- If gas embolism suspected, aspirate central line; if none, consider subcostal insertion of spinal needle attached to large aspirating syringe directly into RV.

Extubation

- Base on usual criteria, extent of operative procedure, pt's age and physical condition, and concurrent disease

Postoperative Period

- Base pain control plan on nature and extent of resection; regional anesthesia an option if coagulation status permits.
- Base monitoring on extent of resection, blood loss, and preop health status.
- Watch for pneumothorax, subphrenic abscess, pneumonia, bronchobiliary fistula, jaundice, hepatic failure, and septicemia.

Anticipated Problems/Concerns

- If the pt is being treated in a nonendemic area, surgical team may be unfamiliar with disease; anthelmintic medications may require special order well in advance of procedure.
- Consider *Echinococcus* in any pt from endemic area presenting for surgical excision of cyst; search for others using US imaging; consider ID consult and serologic testing.
- Cysts may eventually involute and degenerate, hence the conservative nature of treatment in nonemergent cysts.
- Arrange ultrasound imaging in family/neighbors/farm animals capable of being intermediate hosts.
- Examine stool of companion canids for eggs and segments.

Eclampsia

Emily Baird

Risk

- Incidence varies from 0.01–0.1% of pregnancies in developed countries.
- Occurs in 1–3% of pts with preeclampsia.
- Risk factors include age <20 y old, nulliparity, anemia, diabetes, and preexisting heart disease.

Perioperative Risks

- Eclampsia is a factor in approximately 10% of all maternal deaths in developed countries.
- Maternal complications include adult respiratory distress syndrome, acute renal failure, cardiopulmonary arrest, and CVA.
- Fetal complications include respiratory distress syndrome, small for gestational age, preterm birth, and intrauterine growth restriction.

Worry About

- Risk of pulm aspiration and hypoxemia with seizure
- Fetal bradycardia may occur during or following seizure
- 90% of women with eclampsia have manifestations of severe preeclampsia (Htn, proteinuria, renal insufficiency, pulmonary edema, coagulopathy)

Overview

- New onset of generalized, tonic-clonic seizures, and/or unexplained coma during the peripartum period in a woman without a preexisting neurologic disorder.
- Eclamptic seizures can occur during the antepartum (60%), intrapartum (20%), or postpartum (20%) period.
- Onset of eclampsia is generally preceded by signs of severe preeclampsia but approximately 10% occur without Htn.

Etiology

- Precise etiology is unknown, but two models have been proposed based on the central role of Htn in the majority of eclampsia cases.
  - Forced dilation theory: Htn exceeds the upper limit of cerebral autoregulation leading to hyperperfusion, endothelial dysfunction, and interstitial edema.
  - Vasospasm theory: Htn causes overactivation of cerebral autoregulation leading to vasoconstriction, hypoperfusion, localized ischemia, and cerebral edema.

Usual Treatment

- Establish patent airway and maintain maternal oxygenation.
- Maintain left uterine displacement.
- Seizure treatment/prophylaxis: Magnesium sulfate (4–6 g bolus over 20 min followed by 1–2 g/h infusion ± 2 g bolus over 10 min for recurrent seizure).
- Antihypertensive treatment for SBP ≥160 mm Hg and/or DBP ≥110 mm Hg: Labetalol (10–20 mg IV) and/or hydralazine (5–10 mg IV).
- Expeditious delivery via induction/augmentation of labor (preferred) or cesarean delivery (if persistent maternal or fetal distress).

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Htn Reduced intravascular volume LV dysfunction (rare)	Dyspnea, peripheral edema	Htn, peripheral edema, decreased CVP	ECHO if suspect LV dysfunction
RESP	Airway edema Pulm edema	Snoring, stridor dyspnea, orthopnea	Tachypnea, dyspnea, hypoxemia, rales	CXR ABG
RENAL	Proteinuria Renal failure Decreased RBF Decreased GFR	Rapid weight gain, decreased urine output	Nondependent edema	24-h urine protein, BUN, Cr, uric acid
HEME	Thrombocytopenia Microangiopathic hemolysis DIC	Mucosal bleeding, easy bruising	Petechiae, bleeding from puncture sites	Hgb, Hct, plt, fibrinogen, and FSP
NEURO	Seizure Coma	Headache, visual disturbances	Hyperexcitability, hyperreflexia	CT/ MRI if focal deficits or prolonged coma
FETUS	Fetal distress IUGR Oligohydramnios			Fetal heart monitor Fetal ultrasound

Key References: Leffert LR: What's new in obstetric anesthesia? Focus on preeclampsia, *Int J Obstet Anesth* 24(3):264–271, 2015; Parthasarathy S, Kumar VR, Sripriya R, et al.: Anesthetic management of a patient presenting with eclampsia, *Anesth Essays Res* 7(3):307–312, 2013.

Perioperative Implications

Monitoring

- Standard maternal monitors including noninvasive BP, pulse oximetry, and UO.
- Indications for invasive BP monitoring: (1) BP poorly controlled; (2) frequent blood sampling; or (3) infusion of potent vasodilators (nitroprusside or nitroglycerin).
- Indications for invasive central venous monitoring: (1) infusion of potent vasoactive agents; (2) pulmonary edema; and (3) cardiomyopathy.
- Electronic fetal heart monitoring.

Regional Anesthesia for Labor and Delivery

- Benefits of an early epidural: (1) high-quality analgesia (attenuates hypertensive response to pain); (2) improvement in uteroplacental circulation; and (3) avoidance of general anesthesia if emergency cesarean delivery indicated.
- Assessment of coagulation status, as outlined previously, should be checked prior to both placement *and* removal of epidural cath.
- Avoid IV fluid boluses prior to neuraxial anesthesia because of the increased risk of pulm edema.

- 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.