Vascular Liver Disorders, Portal Vein Thrombosis & Procedural Bleeding

Hemostasis in cirrhosis, procedural bleeding risk, PVT evaluation/treatment/anticoagulation, and hepatic vein thrombosis (Budd-Chiari syndrome).

2021 AASLD Practice Guidance
Vascular Liver Disorders, Portal Vein Thrombosis, and Procedural Bleeding
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FORMAT *52 guidance statements — expert-consensus format (not GRADE-rated); grouped by clinical theme.*

1 Hemostasis in Cirrhosis — Key Concepts

GS	STATEMENT
1	Simultaneous procoagulant/anticoagulant changes create complex hemostasis not captured by PT, aPTT, or platelet count alone
2	Distinct hypercoagulable and hypocoagulable features may coexist in the same patient, contributing to both thrombosis and bleeding
3	Global hemostasis tests (thrombin generation, TEG/ROTEM) better capture overall status but are not yet clinically validated

2 Procedural Bleeding Risk Assessment

GS	STATEMENT
4	Determining procedural bleeding risk is complex — requires multidisciplinary collaboration before procedures
5	No data-driven INR or platelet cutoff reliably predicts procedural bleeding risk
6	Identify and correct modifiable bleeding risk factors before elective procedures (antithrombotics, AKI, infection)
7	Given low bleeding risk of most common procedures, routine prophylactic platelet transfusion is not evidence-supported
8	Individualize approach to severe thrombocytopenia pre-procedure — no definitive evidence for correction safety/efficacy
9	INR should NOT be used to gauge procedural bleeding risk in patients not on vitamin K antagonists (VKAs)
10	Measures to reduce INR (e.g., FFP) are not recommended pre-procedure in patients not on VKAs
11	FFP transfusion pre-procedure carries risk without proven benefit
12	Low fibrinogen is associated with increased bleeding risk in critically ill cirrhosis patients
13	Cryoprecipitate/fibrinogen concentrate are effective low-volume options to raise fibrinogen when indicated

3 Portal Vein Thrombosis — Evaluation

GS	STATEMENT
14	Test for occult myeloproliferative neoplasm (MPN) even when CBC is not overtly suggestive
15	MPN testing negative + thrombocytosis or polycythemia vera concern → pursue further testing
16	Outside LT candidates, unclear whether PVT independently worsens mortality vs. reflects portal hypertension severity
17	PVT at time of transplant associates with increased posttransplant mortality; insufficient data to recommend pretransplant PVT treatment specifically to improve posttransplant outcomes

4 PVT — Treatment

GS	STATEMENT
18	Recent PVT + intestinal ischemia concern: immediate multidisciplinary consult (surgery, critical care, IR, hematology); anticoagulation essential; surgery for infarction
19	Non-cirrhotic recent PVT: directed antithrombotic therapy to avoid ischemia and prevent chronic PVT/portal hypertension
20	Cirrhotic PVT without ischemic symptoms: weak trial data — decide case-by-case, weighing clot-extension risk vs. bleeding/LT candidacy
21	Recent small intrahepatic sub-branch thrombosis or <50% main PV occlusion: observation with serial imaging q3mo is reasonable
22	Recent occlusive or >50% partial occlusion of main PV/mesenteric veins: anticoagulation favored to prevent progression
23	Thrombolytic therapy (local/systemic) only for selected recent PVT with persistent ischemia despite anticoagulation
24	PVR + TIPS for LT candidates with chronic PVT hindering physiological graft anastomosis (multidisciplinary decision)
25	PVR + TIPS for chronic PVT with recurrent bleeding/refractory ascites unmanageable medically or endoscopically

5 PVT — Anticoagulant Selection

GS	STATEMENT
26	Choice of anticoagulant (LMWH, VKA, DOAC) should be individualized; consult hematology/expert hepatology
27	Therapeutic anticoagulation bleeding rates in cirrhosis resemble the general population; portal-hypertensive bleeding is not increased by anticoagulation
28	DOAC data in PVT remain limited — caution with advanced portal hypertension; seek expert consultation

6 Hepatic Vein Thrombosis / Budd-Chiari Syndrome

GS	STATEMENT
29	Consider HVT/BCS in any liver disease of unknown etiology, especially with a known prothrombotic disorder
30	Doppler US (experienced operator) is first-line imaging; MR/CT for confirmation and intervention planning
31	Full thrombophilia workup at diagnosis — investigate additional factors even after one cause identified; hematology consult
32	HCC surveillance as in general cirrhosis population — ultrasound q6mo ± AFP
33	Progressive "step-up" therapy (least→most invasive); early referral to expert tertiary centers
34	All HVT/BCS patients should receive therapeutic anticoagulation, even without an identified prothrombotic disorder
35	TIPS/DIPS with PTFE-covered stents is treatment of choice when medical therapy/angioplasty fail or are infeasible

7 Sinusoidal Obstruction Syndrome (SOS)

GS	STATEMENT
36	Consider SOS 1–3 weeks post-HSCT with RUQ pain, edema-related weight gain, hepatomegaly, and jaundice
37	UDCA is recommended prophylaxis for SOS in all patients undergoing HSCT

8 Liver Vascular Malformations (HHT)

GS	STATEMENT
38	Suspect HHT-associated LVMs with liver bruit + CT/MR showing enlarged hepatic artery and heterogeneous enhancement, especially with family history
39	Asymptomatic LVMs do not require therapy or surveillance imaging
40	Manage symptomatic LVMs by treating the specific complication (heart failure, portal hypertension, biliary ischemia)
41	Symptomatic patients need multidisciplinary specialized-center care; consider bevacizumab and/or LT for standard-therapy nonresponders

9 Idiopathic Noncirrhotic Portal Hypertension

GS	STATEMENT
42	Consider INCPH in any patient with portal hypertension evidence but without cirrhosis or other known cause
43	Liver biopsy required to exclude cirrhosis; reticulin stain and specialized liver pathology review helpful
44	Evaluate for venous thrombosis risk factors, immune disorders, and inherited conditions associated with INCPH
45	PVT may be more frequent in INCPH than cirrhosis, but routine PVT screening is not recommended; HCC is rare

10 Hepatic & Splenic Artery Aneurysms

GS	STATEMENT
46	Doppler US or CT for detection; multidetector CT angiography for characterization/treatment planning
47	If intervention needed, endovascular repair first-line (open repair if inappropriate) — multidisciplinary decision
48	Aneurysms <2 cm: early follow-up imaging at 3 and 12 months to assess growth; significant growth prompts intervention
49	Urgent intervention for symptomatic or complicated HAAs/SAAs

11 Pediatric Considerations

GS	STATEMENT
50	Congenital/acquired pediatric vascular liver disease: early referral to centers with pediatric hepatology/hepatobiliary surgery/LT expertise
51	Children with EHPVO: evaluate for early intervention at the presymptomatic stage at an expert center
52	Infantile hepatic hemangiomata (~30–50% of cases): propranolol (beta-blocker) is recommended

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