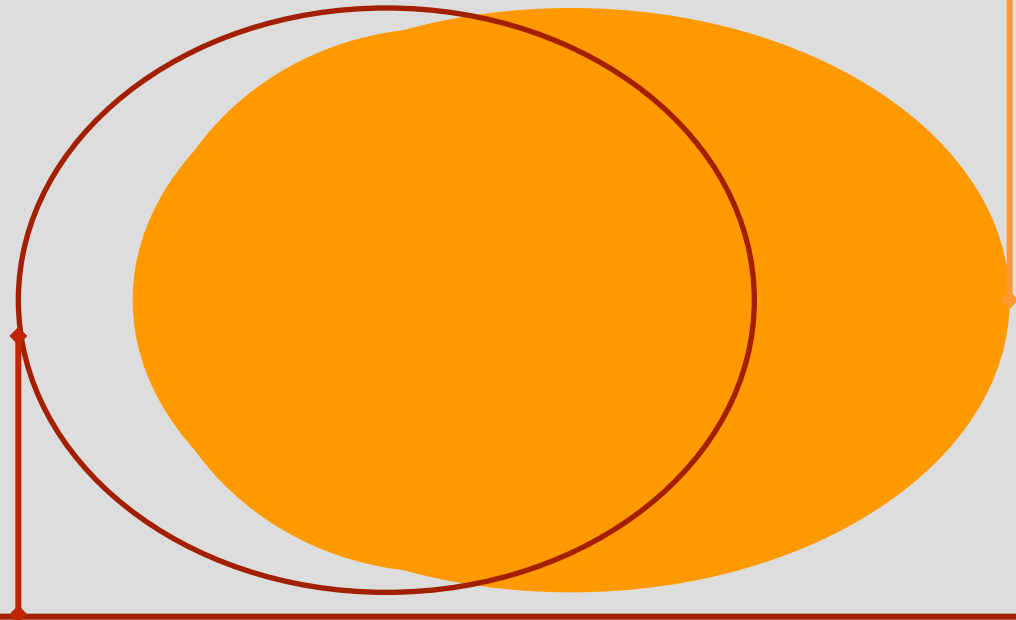
IPH and related entities

Pathology

- Obliterative portal venopathy
- Hepatoportal sclerosis
- Nodular regenerative hyperplasia

