Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	CHF	FTT, difficulty feeding	"Machinery" murmur Wide pulse pressure Bounding pulses Pulsus bisferiens	ECHO
	Pulm Htn		Tachycardia Diaphoresis	
RESP	Pulm edema	Recurrent respiratory infections Increased O_2 requirement	Worsening mechanical ventilation parameters Rales	CXR
GI	NEC	Abdominal distention Poor feeding Blood in stool Free air in peritoneum	Distended tense abdomen Edema of abdominal wall Tender abdomen	Abdominal x-ray
RENAL	Oliguria	Decreased UO due to decreased renal blood flow		Serum chemistry
CNS	CNS hemorrhage	Increased fontanel pressure Decreased Hct	Increased fontanel size and tension	Head US

Key Reference: Jacobs JP, Giroud JM: Evolution of strategies for management of the patent arterial duct, *Cardiol Young* 17(Suppl 2):68–74, 2007.

Perioperative Implications

Preoperative Preparation

- Surgery:
  - Unstable neonate, bedside: Cross-matched blood at bedside, adequate IV access with extension tubing. Caretakers must be familiar with ventilator function and settings and must check current running infusions (TPN, vasopressors).
  - Stable child: Cross-matched blood in the OR.
- Cardiac cath lab: Routine setup for general ET anesthesia.

Preinduction/Induction

- Unstable neonate: Induce with fentanyl (10–30 mcg/kg)
- Stable child in cath lab/OR: Premedication and mask induction

Monitoring

- Standard ASA monitors.
- Unstable neonates require an arterial line for continuous BP measurement and blood gas analysis and central venous access for inotrope drug delivery.
- Stable older children do not require invasive monitoring.

Airway

- Critically ill neonates are already intubated and ventilated. Check tube size for leak and confirm position on CXR.
- OR cases: Intubate for single-lung ventilation (right main stem, a single-lumen ETT, bronchial blocker, or double-lumen tube).
- Cath lab cases: Young children often require intubation; older cooperative children may be treated with a natural airway.

Maintenance and Extubation

- Critically ill neonate, bedside: Fentanyl and paralytics; patient should remain intubated at the end of the procedure.
- Stable child, OR: Balanced anesthetic technique with the goal of early extubation and adequate analgesia (consider regional techniques).
- Stable child, cath lab: Balanced anesthetic technique and extubate at the end of the procedure. Analgesic requirements are minimal and related to the femoral vessel puncture sites.

Adjuvants

- Antibiotic prophylaxis for all cases (usually cefazolin 30–50 mg/kg)

Postoperative Period

- Adequate analgesia

Anticipated Problems/Concerns

- Critically ill neonates: Often require a transient increase in BP and respiratory support.
- Stable children: Postop surgical ligation via a thoracotomy; such patients may have atelectasis from single-lung ventilation and also thoracotomy pain.

Patent Foramen Ovale

Ronit Lavi | Daniel Bainbridge

Risk

- Incidence: 25–30% at autopsy.
- Atrial septal aneurysm (a deformity of the septum that results in deviation of the septum more than 15 mm into either atrium) is associated with at least 50% of PFOs and is considered an additional risk factor for stroke.

Perioperative Risk

- Unclear if a PFO increases the risk of stroke or cognitive dysfunction in the periop period.

Worry About

- R-to-L shunting of blood leading to profound hypoxemia
- Paradoxical embolization of air, blood clot, or tissue fragments, potentially resulting in stroke

Overview

- The foramen ovale directs oxygenated blood returning from the placenta and into the right atrium across the intra-atrial septum to the left ventricle.
- As right-sided pressures decrease after birth, the foramen ovale flap is pressed against the septum secundum.
- This results in the fusion of the ovale flap to the septum secundum; irreversible closure of the ovale occurs in 75% of pts.
- Diagnosed by:
  - Right heart cath, with the ability to cross a guide wire across the atrial septum.
  - TEE is considered the "gold standard" imaging technique, using a contrast agent (bubble study) and provocative technique (Valsalva maneuver).
  - TCD is less invasive than TEE with similar sensitivity but reduced specificity.
  - TTE: Sensitivity 50% of TEE, with similar specificity.
- See also Atrial Septal Defect.

