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.

- For regional technique, assess any possibility of AVMs in the neuraxial region prior to performing the technique.

Monitoring

- Avoid or use with great caution TEE, gastric suctioning or esophageal stethoscope if esophageal varices or AVMs are present, and avoid nasal temperature probes.

Airway

- If oropharyngeal AVMs are present, there is a high risk of airway bleeding.
- Nasal intubation contraindicated if nasal telangiectasias are present.
- Well-lubricated smaller size ETT to prevent any tissue trauma.

Maintenance

- When there is a risk of high-output heart failure and liver failure, modify anesthetic management.
- Pulm AVMs could be large enough to lead to heart failure and polycythemia.
- Key aspects of anesthetic management are interventions to maintain nml hemodynamic parameters and to prevent bleeding and the formation of emboli.

Postoperative Period

- Avoid immobilization for prolonged periods of time to avoid thromboembolism to CNS.

Adjuvants

- Watch for incompatible drugs in IVs or peripheral veins to avoid particulate matter precipitation and embolization to the brain.
- Broad-spectrum antibiotic prophylaxis to decrease risk of CNS infections.
- NSAIDs may precipitate GI or mucosal bleeding and impair renal function.

Anticipated Problems/Concerns

- Anemia due to recurrent bleeding; most commonly epistaxis.
- Transfusion is complicated: Low Hct may increase the risk of high-output CHF by increasing extent of arteriovenous shunting (decreasing viscosity effect), but a high Hct may increase risk of thromboembolism.
- Coagulopathy: Multiple hemostatic defects, including low-grade DIC, reduced plt aggregation, and factor XI deficiency, may aggravate bleeding caused by local vessel wall pathology.
- Paradoxical embolism: Owing to pulm AVMs, peripheral microemboli (air, bland, or septic) bypass nml pulm capillary filtering and embolize, causing transient or permanent neurologic defects or brain abscess.
- Special attention should be paid to pregnant women with the diagnosis of HHT. In the rare instances, deterioration of preconception AVMs and the development of new AVMs will present with clinically silent but potentially life threatening complications of the disorder. These are most commonly located in the pulm vasculature, followed in frequency by the cerebral, GI, and spinal circulation. With CV and hormonally induced enlargement of certain AVMs, there is concurrent risk of rupture, as well as shunt-induced high cardiac output failure and systemic embolism.

Herniated Nucleus Pulposus

Christine Peeters-Asdourian | Efrain I. Cubillo

Risk

- Incidence of symptomatic disc herniation is 1–2% in the general population.
- Most common age of presentation is during third and fourth decades of life.
- Smoking leads to reduced O_2 tension secondary to vasoconstriction, significant inhibition of cell proliferation and extracellular matrix synthesis, and increased abnormal type I collagen versus type II collagen overall, leading to disc degeneration.
- Chronic stress (e.g., chronic coughing, sitting without lumbar support, heavy lifting) increases strain on disc.
- Poor posture combined with poor body mechanics places stress on the lumbar spine and affects the distribution of body weight.
- Obesity and largely sedentary lifestyle.

Overview

- Structurally the lumbar disc has three components: the annulus fibrosus, forming the circumferential rim of the disc; the nucleus pulposus, composing its central core; and the cartilaginous end plates on the adjacent vertebral bodies.
- The intervertebral disc is the largest avascular structure in the body.
- The nucleus pulposus is composed of H_2O, collagen, and PGs. PG molecules are important because they attract and retain H_2O, constituting a hydrated gel-like matter that resists compression. The amount of H_2O in the nucleus varies throughout the day, depending on activity. It decreases with age, leading to degenerative disc disease.
- The annulus fibrosus is an annular structure composed of concentric sheets of collagen fibers connected to the vertebral end plates. The sheets are oriented at various angles and enclose the nucleus pulposus.
- Disc herniation occurs when the annulus fibrosus breaks open or cracks, allowing the nucleus pulposus to escape. This is called a HNP or herniated disc. Escaping material initiates an inflammatory reaction.
- Disc herniation typically gives rise to radicular pain, which is pain in the distribution of the nerve root affected by the herniation. This pain has strong inflammatory and neuropathic components with or without neurologic change. If radicular changes take place, the presentation is that of a radiculopathy.
- Lumbar region L4–L5 is most common site (59%), followed by L5–S1 (30%) and L3–L4 (9%).
- Natural history of disease is favorable.
- Most pts have substantial improvement of symptoms within a few mo.

Etiology

- Ability of the nucleus pulposus to retain H_2O declines progressively with age.
- Displacement of nuclear material initiates a robust inflammatory response, eliciting known inflammatory mediators such as IL-1, IL-8, IL-17 and TNF α in addition to several recently identified contributing mediators including NGF, IFNγ, and Th1 lymphocyte activation.
- The inflammatory response results in migration and activation of macrophages, leading to scar production and an increase in substance P.
- Symptoms do not always correlate with herniation size (asymptomatic herniation is frequent).

Disease Presentation

- May begin either suddenly, with physical activity, or slowly.
- Frequently presents with a combination of back pain and radicular symptoms; neurologic signs such as weakness or sensory deficits are possible. Isolated low back pain may also be the sole presentation.
- Pts often describe a popping sensation prior to onset of radicular symptom.
- Neural impingement is responsible for dysfunction. Compression of a motor nerve results in weakness in less than 50% of pts; compression of a sensory nerve results in numbness.
- Radicular pain is caused by inflammation of the nerve (which can explain the lack of correlation between the size of a herniation and symptoms of pain).
- Ideal imaging modality is MRI, although CT may also be helpful, EMG/NCS can help to identify the nerve root involved. However, there is not always a correlation between findings on imaging studies and clinical presentation.
- Maneuvers that increase intrathecal pressure (coughing, sneezing, prolonged sitting) can aggravate pain.

Usual Treatment

- Conservative therapies:
  - NSAIDs are supported by the literature.
  - Systemic corticosteroids have not been shown to be superior to placebo.
  - Opioids, muscle relaxants, neuropathic agents (empirical data, limited EBM data)
  - Contrary to prior belief, activity is now preferred over bed rest.
  - Physical therapy.
  - Several other modalities, poorly supported by the literature, include bracing, traction, acupuncture, chiropractic manipulation, behavioral therapy, and biofeedback.
  - Favorable outcomes are more common among better-educated pts and those who are self-motivated. A second neurologic examination within 12 wk is suggested. nml psychological profile, and absence of a workers compensation claim or litigation
- Injection therapy:
  - Epidural injections utilizing fluoroscopy are the standard of care as fluoroscopy is one modality that may reduce catastrophic neurologic injuries, including stroke and spinal cord injury
  - Interlaminar epidural steroid injections are commonly performed.
  - Transforaminal epidural steroid injections target a given area more precisely (commonly performed in the lumbar region but controversial in the cervical region).
  - Investigational studies have shown evidence for lumbar intradiscal PRP.
- Percutaneous discectomy:
  - Does not directly remove the herniated portion but rather removes only the nucleus pulposus in the hope that the herniation portion will regress (limited studies showed a success rate of about 30%).
  - Nucleoplasty.
  - Laser disc decompression.
  - Endoscopic discectomy.