Clinics &
Pathology

Idiopathic portal hypertension
Noncirrhotic intrahepatic portal hypertension



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- HVOTO
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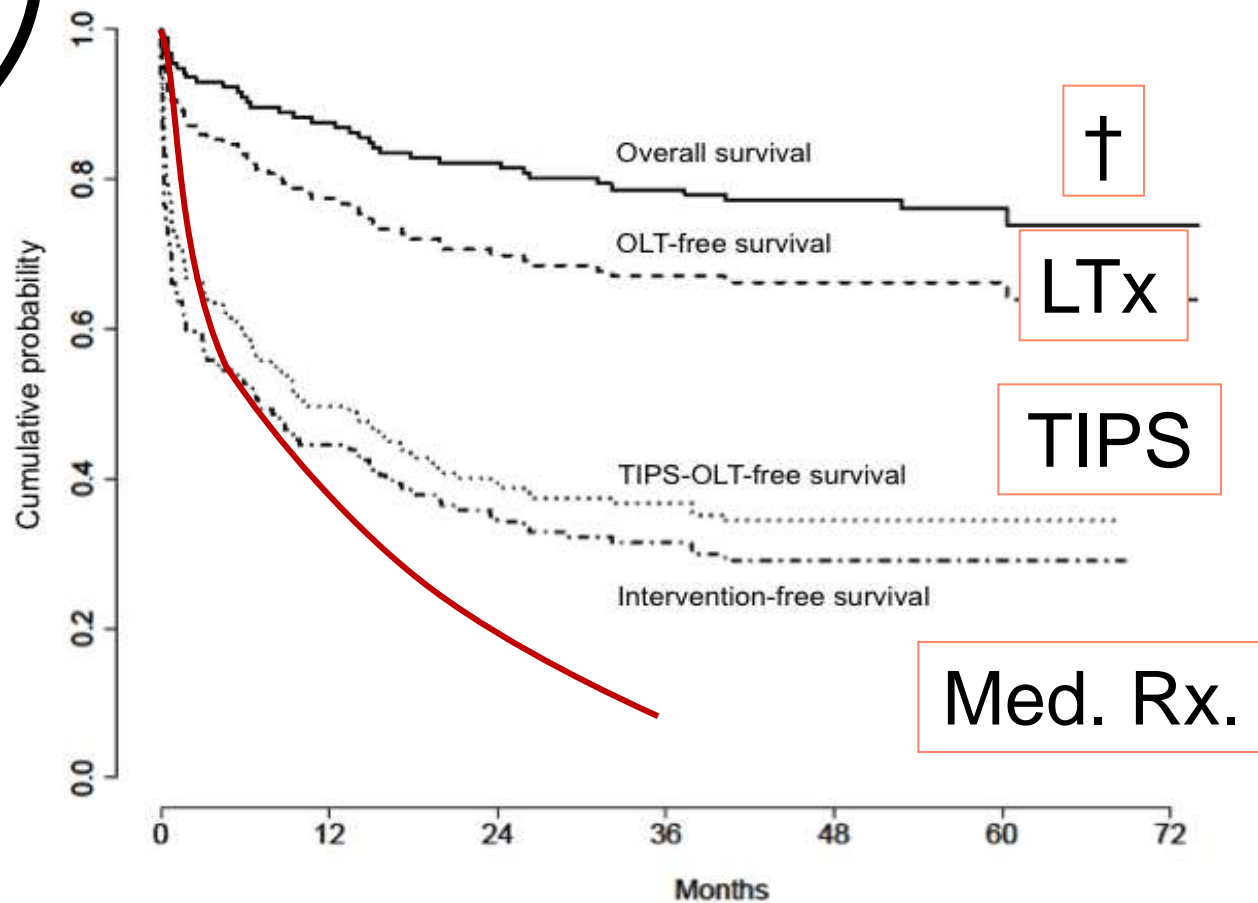
BCS/HVOTO

Anticoagulation
Medical therapy

Angioplasty

TIPS

Transplantation



BCS/HVOTO

Treatment and outcome issues

- Complications of therapy
 - Encephalopathy related to TIPS
 - Bleeding related to anticoagulation therapy
- Prediction of treatment response
- Underlying blood disease
- Regenerative nodules and HCC

