Surgical Treatment

- With PA/VSD (usually RV well developed), goal is "two-ventricle repair":
  - Implement a staged repair with BTS early on, followed by VSD closure and valved RV-PA conduit (Rastelli procedure).
  - If multiple MAPCAs present, then implement "unifocalization" (surgically creating a neopulmonary artery by fusing all MAPCAs), followed by VSD closure and valved conduit from RV to neo-PA (Rastelli procedure).
- With PA/IVS (often RV maldeveloped and coronary artery obstruction), goal is "two-ventricle repair," if possible, or else "single-ventricle repair" and eventual heart transplantation. Monitor size/development of RV and extent of coronary artery malformation/obstruction:
  - For adequate RV size for future growth and no coronary malformation, transannual RV outflow patch or radiofrequency assisted valvotomy and dilatation and a systemic-to-PA shunt for future "two-ventricle repair."
  - For small RV (monopartite) and/or coronary malformation, RV outflow patch or dilatation not recommended so as to maintain high RV pressure necessary for coronary blood flow. Stage surgery with BTS, Glenn shunt, total cavo-pulmonary connection (single-ventricle repair), and/or heart transplant.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	RV failure Hypoxemia/metabolic acidosis	SOB	Cyanosis	ECG: RA enlargement, ECG-QRS axis 30–90° CXR: Decreased pulm vascular markings ABG ECHO: PV annulus size, flow across PV, TV size and function, RVOT obstruction, ductus arteriosus, PA anatomy Cardiac cath: Confirm ECHO findings and detect state of coronary blood flow, MAPCAs
RESP	Decreased pulm blood flow	SOB	Tachypnea	ECHO, cardiac cath
SYSTEMIC	Signs of RV failure	Hepatomegaly		

Key References: Malhotra SP, Hanley FL: Surgical management of pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals: a protocol-based approach, *Semin Thorac Cardiovasc Surg Card Surg Annu* 12:145–151, 2009; Mi YP, Chau AK, Chiu CS, et al.: Evolution of the management approach for pulmonary atresia with intact ventricular septum, *Heart* 91(5):657–663, 2005.

Perioperative Implications

Preoperative Preparation

- Ascertain physiologically predominant PA/IVS versus pulm valve stenosis or PA/VSD versus TOF.
- Cath suite intervention versus surgical intervention.
- Patency of ductus arteriosus.
- Maintain pulm blood flow.
- Avoid hyperoxia and hyperventilation in PS/VSD.
- RV dysfunction.
- Presence of RV-dependent coronary circulation.
- Type of shunt to be performed.
- Which systemic artery is to be used for shunt? Avoid during CVP attempts.
- Degree of hypoxemia, metabolic acidosis.

Monitoring

- Standard ASA monitors.
- A-line:
  - Use umbilical artery if good trace.
  - For radial artery, use opposite side of shunt.
  - Use partial clamping of subclavian artery for BTS, with the same implication for pulse oximeter placement and preductal and postductal pulse oximeter.
- CVP for resuscitation drugs.
- Temp/warmers.
- Bubble precaution.

Airway

- ET

Preinduction/Induction

- Continue prostaglandin infusion (0.03–0.1 µg/kg per min) in ductal dependent pts.
- Inhalational induction may be chosen to relax RV infundibulum versus IV ketamine to maintain vascular resistance, depending on presenting predominant physiology and physician preference.
- $ETCO_2$ may significantly underestimate $PaCO_2$ due to limited pulm blood flow.
- Goals: Decrease PVR (to help maintain pulm blood flow), and maintain normal SVR (to avoid desaturation and hypoxemia due to shunt reversal of L-to-R shunt across the VSD by drop in SVR) in PA/VSD.
- Avoid increases in pulm vascular resistance (coughing, bucking; increased PEEP; increased CO_2; decreased PO_2).
- Avoid extreme hyperoxia/hypocarbia in PA/VSD, as this may result in increased L-to-R shunting and systemic hypotension.