Etiology

- Unknown what, if any, risk factors predispose to patent foramen ovale
- A higher incidence of PFO was found in pts who suffer migraine with aura; unclear whether this represents coexistence or a causal relationship between the two entities.

Usual Treatment

- Anticoagulation has not been shown to reduce cryptogenic stroke.
- Percutaneous closure for pts with history of cryptogenic stroke.
- Surgical closure in pts who are not candidates for percutaneous closure.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Rarely RV overload secondary to chronic L-to-R shunting	Fatigue, abdominal pain	Hepatic enlargement, elevated JVP, peripheral edema	ECG showing right axis deviation ECHO showing large RV PFO
CNS	Assoc with cryptogenic stroke, migraine	Hx of migraine headache, stroke unrelated to carotid Dx, AFIB		

Key References: Sukernik MR, Mets B, Bennett-Guerrero E: Patent foramen ovale and its significance in the perioperative period, *Anesth Analg* 93(5):1137–1146, 2001; Kutty S, Sengupta PP, Khandheria BK: Patent foramen ovale: the known and the to be known, *J Am Coll Cardiol* 59(19):1665–1671, 2012.

Perioperative Implications

Preoperative Preparation
- Deairing of all IV lines/syringes.
- No indication for antibiotic prophylaxis.
- Use regional technique when possible as a sole anesthetic or in combination with GA for pain control.

Monitoring
- TEE and/or TCD to check for PFO has been advocated, especially for pts undergoing surgery in the sitting position or when the surgical field is above the level of the right atrium.

Airway
- Careful airway assessment, optimal pt positioning, and all necessary airway management tools available

Induction
- Proper preoxygenation avoid hypoxemia, hypercarbia, and acidemia
- Five-lead ECG and low threshold for arterial line cath to monitor BP, metabolic balance, and oxygenation throughout the case and in PACU.
- Avoid systemic hypotension/pulm Htn, as this increases the potential for R-to-L shunting.
- If appropriate, use an induction regimen that maintains or decreases PVR and maintains systemic vascular resistance, sinus rhythm, and contractility.

Maintenance
- Avoid high peak airway pressures during PPV as rapid changes in pressure may predispose to R-to-L shunting.
- If hypoxemia worsens with PEEP, suspect shunt.
- Laparoscopic procedures with increased intra-abdominal pressure might not be tolerated.

Extubation
- Hypercarbia and/or hypoxia may predispose to R-to-L shunting. Extubate when pt is fully awake and obeys commands.
- Smooth emergence is indicated to prevent shunting.

Postoperative Period
- Pulm embolus in the postop period may present as severe hypoxemia with preserved systolic blood pressure owing to an increase in R-to-L shunting.
- Adequate pain control with a multimodal approach and/or regional anesthesia as indicated.

Anticipated Problems/Concerns
- Intraoperative and postop hypoxemia.
- Potential increased risk for periop TIA/stroke.
- Positive-pressure ventilation and laparoscopic procedures with increased intra-abdominal pressure; neurosurgery, orthopedic surgery, thoracic surgery, and cardiac procedures might precipitate R-to-L shunt with hypoxemia.
- Atrial arrhythmia should be managed promptly to decrease the chance of shunt and embolism.
- Excessive pain might increase the chances of developing shunt; an adequate pain control regimen should be in place.
- All lines should be de-aired and special air-trapping filters used.
- Insertion and removal of a central line impose a higher risk for air embolism.

Pemphigus

Megan K. Werntz | Brandon M. Togioka

Risk
- Incidence in USA: 0.1–0.5:100,000 per y for pemphigus vulgaris (the most common form of pemphigus)
- Individuals from ages 40-60 y most commonly affected.

