- Pts may display greater sensitivity to vasopressors; systemic and neuraxial administration of vasopressors should be used with caution.

General Anesthesia for Cesarean Delivery

- Neuraxial anesthesia preferable to general anesthesia for cesarean delivery.
- Potential for difficult intubation secondary to airway edema.
- Htn accompanying laryngoscopy increases the risk of cerebral hemorrhage and pulm edema.
- Induction with propofol increases seizure threshold and reduce $CMRO_2$ and CBF.
- Magnesium sulfate increases the potency and duration of depolarizing and nondepolarizing muscle relaxants.
- Avoid hypercarbia, which lowers seizure threshold.
- Maintain CPP (MAP – ICP) and avoid hypoxia, hyperthermia, and hyperglycemia to prevent further neurologic injury.

Postoperative Period

- Continue magnesium sulfate infusion for 24 h after delivery and/or last seizure.
- Increased risk of pulm edema as extracellular fluid is mobilized leading to increased intravascular volume.
- Most eclamptic pts have complete resolution of neurologic abnormalities.

Anticipated Problems/Concerns

- 10% will have recurrent seizures in the absence of prophylaxis with initial seizure.
- Eclamptic seizures can occur up to 4 wk postpartum.
- Cerebral hemorrhage accounts for 15–20% of deaths from eclampsia.

Ehlers-Danlos Syndrome

Christopher J. Cullom | Alan David Kaye

Risk

- EDS has an overall incidence of 1:10,000-25,000, with no ethnic predisposition.
- Six major subtypes, each with slightly different and unique phenotypes.
- Symptoms involve skin, ligaments, joints, and vessels.

Perioperative Risks

- Valvular abnormalities or major vessel dissection/aneurysm
- Unstoppable bleeding
- Pneumothorax from positive pressure ventilation or pneumoperitoneum
- Neuropathy or musculoskeletal injury from positioning
- Airway difficulty from atlanto-occipital instability

Worry About

- Musculoskeletal injury from positioning.
- Airway damage due to repeat intubations.
- TMJ luxation from intubation or mask ventilation.
- Postural orthostatic tachycardia syndrome possible in EDS; thus preop crystalloid and early use of vasopressors recommended.
- Initiate preop crossmatching of RBCs and use of cell-saver for major surgery. DDAVP improves bleeding time and transfusion requirement.
- Use ultrasound when performing central lines or arterials lines to avoid vessel dissection.
- Generally avoid neuraxial blockade due to risk of bleeding.

Overview

- EDS I, EDS II, and hypermobile type (EDS III) is found in 90% of cases.
- Vascular type (EDS IV) is found in 3–10% cases.
- Kyphoscoliotic (EDS VI), arthrochalasis (EDS VIIA/B), and dermatosparaxis (EDS VIIC) types are rare cases. Principal clinical features include tissue fragility, easy bruising, skin hyperextensibility, delayed wound healing, joint hypermobility, and atrophic scarring.
- Initial manifestation is usually easy bruising. Bleeding from gums after brushing or bleeding after minor trauma is common.
- Platelet count or bleeding time is normal, yet a Rumpel-Leede test may be positive.
- Cardiac manifestations include arterial aneurysms, arterial rupture, varicose veins, aortic regurgitation, mitral valve prolapse, or conduction disturbances.
- Other important manifestations include pneumothorax, diverticula of intestine, megaesophagus, or megacolon.
- EDS types I and II notably present with very soft, fragile skin.
- Frequent joint dislocations happen at shoulder, hip, and patella, typically with EDS III.
- EDS IV has the most severe presentation and only forms with increased risk of death due to cardiac pathology.
- EDS VI is recognized by kyphoscoliosis, muscle hypotonia, and joint hypermobility.
- Arthrochalasia type (EDS VII A/B) presents with joint hypermobility and congenital bilateral hip dislocation.
- Dermatosparaxis type (EDS VII C) presents with severe bruising, extreme skin fragility, large fontanels, and short stature.

Etiology

- Mutation in gene encoding for fibrillar collagen proteins or enzymes can be involved in modifications of these proteins.
- Type I collagen is the predominant type in body. Mutation in type I results in EDS VIIA/B. Mutation in type V collagen that is coexpressed with type I results in EDS I/II.