Obstruction of the splanchnic venous systems

Definition

Etiology

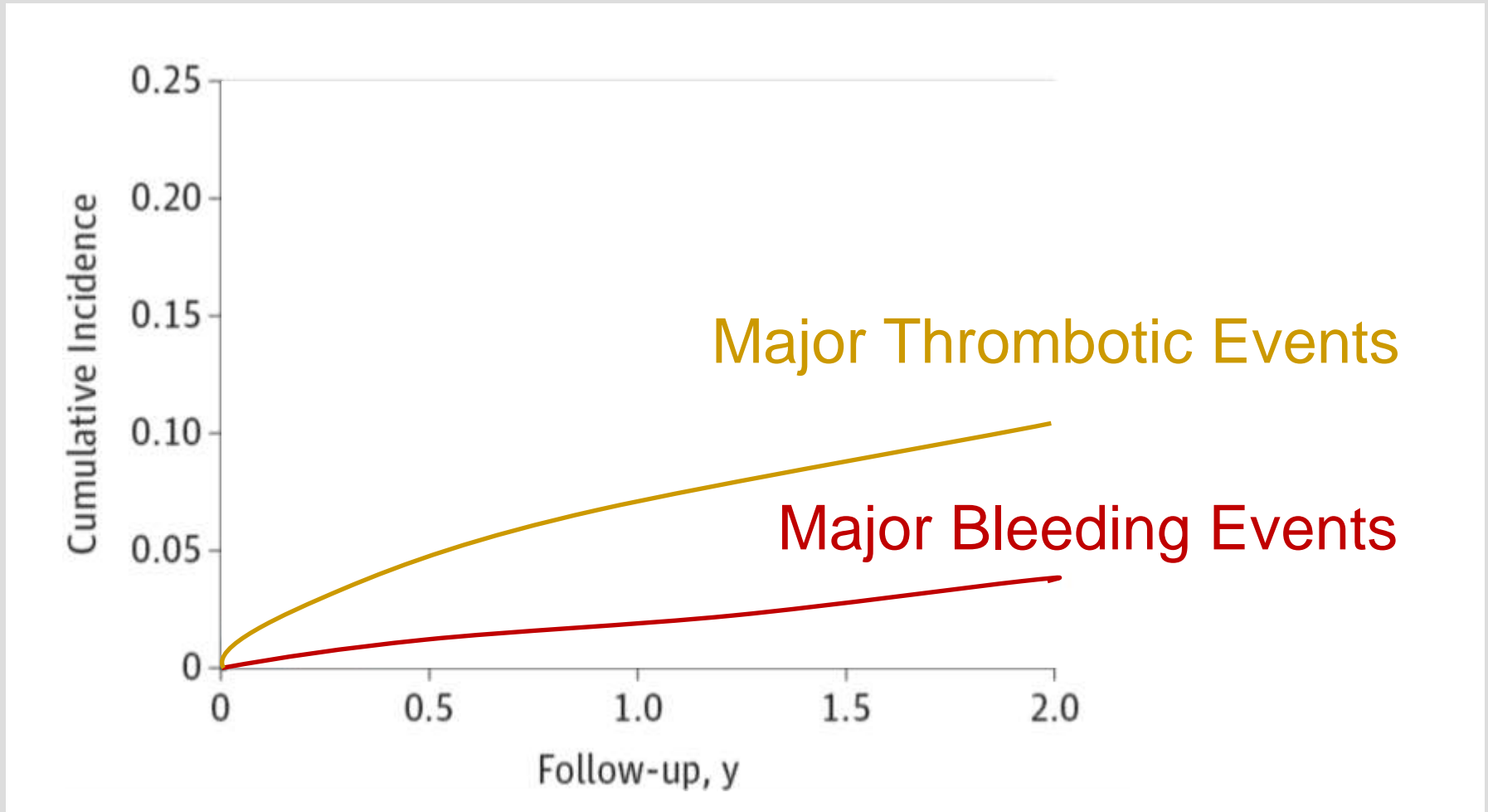
Diagnosis

Treatment

Outcome – Prognosis

- HVOTO
- **EHPVO**
- IPH

Noncirrhotic Splanchnic Vein Thrombosis



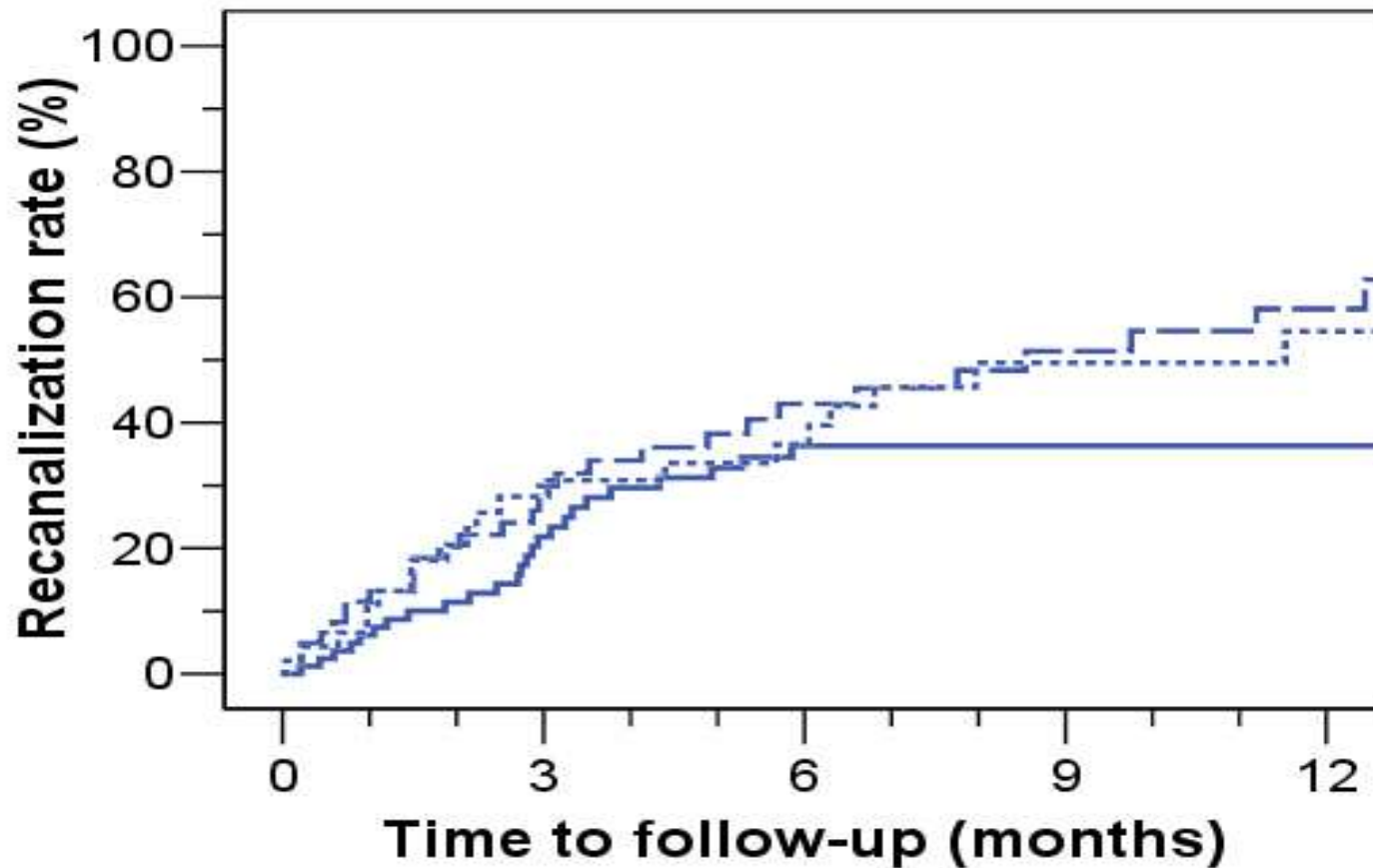
From Ageno, JAMA Intern Med 2015

EHPVO – Therapy : Baveno consensus

- Recent PVT → Immediate anticoagulation
 - Chronic EHPVO → long-term anticoagulation after prophylaxis of bleeding related to PHT
- If
- documented prothrombotic state or
 - recurrent thrombosis or
 - intestinal infarction
-

Recent Portal Vein Thrombosis

EN-Vie Cohort: 95 anticoagulated patients



Sup. mesenteric
Splenic
Portal

EHPVO – Therapy : Baveno consensus

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-

EHPVO – Therapy : Baveno consensus

Management of portal hypertension

- NSBB and endoscopic variceal banding as recommended for cirrhosis
 - Meso-Rex shunt in children
-

EHPVO – Therapy : Issues

Improving efficacy and safety

- Adapting anticoagulation to risk factors
 - Adding antiplatelet agents on
 - Anticoagulation when causes controlled ?
 - New oral anticoagulants
 - Role of TIPS and Meso-Rex in adults
-

