- Acute liver failure is not likely to reduce plasma cholinesterase levels, so succinylcholine may be used if indicated.
- Increased bioavailability of IV drugs if serum albumin concentration is decreased.
- Limit sedative drugs.

Maintenance

- Inhalational agent with high inspired O_2 concentration is useful for maintaining hepatic blood flow and O_2 supply; halothane should probably be avoided.
- Effect of muscle relaxants with hepatic clearance may be prolonged.
- Increased blood loss with coagulopathy.

Extubation

- Postop mechanical ventilation to ensure time for adequate metabolism of depressant drugs

Adjuvants

- Hypocalcemia can occur with citrate administration.

Anticipated Problems/Concerns

- Worsening of hepatic or renal function
- Delayed awakening from prolonged drug metabolism or encephalopathy
- Need to protect airway with reduced consciousness
- Hypoglycemia

Hepatopulmonary Syndrome

Gregory Hertel | Gaurav Malhotra

Risk

- Occurs in up to 10–32% of pts with cirrhosis.
- Dyspnea is present in up to 70% of cirrhotic pts for varying reasons (ascites, ILD, volume overload, anemia).

Perioperative Risks

- Hypoxemia, often worsened on induction and post LT
- Aspiration
- Hemodynamic instability and CV collapse
- Acute/chronic renal insufficiency
- Myocardial infarction

Worry About

- Full stomach and aspiration risk in presence of ascites and increased intraabdominal pressures
- Hypoxemia (exacerbated in immediate post LT period)
- Severe post LT hypoxemia and possible RV failure related to pulm vasoconstriction from an abrupt change in vascular mediators from the new liver
- Hemodynamic instability, especially related to reperfusion during LT

Overview

- Pulm complication of cirrhosis resulting in arterial hypoxemia.
- Defined as triad of liver disease, intravascular pulm vasodilatation, and abnormal gas exchange.
- Cirrhosis pts with hepatopulmonary syndrome have a higher mortality than those without it.
- Liver transplant is the only definitive treatment.

Etiology

- Involves a widespread vasodilatation of the precapillary pulm arterioles up to 100 μm.
- Overall understanding of the pathogenesis of HPS is limited.
- Animal models suggest increased endothelin (and subsequently increased NO), pulm monocytes, and VEGF all contribute to pulm vasodilatation and angiogenesis, which in turn contribute to oxygen impairment.
- Increased vessel diameter in addition to impaired hypoxic pulm vasoconstriction results in increased flow across the capillary bed without an increase in alveolar ventilation, causing a V/Q mismatch.
- Hypoxemia is exacerbated by inability of oxygen at room air concentration to diffuse to blood at the center of the dilated vessels.

Usual Treatment

- Liver transplantation remains the only effective treatment for HPS.
- Angiogenesis hypothesis is supported by the observation that correction of hypoxemia is not immediate post LT and may take up to a year; NO returns to normal post LT.
- TIPS has not been shown to be consistently beneficial in HPS.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Volume overload Hypotension Cirrhotic cardiomyopathy (increased CO, decreased ventricular response to stress) Postreperfusion syndrome (LT) CAD LVOT obstruction	Poor exercise tolerance CAD/MI	Vital signs Pitting edema	Monitor BP and HR ECG and ECHO
RESP	V/Q mismatch Hypoxemia Hepatic hydrothorax Portopulmonary Htn	Dyspnea, tachypnea	Orthodeoxia- worsened hypoxia with standing as flow increases to larger vessels in lung bases	Monitor pulse oximetry and RR, increased A-a gradient (>15 mm Hg) ABG Contrast-enhanced TTE
GI	Ascites Portal Htn Esophageal varices SBP	Abdominal distension Hematemesis, melena	Fluid wave Guarding and rebound (SBP)	Abdominal x-ray, CT scan, US WBC count, peritoneal fluid analysis
RENAL	Hepatorenal syndrome Hypervolemia Hyponatremia	Oliguria Peripheral edema	Vitals signs	UA, serum lytes, BUN and Cr
HEME	Coagulopathy (decreased clotting factors and thrombocytopenia)	Easy bruising/bleeding	Purpura	INR/PT, PTT, fibrinogen, CBC Thromboelastogram
CNS	Hepatic encephalopathy Intracranial Htn Cerebral edema	Confusion Coma	GCS Asterixis	Head CT Serum ammonia

Key References: Dalal A: Anesthesia for liver transplantation, *Transplant Rev (Orlando)* 30(1):51–60, 2016; Koch DG, Fallon MB: Hepatopulmonary syndrome, *Clin Liver Dis* 18(2):407–420, 2014.

Perioperative Implications

Preoperative Preparation

- Baseline ABG to evaluate severity of hypoxemia
- ECHO to evaluate cardiac function
- Thorough H+P and consent, including risks of anesthesia and full anticipated lineup

Monitoring

- Standard monitors
- Urinary cath
- Arterial line for frequent ABGs to assess hypoxemia and hemodynamic monitoring
- Consider central line with pulm arterial cath and SvO_2.
- Consider possible VV bypass for LT.
- Consider intraop TEE.

Airway

- Ensure adequate preoxygenation.
- Ideally utilize tools for rapid intubation (video laryngoscopy).
- Full stomach precautions.

Induction
- Rapid sequence induction with cuffed ETT in setting of ascites or full stomach.
- Induction alone may worsen hypoxemia.
- Anticipate hypoxemia and hemodynamic instability in setting of decompensated cirrhosis and HPS.
- Ketamine decreases hepatic blood flow; propofol increases it.