Usual Treatment

- No treatment for EDS; however, there are preventative guidelines.
- Protective pads for pressure points during positioning.
- Avoid antiplatelet drugs and use DDAVP for pts at high risk of bleeding.
- Baseline ECHO and ECG, especially for EDS type IV.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Mitral valve prolapse, aortic regurgitation, arterial aneurysm/rupture, conduction abn	Dizziness Palpitations Chest pain	Orthostatic hypotension Arrhythmia	ECG, ECHO
RESP	Pneumothorax, airway difficulty		SOB, wheezing	CXR
GI	Intestinal diverticula, megacolon			Abdominal x-ray
HEME	Bleeding propensity			CBC, Rumpel-Leede test
MS	Skin fragility, joint dislocations, TMJ dislocation		Skin fragility testing	X-ray of extremities
NEURO	Atlanto-occipital instability		Cervical spine eval	
HEENT	Retinal detachment		Visual acuity, ophthalmoscopic exam	

Key References: Weismann T, Castori M, Malfait F, et al.: Recommendations for anesthesia and perioperative management in patients with Ehlers-Danlos syndrome, *Orphanet J Rare Dis* 9(109):1–9, 2014; Johnston B, Occhipinti K, Baluch A, et al.: Ehlers-Danlos syndrome: complications and solutions concerning anesthetic management, *Middle East J Anaesthesiol* 18(6):1171–1183, 2006.

Perioperative Implications

Preoperative Preparation

- Genetic counseling and identification of EDS type
- Standardized bleeding history
- History of intubation difficulty
- Echo to exclude cardiac pathology
- Type and crossmatch of RBCs as well as avoidance of antiplatelet drugs

Monitoring

- Avoid invasive monitoring whenever possible due to bleeding risk.

Airway

- Atlanto-occipital instability creates airway difficulties.
- Use fiberoptic intubation to avoid multiple intubation attempts in the setting of fragile mucosa.

Preinduction/Induction

- Use of padding during position and appropriate tape to avoid shear forces and external tissue pressure
- Eye protection due to risk of retinal detachment

Maintenance

- No anesthetic or pharmacotherapy contraindications.
- BP control due to fragile vessels, particularly if there are undiagnosed aneurysms.
- Monitor for pneumothorax due to barotraumas.

Extubation
- Usual criteria

Postoperative Period
- Careful positioning and use of tape in PACU
- Early mobilization to avoid loss of strength
- Postop N/V prophylaxis to avoid esophageal tears

Regional Anesthesia
- Generally avoid due to potential risks.

Anticipated Problems/Concerns
- Cardiac pathology
- Hemodynamic instability and hypotension
- Airway difficulty
- Bleeding propensity
- Nerve and soft tissue injury due to positioning
- Tissue or vessel fragility
- Pneumothorax

Eisenmenger Syndrome

Inna Maranets

Risk
- 8% of all CHD pts.
- 11% of pts with intracardiac or aortopulmonary shunt, allowing continuous exposure of pulm vasculature to systemic arterial pressure.
- VSD is the most common lesion.

Perioperative Risks
- High risk of cardiovascular complications when undergoing noncardiac surgery; mortality reaching 30%.
- Severity of pulm Htn cyanosis, tricuspid regurgitation, and right ventricular dysfunction are important factors.
- Additional acquired cardiac and systemic diseases, such as CAD and renal dysfunction.
- Underlying pathology, urgency, duration of surgery, and anesthetic choice contribute to the risk.
- Bleeding due to platelet dysfunction.
- Mortality rate of pts with ES carrying pregnancy to viability is 27–30%, most often at delivery or postpartum.
- Fetal risks: Increased risk of preterm labor and intrauterine growth retardation; fetal demise of 75%.
- Cesarean section carries higher mortality: 70% versus 30% for vaginal delivery.

Worry About
- R-to-L shunt, pulm Htn, RV and LV ventricular failure, hypoxemia, polycythemia.
- Minor decrease in SBP can cause increase in R-to-L shunt, decreased pulm blood flow, hypoxia, and cardiovascular collapse.
- Increased blood viscosity can lead to thromboembolic phenomena, paradoxical emboli, hemoptysis.
- Arrhythmias, ventricular and supraventricular.
- May not tolerate positive pressure ventilation.
- Decreased systemic vascular resistance of pregnancy worsens R-to-L shunt.
- Inability to meet increased demand for O_2 with gestation and labor.
- Delivery produces autotransfusion with RV failure.
- Excessive bleeding with previous heparinization.
- Postpartum increase in PVR.