Obstruction of the splanchnic venous systems causing portal hypertension

Definition – Nosology

Etiology – Work-up for causes

Manifestations – Diagnosis

Treatment

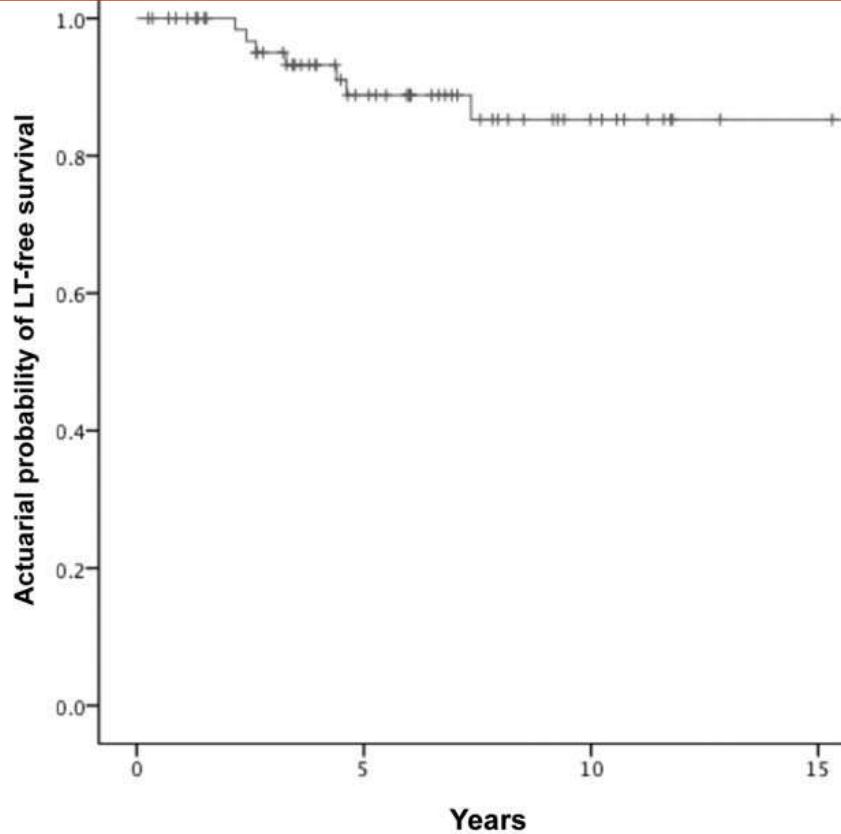
Outcome – Prognosis

- BCS/HVOTO
- EHPVO
- **IPH**

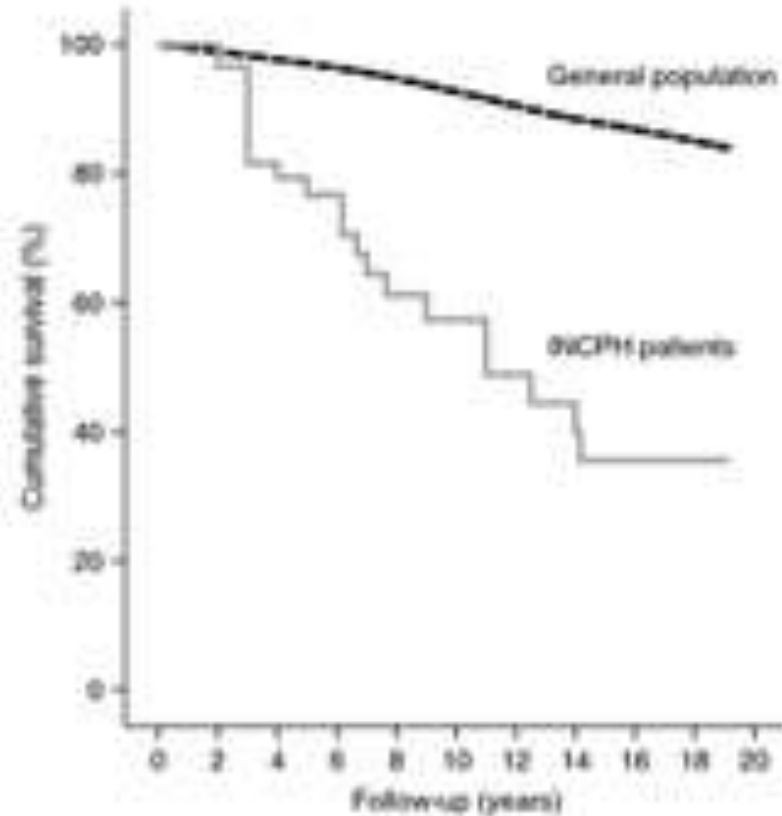
IPH – Therapy : Baveno consensus

- Manage portal hypertension as in patients with cirrhosis
 - Screen for portal vein thrombosis q. 6 mos.
 - Anticoagulation for portal vein thrombosis
-

Survival in patients with IPH



Patients at risk 69 37 14 6



