- Pulmonary rehabilitation should include inspiratory muscle training and pt education, supplemental oxygen, and vaccination for seasonal influenza and pneumococcus.
- No current FDA-approved therapy has been shown to be efficacious in IPF, and consequently management includes primarily supportive care, as described previously.
- Some clinical benefit is described with medications pirfenidone and nintedanib in terms of potentially slowing disease progression.
- Lung transplantation is a possible consideration, but mainstay of therapy more commonly involves aggressive management of associated comorbidities.
- A multitude of pharmacologic trials, including a broad spectrum of medications (cytotoxic agents, antifibrotic agents, anti-inflammatory and immunosuppressive agents, and PDEinh), have unfortunately not proven beneficial and in fact were often associated with intolerable toxicities.
- Pts may present on these medications, and therefore it is important to have an awareness of them and review them preop.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	RV strain and failure, pulm Htn and cor pulmonale	Easy fatigue and swelling peripherally	Edema in legs and feet and hepatic congestion	ECG, ECHO, and consider RV heart cath
RESP	Diminished pulm capacity and function	Progressive shortness of breath and diminished functional capacity Dry persistent cough	Inspiratory crackles, clubbing of digits; rapid and shallow breathing; absence of signs of acute/chronic infection	PFTs (restrictive physiology with low lung volumes and decreased DLCO); CXR (diffuse patchy fibrosis and pleural based honeycombing), 6-min walk test, oximetry, hypoxemia
GENERAL	Metabolic wasting and weight loss, malnourishment	Diminished exercise and functional capacity	Progressive weight loss	Albumin
RENAL	Renal insufficiency			Cr, Cr clearance

Key Reference: King TE Jr, Pardo A, Selman M: Idiopathic pulmonary fibrosis. *Lancet* 378 (9807):1949–1961, 2011.

Perioperative Implications

Preoperative Preparation

- Review systems. It is important to thoroughly investigate through questioning and appropriate testing for associated comorbidities that may be optimized to improve potential for periop success. One must be vigilant to address all coexisting disease to the extent possible.
- Assess pulm physiologic reserve to guide periop risk discussion.
- Consider common misdiagnoses that may be treatable preop if surgery is elective (e.g., bronchitis/pneumonia, bronchospasm, PE, exposures).
- Evaluate progression of disease as assessed by longitudinal measurements: FVC, TLC, DLCO, oximetry, 6-min walk test.
- Risk is further increased for periop pulm insufficiency if:
  - Obesity BMI >27 kg/m^2
  - Smoking within 8 wk of surgery
  - Productive cough or wheezing within 5 d of surgery
  - FEV_1/FVC ratio <70% and $PaCO_2$ >45 mm Hg
- Consider need for ICU and invasive hemodynamic monitoring/management periop.
- Aggressive management of COPD, smoking cessation, steroids, and antibiotics as indicated.

Monitoring

- Routine ASA monitors plus consideration of invasive hemodynamic monitors and/or TEE as dictated by pt status, pulm Htn, RV and LV functional status, and case type/duration.
- Consider possible need for postop ventilation in the ICU and ABG monitoring for weaning from ventilator support.
- Type and duration of surgery will affect the rate of postop pulm insufficiency. Proximity to the diaphragm and intrathoracic procedures are the most likely to result in postop respiratory complications.

Airway

- If intubation is necessary, proceed with meticulous antiseptic technique to minimize risk for postop respiratory infection.

Maintenance

- Low tidal volume lung protective strategies intraop to minimize postop respiratory complications.
- Judicious intraop fluid management.
- High FiO_2 likely required to maintain adequate oxygenation.
- If pulm Htn is present, then use additional consideration/preparation for treatment and hemodynamic management for RV failure and pulm Htn.
- Efforts to minimize bronchospasm and optimize bronchodilation: beta-adrenergics, steroids, potent inhalational agent, and adequate anesthetic depth.

Extubation

- Respiratory mechanics and physiology impaired by inhalational anesthetics, narcotics, NMB agents, interscalene block, and high neuraxial blocks, and therefore particular caution regarding residual respiratory depression and muscle weakness on emergence.
- Ensure pt is fully awake and strong with return of baseline respiratory mechanics and particular attention to completely reverse residual muscle relaxant effects.
- If postop ventilation is required, consider early extubation if able to minimize complications of prolonged intubation and ventilation.
- Use bronchodilators and steroids as indicated
- Careful titration of opioids but inadequate treatment of surgical pain will contribute to splinting and insufficient respiratory effort, so consider adjuvants as able.

