

**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

**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 <sup>+</sup> Glucose
ENDO	Hyperglycemia (due to steroids)			
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	Guaic 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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