Siramolpiwat, Hepatology 2014

Schouten, APT 2012

IPH – Therapy : Issues

- Mechanisms of disease(s)
 - Anticoagulation for blocking disease progression and preventing complications
-

Obstruction of the splanchnic venous systems

~~HVOTO~~

EHPVO

~~IPH~~

- In patients with cirrhosis

Partial PVT

10% (5-16)

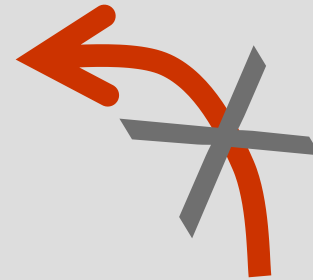
Occlusive PVT

3% (1-4)

PVT in cirrhosis: Baveno consensus

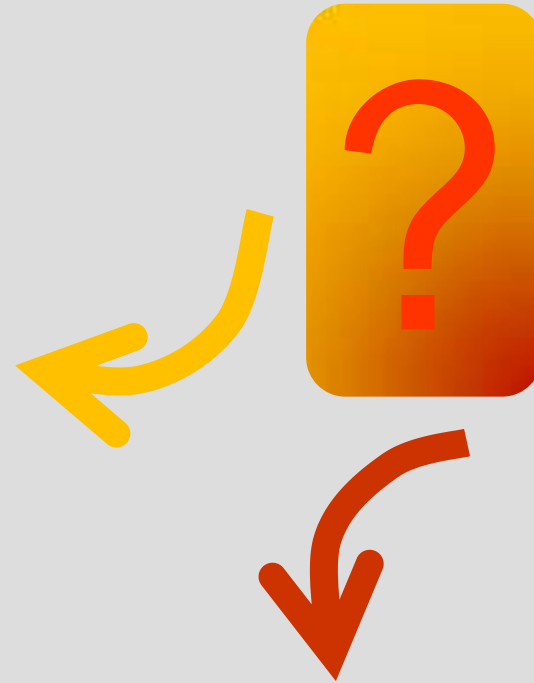
- Screening q. 6 mos in LT candidates
 - Anticoagulation in LT candidates with PVT
-