Adjuvants

- NSAIDs, beta-adrenergic agents, and steroids.
- Consider neuroaxial and/or regional anesthetic technique if possible to minimize requirements for neuromuscular blockade and potentially avoid airway instrumentation and ventilation and to assist in postop pain management.
- Judicious intraop fluid management.
- Vasopressor and inotropic support may possibly be required.
- Consider iNO, or prostacyclin or nebulized iloprost for pts with pulm Htn.

Postoperative Period

- Confusion and decreased LOC secondary to hypoxia and hypercarbia.
- Periop respiratory insufficiency and failure; prolonged intubation/ventilation.
- Low threshold for postop observation in the ICU
- Consider NIPPV or CPAP assistance.
- Nasogastric decompression to potentially improve respiratory mechanics and minimize rates of pneumonia and atelectasis.
- Nutrition: Consider early TPN support, especially in malnourished pts and those in whom prolonged hospitalization is likely.
- Early ambulation and lung expansion maneuvers.
- Adequate but cautious pain control.
- DVT prophylaxis.

Anticipated Problems/Concerns

- Pulm insufficiency; reintubation/ventilation for respiratory failure
- Pulm Htn and RV failure leading to LV decompensation and hemodynamic compromise

Pulmonary Hypertension

Michael Wollenberg | Jeffrey D. Davis

Risk

- Relatively uncommon disease process, with an estimated incidence of 1–5:100,000.
- Frequently identified as a contributing cause of death in USA, resulting in 6.5:100,000 deaths (2010).
- Left heart disease underlies 60–85% of pHTN cases.
- Primary pulmonary disease (e.g., COPD/OSA) is the second most common etiology.
- Chronic thromboembolic disease causes pHTN in 2–4% of pts after acute PE.
- Primary PAH is rare but most amenable to medical therapy.

Perioperative Risks

- RV failure
- Atrial tachyarrhythmias
- Hemodynamic instability

Worry About

- Hypoxia/hypercarbia: Causes pulm vasoconstriction and decreases myocardial contractility, which can lead to RV pressure and volume overload and ultimately RV failure.
- PE: Consider urgent intervention (surgical or thrombolytics) if hemodynamically unstable.
- Hypotension: Decreases RV perfusion and preload, which can worsen failure.
- Atrial tachyarrhythmias: Atrioventricular coupling ensures adequate preload.
- Sympathectomy (if neuraxial blockade present): Disrupts RV homeometric autoregulation in addition to systemic vasodilation.

Overview

- Defined by mean PA pressure (MPAP): ≥25 mm Hg
  - Mild: 25–40 mm Hg
  - Moderate: 41–55 mm Hg
  - Severe: >55 mm Hg
- pHTN is often occult but presents symptomatically with increasing DOE (graded by NYHA classification).
- Diagnosed with RHC.
- PA pressures can be estimated on ECHO by utilizing the modified Bernoulli equation and maximal velocity of the TR jet, if present. (RV systolic pressure >40 mm Hg, which roughly correlates to MPAP >25 mm Hg.)
- Primary periop morbidity and mortality results from RV failure, organ hypoperfusion, and arrhythmias.

Etiology

- WHO classification:
  - PAH: Idiopathic, heritable, drug/toxin-induced, HIV, connective-tissue disease
  - Due to left heart/valvular/congenital heart disease
  - Due to primary lung disease (e.g., COPD, interstitial lung disease, OSA)
  - Chronic thromboembolic pHTN
  - Unclear multifactorial mechanisms: Incorporating chronic hemolysis, metabolic diseases, rheumatologic disorders

Usual Treatment

- Avoid hypoxia/hypercapnia.
- Maintain adequate coronary blood flow (potential role for norepinephrine, vasopressin).
- Maintain sinus rhythm.
- In RV failure, support the right heart with inotropes (dobutamine, milrinone), judicious volume management, and decrease pulm vascular resistance.
- Consider pulm artery cath.
- There is a potential role for nitric oxide and inhaled prostacyclins in acute management of decompensation.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Right heart failure	Poor exercise tolerance	Dependent edema, JVD, systolic murmur (TR)	RHC (gold standard), TTE, TEE, ECG, 6-min walking distance
	Left heart failure, MR/MS, AR/AS, congenital heart disease	DOE, orthopnea, PND	Rales, S_3, cold extremities, prolonged capillary refill	TTE, TEE, ECG, CXR, nt-Pro-BNP
RESP	COPD, sarcoidosis, interstitial pulm fibrosis, chronic thromboembolic pulm disease	DOE, fatigue, cough	Rales, wheezes	CXR, PFTs, sleep study, VQ scan, CT pulm angiogram
	OSA	High STOP-BANG score	Large neck (>40 cm circumference)	Sleep study
GI	Portopulmonary Htn	ETOH abuse, IVDU	Ascites, effusions	Liver US
IMMUNE	Lupus, scleroderma, HIV			ANA, HIV