Maintenance
- Higher FIO_2 and PEEP throughout case augment oxygenation.
- Standard maintenance with adequate muscle relaxation.
- Trendelenburg positioning if tolerated by surgical needs.
- All inhaled anesthetics decrease MAP and portal blood flow.
- Allow adequate preparation for transfusion, adequate access, and readily available products (RBCs, FFP, plts, cryoprecipitate).

Extubation
- Extubate only if conditions optimized with pt awake, strong, and with assuring ABG, with caution given to potential for severe postop hypoxemia
- Low threshold to remain intubated with plan for SICU postop
- Postop period
- Supplemental oxygen therapy
- May require PEEP to improve oxygenation
- In case of severe postop hypoxemia:
  - Trendelenburg positioning
  - Inhaled vasodilators (epoprostenol and NO, selectively targeting constricted normal vessels in the more ventilated middle and upper lobes);
  - IV methylene blue (vasoconstrictor preferentially targeting dilated vessels in the bases);
  - Embolization of lower lobar pulm vessels;
  - ECMO.

Anticipated Problems/Concerns
- Hypoxemia
- CV instability
- Coagulopathy

Hereditary Hemorrhagic Telangiectasia

(Osler-Weber-Rendu Disease)

Rishi Chokshi | Lee A. Fleisher

Risk
- Effects vary in racial and ethnic groups, with a wide geographic distribution.
- Men and women affected equally.
- In Vermont, frequency is 1:16,500.
- In Europe and Japan, frequency is 1:5000–8000.

Perioperative Risks
- Excessive bleeding
- Paradoxical air, bland, or septic embolism to brain

Worry About
- Chronic anemia due to hemorrhage, especially recurrent epistaxis.
- Due to danger of intrapartum or postpartum pulm hemorrhage, a pregnant woman with HHT who has not had a recent pulm evaluation should be evaluated as soon as pregnancy is recognized.

Overview
- Mucocutaneous and visceral vascular dysplasia can occur.
- Combination of defective perivascular connective tissue, insufficient smooth muscle contractile element, endothelial cell junction defects, and increased endothelial tissue plasminogen activator impairing thrombus formation in case of vascular damage.
- International consensus diagnostic criteria (Curacao criteria) indicates HHT diagnosis classified as definite if three criteria present, possible or suspected if two criteria present, and unlikely if one criterion present. The criteria are:
  - Epistaxis: Spontaneous recurrent nosebleeds.
  - Mucocutaneous telangiectasia.
  - Visceral involvement (i.e., GI telangiectasia, pulm AVM, hepatic AVM, cerebral AVM, spinal AVM).
  - Affected primary relative.
- Manifestations of HHT are not present generally at birth but develop with increasing age, with epistaxis usually being the earliest sign that may lead to chronic anemia. About 90% of pts have signs and symptoms by age 40.

Etiology
- Autosomal dominant trait with varying penetrance and expressivity

Usual Treatment
- Epistaxis is medically treated with Fe supplementation, estrogen therapy, and humidification. With intractable epistaxis ablative therapy with Nd:YAG laser is effective, although multiple treatments are required.
- Multiple transfusions.
- Pulm AVMs with feeding artery diameter ≥3 mm require treatment with transcatheter embolotherapy with coils.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Telangiectasia of nasal mucosa, conjunctival telangiectasias, retinal vascular malformations	Recurrent frequent epistaxis		
CV	High-output heart failure, thromboembolism	Fatigue, SOB	Rales, neurologic deficits	CXR
RESP	AVMs with R-to-L shunt leading to hypoxemia, absence of filtering capillary bed allowing particulate matter to reach systemic circulation, fragile vessels may hemorrhage into bronchus or pleural cavity	Fatigue, dyspnea on exertion, hemoptysis, embolic cerebral events	Cyanosis, clubbing, neurologic deficits	CXR, CT, detection of R-to-L shunt via radionuclide perfusion scans or contrast ECHO
HEME	Anemia, coagulopathy, associated with von Willebrand disease	Recurrent epistaxis	Pallor	CBC, PT/INR, PTT
CNS	Cerebral AVM, aneurysms, cavernous angiomas paradoxical embolism, spinal AVM, migraines	CVA, brain abscess	Headache, seizure, hemorrhage, ischemia of the surrounding tissues due to a steal effect	MRI
HEPAT	Hepatomegaly, high output heart failure, portal Htn, encephalopathy, biliary disease	Hemorrhage, sepsis	Jaundice	LFTs, PT/INR, PTT

Key References: Lomax S, Edgcombe H: Anesthetic implications for the parturient with hereditary hemorrhagic telengiectasia, *Can J Anaesth* 56(5):374–384, 2009; Weingarten TN, Hanson JW, Anusionwu KO, et al.: Management of patients with hereditary hemorrhagic telengiectasia undergoing general anesthesia: a cohort from a single academic center's experience, *J Anesth* 27(5):705–711, 2013.

Perioperative Implications

Preoperative Preparation
- Preop cardiac and pulm evaluation to exclude high-output cardiac failure and pulm AV malformations, which are often asymptomatic.
- CBC for anemia from bleeding or polycythemia from pulm shunt.
- Check liver and renal function.
- Perform neurologic assessment to exclude previous paradoxical emboli and severe brain AVM.
- Debubble IV lines and add air filters to prevent paradoxical air emboli.
- Use meticulous aseptic technique.