Overview
- ES is defined as pulm Htn at systemic level due to high PVR with reversed or bidirectional shunt through communication between the two circulations.
- Communication may be at aortic level (PDA, aortopulmonary window), intracardiac (ASD, VSD, AV canal, TAPVR) or single ventricle.
- Uncorrected L-to-R shunt leads to irreversible fixed pulm vascular obstructive disease.
- Characterized by pulm Htn, R-to-L shunt, and RV dysfunction.
- Overall poor prognosis; mean age at death: 25 y.
- Syncope, increased right-sided filling pressures, and systemic arterial desaturation below 85% indicate poor prognosis.
- 50% of pregnant pts die in association with pregnancy.
- Some pulm vascular reactivity may exist in the pulm vasculature of pregnant women; may be due to systemic hormonal changes of pregnancy.

Etiology
- Individuals with large unrestricted intracardiac or aortopulmonary communication have large L (systemic)-to-R (pulm) shunts.
- Uncorrected L-to-R shunt overloads pulm vasculature and RV.
- Continuous exposure to systemic pressure leads to pulm arteriolar medial hypertrophy, intimal proliferation, and fibrosis.
- Progressive pulm capillary and arteriolar occlusion leads to fixed increased PVR.
- As pulm pressure exceeds systemic, shunt reverses to R to L.

Usual Treatment
- Repair of intracardiac lesion is contraindicated.
- Supplemental oxygen to decrease PVR.
- Avoidance of medications that can cause hypotension, worsening cyanosis or hemorrhage (calcium channel blockers, antiplatelet agents, anticoagulants).
- Phlebotomy to treat hyperviscosity, extreme erythrocytosis (Hc >65%), and bleeding diathesis.
- Single or bilateral lung transplantation with repair of the primary cardiac defect.
- Combined heart-lung transplant in select pts.
- With expected high maternal mortality, pregnant pts with ES should initially be counseled to terminate pregnancy.
- For the pt who wishes to continue with pregnancy:
  - Hospital admission early in third trimester.
  - Anticoagulation with heparin: SQ heparin 5000–10,000 U bid.
  - Pts with O_2 sat <80% on room air should be fully anticoagulated.
  - O_2 Rx.
  - Monitor for preterm labor
  - Medical Rx: Diuretics, antiarrhythmics, inotropes

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	R-to-L shunt Right and left ventricular enlargement/failure	DOE, fatigue, syncope edema, orthopnea, anginal chest pain, arrhythmias	Elevated jugular venous pressure, increased intensity of S_2, split S_2 and S_3; decrescendo murmur of pulmonic regurgitation, holosystolic murmur of tricuspid regurgitation; rales; right parasternal heave	ECG CXR ECHO MRI Cardiac cath
RESP	Pulm Htn	Dyspnea, hemoptysis	Palpable pulm artery Cyanosis, clubbing	Pulse oximetry ABG, Hct (polycythemia)
NEURO	Neurologic abnormalities	Headache, dizziness, visual disturbances, CVAs	Neuro exam	CT scan, MRI
HEME	Polycythemia, Hyperviscosity	Headache, weakness, blurred vision, pruritus	Splenomegaly, facial erythema, bleeding gums	CBC

Key References: Ammash NM, Connolly HM, Abel MD, et al.: Noncardiac surgery in Eisenmenger syndrome, *J Am Coll Cardiol* 33(1):222–227, 1999; Bennett JM, Ehrenfeld JM, Markham L, et al.: Anesthetic management and outcomes for pts with pulmonary hypertension and intracardiac shunts and Eisenmenger syndrome: a review of institutional experience, *J Clin Anesth* 26(4):286–293, 2014.

Perioperative Implications

Preoperative Preparation
- Continue antiarrhythmic medications and withhold diuretics.
- Discontinuation of heparin; consider reversal with protamine.
- Endocarditis prophylaxis depends on the type of operation (AHA Guidelines).
- In pregnant pts avoid aortocaval compression at all times.
- IV lines must be carefully de-aired, consider placing air filters.

Monitoring
- Pulse oximetry.
- With uncorrected patent ductus arteriosus, use simultaneous right hand (preductal) and foot (postductal) pulse oximetry to estimate changes in shunt fraction.
- Arterial line for early recognition of sudden alteration of BP and repeated blood gas sampling.
- CVP line.
- PA cath use must be balanced against potential complications:
  - Difficult to position in PA.
  - High risk of arrhythmias, thrombi, paradoxical emboli, and PA hemorrhage.
  - Misleading data: Unreliable PCWP and measurement of CO with shunt.

Airway
- Preop administration of Bicitra, metoclopramide, and ranitidine if needed
- NPO for 8 h (if possible)