- Heterozygotes may develop venous thrombosis and thromboembolism (rare prior to age 20 y). Protein C levels are 35–65% of normal.
- Acquired causes: Hepatic dysfunction, vitamin K deficiency, DIC

## Usual Treatment

- Heterozygotes:
  - If acute thrombosis, start heparinization (heparin or high-dose LMWH).
  - Long-term anticoagulation with warfarin in pts with Hx of thrombosis. (Heparin therapy should be continued until warfarin is at therapeutic levels to prevent skin necrosis.)
  - Acute thrombosis may require transfusions of FFP to increase protein C levels.
  - During pregnancy, treat with LMWH during and for 4–6 wk after delivery.
- Homozygotes: Periodic FFP or purified protein C concentrate transfusions to provide protein C.
- Acquired:
  - Vitamin K deficiency: Parenteral vitamin K
  - DIC: Treatment of underlying cause

## Assessment Points

| System | Effect | Assessment by Hx | PE | Test |
|---|---|---|---|---|
| HEENT | Retinal vein thrombosis | Hx of vision problems | Ophthalmoscopic exam | |
| CV | MI<br>Angina<br>Peripheral arterial disease | Hx of MI, angina<br>Peripheral vascular thrombosis | Peripheral pulses | ECG |
| RESP | Pulm embolism | Hx of previous pulm embolism | | |
| GI | Mesenteric thrombosis | Hx of bowel infarction | | |
| HEME | Thrombophlebitis | Hx (and family Hx) of thrombophlebitis, pulm embolism | Exam of veins in legs, evidence for lower extremity postthrombotic syndrome | Screen for hypercoagulable state: PTT, proteins C and S, factor V Leiden, antiphospholipid antibody, antithrombin 3 |
| RENAL | Renal vein and artery thrombosis | Hx of renal problem | | BUN/Cr<br>Urine protein |
| DERM | Necrosis | Cutaneous necrosis after warfarin is begun | Cutaneous necrosis | |
| CNS | Intracerebral artery thrombosis | Hx of CVA, TIA | Neurologic exam | |

**Key References:** Goldenberg NA, Manco-Johnson MJ: Protein C deficiency, *Haemophilia* 14(6):1214–1221, 2008; Lipe B, Ornstein DL: Deficiencies of natural anticoagulants, protein C, protein S and antithrombin, *Circulation* 124(14):c365–c368, 2011.

## Perioperative Implications

### Preoperative Preparation

- For homozygotes and symptomatic heterozygotes, FFP and protein C concentrates can be administered to increase protein C levels.
- Warfarin can be stopped a few days before surgery to allow PT to return to normal range and heparin administered until surgery.
- Intermittent pneumatic compression stocking can be placed prior to induction of anesthesia.

### Airway

- Some have suggested that the ETT cuff not be inflated to prevent tracheal venous thrombosis.
- In neonates, there should be an audible leak.

### Preinduction/Induction

- RA preferable if possible

### Maintenance

- Special attention can be paid to positioning to reduce venous and arterial stasis.
- Maintain adequate hydration to reduce thrombosis risk.
- Have propensity to thrombose central venous cath, so minimize use. If cath required, continually ensure patency.
- FFP and/or protein C concentrates should be given to pts with prior thrombotic manifestations and for prolonged operations.

### Adjuvants

- Intermittent pneumatic compression stockings can be used.
- Postop heparinization should be started as soon as deemed safe.

## Anticipated Problems/Concerns

- Increased risk of thrombosis, especially thrombophlebitis and pulm embolism
- When switching from heparin anticoagulation to warfarin, heparin should be continued until warfarin has achieved therapeutic effect to decrease risk of skin necrosis.

# Pulmonary Atresia

Nirvik Pal | Mark T. Nelson

## Risk

- PA/IVS occurs in 3% of all CHD and has a prevalence of 0.07:1000 live births.
- PA/VSD occurs in 3.4% of all CHD.
- 20% of all cases of TOF are physiologically similar to PA/VSD due to extreme pulm artery stenosis.
- Males are affected more than females.

## Perioperative Risks

- RV failure (due to volume overload, pressure overload or both)
- Hypoxemia (leading to metabolic acidosis)
- Myocardial ischemia secondary to aberrant coronary circulation

## Worry About

- RV-dependent coronary circulation in PA/IVS (rapid boluses of fluid through central line may precipitate myocardial ischemia).
- Maintain a patent ductus arteriosus (continue prostaglandin infusion).
- "Suicide RV" is sudden release of pulm valve obstruction leading to hyperdynamic RV and subpulmonic obstruction of RV outlet. Treatment involves a β blockade.
- Hyperventilation and hyperoxia when excess "pulmonary-steal," leading to low cardiac output syndrome and necrotizing colitis (maintain oxygen saturation to 70–80%), mainly in PA/VSD.

## Overview

- Physiologically, PA/IVS and PA/VSD present as two extremes of the same spectrum. Usually associated with other cardiac lesions (e.g., patent foramen ovale, patent ductus arteriosus, possible VSD, ASD).
- PA/IVS often presents with varying extent of RV maldevelopment, TV hypoplasia and stenosis, and RV-dependent coronary blood flow (due to abnormal coronary arteries arising from sinusoids in the RV outlet musculature).
- PA/VSD, on the other hand, the RV by virtue of 'flow-growth phenomenon due to the presence of VSD, enjoys more blood flow and as a result is more completely developed. However, due to the reduced pulm blood flow in PA/VSD, MAPCA develop compensating for the limited blood flow in the maldeveloped PA.

## Etiology

- Congenital

## Usual Treatment

- Prostaglandin $E_1$ infusion
- Surgical correction
- Infective endocarditis prophylaxis
- Palliative therapy

## 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<br>Hypoxemia/metabolic acidosis | SOB | Cyanosis | ECG: RA enlargement, ECG-QRS axis 30–90°<br>CXR: Decreased pulm vascular markings<br>ABG<br>ECHO: PV annulus size, flow across PV, TV size and function, RVOT obstruction, ductus arteriosus, PA anatomy<br>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).