Maintenance

- Normothermia
- Normal filling volumes
- Normal myocardial contractility
- Aiming for early extubation

Extubation

- As early as reasonably safe

Postoperative Period

- Pediatric cardiac ICU.
- Continue prostaglandin infusion after palliative repair.

Anticipated Problems/Concerns

- Palliative surgery only
- Definitive procedure later (e.g., Fontan, Rastelli)
- Progressive hypoxemia: Inadequate BT shunt flow or ductus closure

Pulmonary Embolism

Ronald G. Pearl

Risk

- Incidence of pulmonary embolism in USA: 600,000/y
- No racial predilection
- Risk factors same as those for DVT

Perioperative Risks

- Presents a risk for hypoxemia and right heart failure.
- Periop mortality of 50–90% for acute thromboendarterectomy and of ~10% for chronic thromboendarterectomy.
- Postop pulm embolism in up to 1% of surgical pts.
- Pulm embolism accounts for 20–30% of deaths associated with pregnancy.

Worry About

- Recurrent pulm embolism (30% mortality if not treated)
- Right heart failure and CV collapse
- Hypoxemia
- Hemorrhage in pts on anticoagulants or thrombolytics

Overview

- Pulm embolism found in ~20% of autopsied pts.
- Clinical presentation may range from asymptomatic to chest pain and hypoxemia to CV collapse depending on magnitude of the embolus.
- Signs (tachycardia, tachypnea, calf swelling) and symptoms (dyspnea, pleuritic chest pain, calf pain) have low sensitivity and specificity.
- Most pts have DVT (and surgical pts may have pelvic vein thrombi).
- Dx should be based on positive CT pulm angiography after use of a validated clinical decision rule; pulm angiogram should no longer be used.
- Negative D-dimer test excludes Dx in selected low probability pts.

Etiology

- Acquired disease.
- Risk factors present in almost all pts include age >40 y, obesity, malignancy, recent surgery, trauma, pregnancy, immobilization, estrogen use, prior Hx of DVT, and hypercoagulable state (factor V Leiden, deficiency of protein C, protein S, or antithrombin III).

Usual Treatment

- Therapy decreases mortality from 30% to <5%.
- LMWH overlaps with warfarin sodium (INR 2–3) for most pts; use unfractionated heparin (PTT 1.5–2.5× normal) in cases of creatinine clearance <20–30 mL/min, high risk of bleeding, or extremes of weight; use fondaparinux if history of HIT.
- Nonvitamin K-dependent oral antagonists for initial and chronic use are as effective and may have decreased bleeding risk.
- Thrombolytic therapy for massive pulm embolism (hypotension).
- Vena caval filter if pt cannot receive anticoagulants.
- Surgical or catheter thrombectomy in selected cases of acute massive pulm embolism.
- Consider reduced dose thrombolytics or catheter-directed thrombolysis in intermediate-high risk pts (normotensive with RV dysfunction).
- Surgical thromboendarterectomy in selected cases of chronic thromboembolic pulm Htn.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	RV failure	Syncope Dyspnea Palpitations	↑ JVP; RV heave Hypotension Tachycardia Hepatojugular reflux	ECG CT pulm angiography ECHO
RESP	Pulm infarction V/Q abnormality Pain from pleural irritation	Hemoptysis Chest pain Shoulder pain Dyspnea Orthopnea	Tachypnea Pleural rub Wheezing	CXR, SaO_2 ABG Lung V/Q scan CT pulm angiography
CNS	Syncope	Syncope		
MS	Phlebitis	Hx DVT Leg edema Leg pain	Leg edema Inflammation Palpable cord	Compression ultrasonography Impedance plethysmography CT scan

Key References: van der Hulle T, Dronkers CE, Klok FA, et al.: Recent developments in the diagnosis and treatment of pulmonary embolism, *J Intern Med* 279(1):16–29, 2016; Banks DA, Pretorius GV, Kerr KM, et al.: Pulmonary endarterectomy: Part II. Operation, anesthetic management, and postoperative care, *Semin Cardiothorac Vasc Anesth* 18(4):331–340, 2014.