Key References: Pilkington SA, Toboada D, Martinez G: Pulmonary hypertension and its management in patients undergoing non-cardiac surgery. *Anaesthesia* 70(1):56–70, 2015; Hosseinian L: Pulmonary hypertension and noncardiac surgery: implications for the anesthesiologist. *J Cardiothorac Vasc Anesth* 28(4):1064–1074, 2014.

Perioperative Implications

Preoperative Preparation

- Optimize RV function with inotropes, vasodilators, and diuresis as needed.
- Optimize any comorbidities that contribute to pHTN (most commonly COPD and left heart dysfunction).
- PAH: Determine if responsive to vasodilators during RHC.

Monitoring

- Consider awake arterial line for beat-to-beat BP monitoring at induction and ABGs intraop.
- Consider PA cath or TEE in pts with severe pHTN or right heart failure; useful to guide optimization of cardiac output, to optimize of volume status, to monitor for RV overload, and to monitor for response to pulm arterial vasodilators.
- PA catheter findings suggest RV failure: Uptrending CVP, unexplained decline in PA systolic pressures, unexpected increase in PA diastolic pressures, or decline in PA pulse pressure.

Preinduction/Induction

- Avoid hypoxia (oxygen is a direct pulm vasodilator), hypercarbia, and acidosis.
- Avoid hypothermia (causes pulm vasoconstriction).
- Avoid high peak inspiratory pressures/excessive PEEP (decreases RV preload).
- Maintain appropriate SVR and coronary perfusion with pressors (norepinephrine or vasopressin preferred).
- Sevoflurane provides the least pulm vasoconstriction.
- All inhaled and many IV anesthetic agents are direct cardiac depressants.
- Avoid nitrous oxide (myocardial depressant and increases pulm vascular resistance).

Maintenance

- Goal: Maintain gradient between aorta and RV with vasopressors and inotropes (MAP >65, PVR/SVR ratio <0.5 [or preop ratio], CI >2.2).
- Norepinephrine and vasopressin increase SVR >PVR.
- Dobutamine and milrinone improve R heart contractility while causing pulm artery vasodilatation (and systemic vasodilatation).
- Epinephrine increases RV contractility, and low doses (<0.02 mcg/kg per min) decrease pulm arterial pressures through primary β2 agonist effects.
- Vasodilators (sildenafil, prostacyclins, inhaled NO) have a role in PAH but must be balanced with hypotension due to systemic vasodilation.

Extubation

- Consider extubation in OR if usual respiratory and hemodynamic criteria are met.
- Withdrawal of PEEP on RV preload may induce volume overload and acute decompensation.
- Hypercarbia and hypoxia may occur post extubation.
- BiPAP/CPAP postextubation provide PEEP and may prevent hypercarbia/hypoxia.

Postoperative Period

- Recommend monitoring in ICU postop.
- Respiratory failure and RV failure are the most common causes of death.
- Atrial tachyarrhythmias may contribute to or result from RV failure.
- VTE prophylaxis is paramount (particularly in pts with chronic pulm thromboemboli).

Anticipated Problems/Concerns

- Acute RV failure: Inotropic and vasopressor support (pulm vasodilators if pt has responsive PAH) and careful volume management. Acute PE may respond to careful volume loading, while RV failure from other causes generally responds best to diuresis.
- Hypercapnea/hypoxia: Increase pulm vascular resistance; treat underlying etiology (e.g., COPD, OSA, pulm edema).
- Atrial tachyarrhythmias (predominantly atrial fibrillation): May cause acute RV failure. Rhythm control is preferred in new onset AFib. Cardioversion in hemodynamically unstable pts. Avoid myocardial depressants (beta blockers, calcium channel blockers) in decompensating or unstable pts.
- Venous thromboembolism: Acute worsening of preexisting increased PVR, highly prevalent in pts with chronic pulm venous thromboemboli as well as PAH. VTE prophylaxis very important postop. Early, definitive treatment via thrombolytics or surgical intervention in hemodynamically unstable pts.