Perioperative Risks
- Infection, sepsis
- Electrolyte abnormalities and dehydration, with extensive lesions

Worry About
- Volume status with oropharyngeal lesions and decreased oral intake
- Skin and/or pharyngeal blisters (lesions may be limited to the oropharynx), sloughing of mucosa, bleeding produced by airway manipulation
- Consequences of steroid treatment (e.g., hypertension, hyperglycemia, gastric or duodenal ulceration, myopathy, infection, psychic disturbances, osteoporosis) or immunosuppressive therapy (bone marrow suppression, susceptibility to opportunistic infections and cancer)

Overview
- An autoimmune intraepidermal blistering disease of the skin and mucous membranes. Oral lesions are most common. Blisters rupture easily but heal slowly.
- There are four types: Pemphigus vulgaris (most common and severe form), pemphigus pemphigus foliaceus, IgA pemphigus, and paraneoplastic pemphigus.
- The 5-y mortality is 5–15% for treated pemphigus vulgaris. The most common cause of death is infection, usually with *Staphylococcus aureus*.
- Occasionally pemphigus can coexist with other autoimmune diseases, a thymoma (with or without myasthenia gravis), or malignancies.

Etiology
- Autoimmune disease, in which autoantibodies are produced against cell adhesion molecules (desmosomal glycoproteins) on keratinocytes. Immune response leads to separation of epidermal keratinocytes and blistering.
- Uncommonly, pemphigus is drug-induced, associated with malignancy, or related to infection.

Usual Treatment
- Corticosteroids (most common therapy).
- Azathioprine and mycophenolate are the most commonly used immunomodulatory agents.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Oral and pharyngeal erosions and blisters	Painful swallowing Decreased oral intake	Oral lesions	Albumin Na^+
ENDO	Hyperglycemia (due to steroids)			Glucose
CV	Htn (due to steroids)		BP	
RESP	At risk for pneumonia	Fever, cough, sputum	Diminished breath sounds, dullness to percussion	CXR
GI	Gastric or duodenal ulcer (due to steroids)	Epigastric pain	Dark stool	Guaiac fecal occult blood test
MS	Myopathy (due to steroids or association with myasthenia gravis)	Fatigability, weakness		
DERM	Skin and mucous membrane blisters	Painful skin lesions	Blisters/denuded skin	BMP

Key References: Mahalingam TG, Kathirvel S, Sodhi P: Anaesthetic management of a patient with pemphigus vulgaris for emergency laparotomy, *Anaesthesia* 55(2):160–162, 2000; Bansal A, Tewari A, Garg S, et al.: Anesthetic considerations in pemphigus vulgaris: case series and review of literature, *Saudi J Anaesth* 6(2):165–168, 2012.

Perioperative Implications

Preoperative Preparation
- Pts may require supplemental steroids.
- Secure IV lines with cloth bandage or suture (avoid tape) and place on lesion-free areas.

Monitoring
- Ensure careful placement of monitors and extra padding under pressure points and BP cuff.

Airway
- Airway management may become more difficult if tissue manipulation leads to bleeding.
- Consider lubricating mask and laryngoscope blade (Macintosh may be less traumatic than Miller blade) to decrease friction (potentially with hydrocortisone cream or ointment).
- Consider small ETT and inflate cuff only minimally; suture ETT or use tube holder instead of taping.
- Consider avoiding LMA owing to risk of pharyngeal trauma.
- Consider use of video laryngoscope for assessment of bullae during intubation.

Preinduction/Induction
- Lubricate eyes; consider goggles instead of tape.
- Use gentle bag mask ventilation.

Maintenance
- Regional anesthesia preferred (when appropriate) to avoid airway manipulation.
- Consider single-shot blocks (spinals or peripheral blocks) to avoid tape.
- Local infiltration is probably contraindicated owing to risk of blister formation.

Extubation
- Minimize coughing during extubation and provide gentle oropharyngeal suctioning.

Postoperative Period
- Gentle pt repositioning.
- Treat pruritus aggressively and avoid pain management that can cause itching.

Adjuvants
- Consider need for steroid supplementation.

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