Perioperative Implications

Preoperative Preparation

- Preop Rx with heparin/LMWH and sequential compression devices, which decrease incidence of periop DVT and PE.
- If active DVT, consider preop vena caval filter.

Monitoring

- Consider PA catheter.
- TEE may demonstrate RV dysfunction and PA thromboembolism.

Airway

- None

Preinduction/Induction

- May develop hypotension due to RV failure.

Maintenance

- Adequate preload essential to RV function.
- Systemic vasoconstrictors for hypotension due to RV failure.
- Inhaled vasodilators (NO, prostacyclin) for refractory RV failure.
- Consider ECMO if cardiac arrest.

Extubation

- None

Regional Anesthesia

- Appropriate, especially if compatible with continued anticoagulation.

Postoperative Period

- Resume anticoagulation as soon as possible (or use IVC filter).

Anticipated Problems/Concerns

- RV failure with systemic hypotension may be initial presentation of PE or may develop with recurrent PE.
- Consider PE in all postop pts with unexplained hypoxemia or hypotension.

Pulmonary Fibrosis, Idiopathic

Andrew Oken

Risk

- Present in ~42.7–63:100,000 and incidence is ~16.3:100,000 in USA; more common with increasing age, with majority >55 y.
- Occurrence higher in males than females; risk factors include smoking, exposures, increasing age, family history, chronic reflux, environmental, viral, and genetics.

Perioperative Risks

- Dependent on degree of underlying lung disease and associated comorbidities.
- Pulm Htn, H/O PE, OSA, CKD, CAD, and NYHA class 2 or above are all associated with increasing periop risk for pulmonary failure.
- Obesity (which further reduces lung volumes and worsens ventilation-perfusion mismatch), hypercarbia, and respiratory failure.
- Preop functional status and exercise capacity; risks include albumin <3.5 mg/dL, smoking, COPD, asthma, and FEV_1 <80%.
- Neurologic impairment and immunosuppressive therapy.
- ABG (PaO_2/FiO_2, 225 mm Hg).
- Type of surgery and location (proximity to the diaphragm), especially thoracic surgery for cancer or lung biopsy, along with anesthetic type and surgical duration (>3–4 h).

Worry About

- Progressive respiratory failure, pulm Htn, and potential RV failure
- Postop pneumonia and respiratory failure; prolonged ventilation
- Increased morbidity and mortality, especially in high-risk pts undergoing thoracic procedures

Overview

- IPF, also known as cryptogenic fibrosing alveolitis, is a chronic and progressive fibrosing interstitial pneumonia of unclear etiology.
- Natural history reveals untreated IPF to have an unpredictable course with an insidious progressive nature and deterioration in physical function and capacity, along with a decline in FVC.
- Pts may also present with acute decompensation.
- Prognosis is poor and associated with ~25% survival at 5 y from time of diagnosis and median survival ~3 y from time of diagnosis.
- Morbidity and mortality can occur in approximately ~3–4% following lobectomy and ~11.6% following pneumonectomy.

Etiology

- Unclear, but possible association with smoking, chronic aspiration, and viral infection.
- Important to rule out common misdiagnoses of interstitial lung diseases (e.g., infectious, drug related, exposures [specifically asbestos], hypersensitivity pneumonitis, rheumatoid arthritis, and systemic sclerosis, and usual interstitial pneumonia).
- Clinical course, presentation, and severity variable from pt to pt.

Usual Treatment

- Smoking cessation and active and aggressive management of COPD with bronchodilators, antibiotics, chest physical therapy, and steroids if indicated for persistent wheezing and airflow limitation.