**Advanced
Cirrhosis**



**Portal vein
Thrombosis**

**Advanced
Cirrhosis**



**Portal vein
Thrombosis**

**Advanced
Cirrhosis**



Enoxaparin

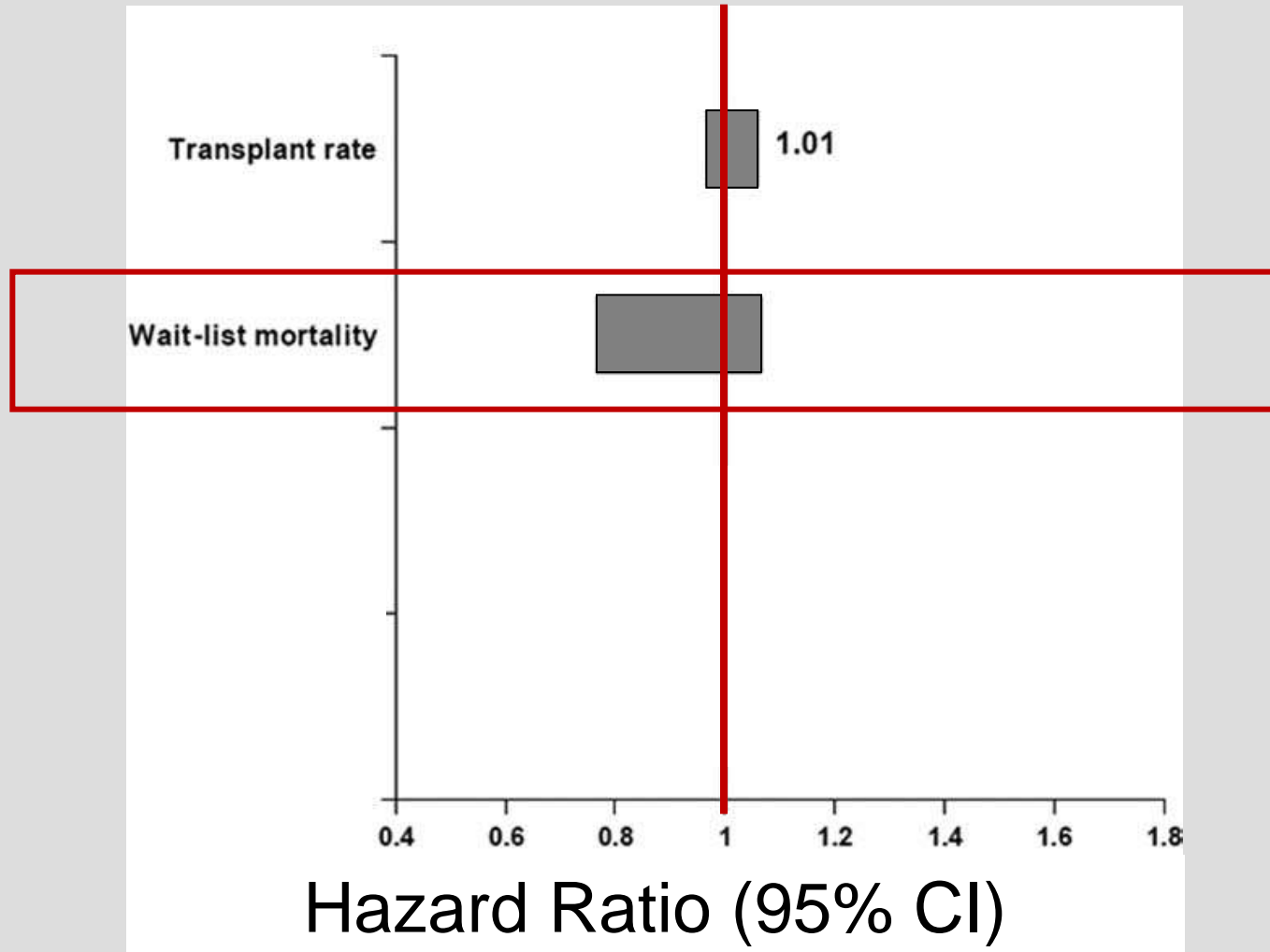
**Portal Vein
Thrombosis**

Villa, E. et al. Gastroenterology 2012
Nery, F. et al Hepatology 2014

PVT in cirrhosis: Issues

- How to anticoagulate/monitor anticoagulation
 - Benefit/risk ratio of anticoagulation therapy for preventing or treating PVT in cirrhosis
 - New oral anticoagulants
-

Impact of PVT before LTx



Englesbe. Liver Transplant 2010. SRTR 22,291 listed candidates. Occlusive PVT 4.02%