Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Difficult or lost airway	Previous intubation, presence of syndrome (e.g., Pierre Robin, Treacher Collins, Klippel-Feil, Goldenhar)	Dysmorphic, micrognathia	Imaging or genetic testing as needed
CV	Cardiac decompensation	Feeding difficulties, cyanotic spells, vital signs, volume status	Cyanosis, poor saturation, murmur, edema	ECHO, ECG, cardiac cath, vital signs including preductal and postductal oxygen saturation, skin turgor
RESP	Respiratory failure	Feeding difficulties, cyanotic spells, vital signs	Air hunger, use of accessory muscles or instability on vent	Pulse oximetry, ABG
RENAL	Hypovolemia or volume overload, renal insufficiency	Review of intake and output and UO	Skin turgor, fontanel	Basic metabolic labs
HEME	Postop apnea, poor tolerance of blood loss, arrest	Recent blood loss or phlebotomy, previous episodes of apnea, Hx of treatment with caffeine	Pallor, air hunger	CBC

Key References: Spaeth JP, Kurth CD: The extremely premature infant (micropremie). In Coté CJ, Anderson BJ, Lerman J, editors: *Cote and Lerman's a practice of anesthesia for infants and children,* ed 5, Philadelphia, 2013, Elsevier; Jones LJ, Craven PD, Lakkundi A, et al.: Regional (spinal, epidural, caudal) versus general anaesthesia in preterm infants undergoing inguinal herniorrhaphy in early infancy. *Cochrane Database Syst Rev* 6:CD003669, 2015.

Perioperative Implications

Preoperative Preparation

- Look for dysmorphic features, micrognathia, and obvious congenital abnormalities.
- Listen for a heart murmur and have a low threshold for obtaining an ECHO. Obtain a feeding history; breast or bottle feeding is the "stress test" of the neonate.
- Make sure pt is metabolically optimized; check basic labs, especially potassium, calcium, and glucose. Consider getting an ABG.
- Warm the room.
- Calculate estimated blood volume (approximately 95 mL/kg) and maximum allowable blood loss (EBV × [HCT initial – HCT minimum]/HCT initial).
- Calculate doses of all medications and consider unit syringes; avoid drug errors.
- Prepare emergency drugs (atropine, succinylcholine, calcium, epinephrine). Consider drips of epinephrine, dopamine, or milrinone.
- De-air all lines; check and recheck.
- For all but the simplest cases, have an active type and cross. For high-risk procedures, have blood in the room. There is generally very little lead time between unexpected surgical bleeding and the need to transfuse.
- If coming from the ICU, transport with backup emergency airway supplies, warmer, and drugs. Check and recheck endotracheal tube position and IV patency before leaving and upon arrival to the OR.

Monitoring

- In addition to standard monitors, consider preductal and postductal pulse oximetry to watch the effect of a PDA on systemic circulation.
- Consider arterial line for continuous BP and for frequent checks of glucose, electrolytes, and hematocrit. Obtain central access if the risk of needing vasoactive drips is high.

Airway

- Always have an LMA for emergencies. Have an advanced airway device if the baby has a history of a difficult intubation or looks dysmorphic.
- Position with a shoulder roll; the large head relative to the rest of the body tends to flex the neck when supine.
- Prepare multiple sizes of tubes and oral airways.
- Preoxygenate.
- Check for an air leak at <20 cm H_2O.
- Consider a nasal tube if a prolonged intubation is anticipated.
- Check and recheck tube position throughout the case. The distance between a mainstem intubation and perfect positioning can be cm or even mm.
- Watch for kinking of the tube.

Induction

- As with any pt, the acuity of the illness will dictate the anesthetic plan.
- IV and arterial line access can be very difficult in these pts.
- Inhalational induction with sevoflurane is reasonable in a relatively healthy baby undergoing a minor procedure. IV induction is more common, however.
- Consider atropine 20 mcg/kg, fentanyl 2 mcg/kg, rocuronium 0.6 mg/kg for induction.

Maintenance

- For maintenance, consider a fentanyl infusion, low-dose inhalational agent, and nondepolarizing paralytic.
- Reduce oxygen levels and maintain saturations between 88–95% to avoid oxygen toxicity.
- Avoid hypercarbia and acidosis, as both raise pressure in the pulmonary arterial tree.
- Check and recheck the tube for dislodgement or kinks throughout the case.
- Maintain normothermia. While it is easy for pts to get cold, it is also very easy to overheat them with warming devices.
- Warm fluids and blood products if given. Consider "spinning" blood in a blood salvage device (CellSaver) to warm and reduce potassium load.
- Run dextrose containing maintenance fluids or keep TPN running. Check glucose during long cases.
- Be careful not to overtransfuse or fluid overload. Use measured aliquots of fluid and blood products.
- Monitor blood loss; communicate with the surgeons.
- Beware of "monitor failure." Loss of multiple monitors may indicate serious decompensation.

Extubation

- Consider postop location when planning extubation; keep intubated for long transports to ICU.
- If extubating, reverse paralytic.

Postoperative Period

- Consider overnight monitoring for any preterm baby who has not yet reached 56 PCA. Caffeine or theophylline can help attenuate the risk of postop apnea.
- Pain management should include opioids, acetaminophen, and nonpharmacologic adjuncts, along with nonnutritive sucking, breastfeeding, and maternal contact.
- Do not use ketorolac in premature infants.

Anticipated Problems/Concerns

- Difficult IV access
- Postop apnea
- Long transport hazards
- Under-resuscitation or over-resuscitation

Protein C Deficiency

Charles Weissman

Risk

- Congenital deficiency. Homozygote is estimated at 1:500,000-750,000 live births. Occurs when gene coding for protein C on both chromosomes #2 is affected.
- Heterozygote ~0.2–0.4% of healthy population; 2–5% in pts with DVT.
- Acquired deficiency also seen.

Perioperative Risks

- Pts with protein C deficiency are at risk for venous thrombosis and pulm embolism (immobility, endothelial damage, and decreased blood flow during periop period may be triggers).

Worry About

- Increased incidence of thrombophlebitis and pulm embolism
- Thrombosis of other vessels, such as intracerebral and coronary arteries, can occur.

Overview

- Protein C is a vitamin K–dependent protein found in blood and synthesized in the liver.
- Activated after forming a complex with thrombin on endothelial cell receptor thrombomodulin; facilitated by binding to endothelial cell protein C receptors.
- Inhibits blood coagulation by proteolytic inactivation of factors V and VIII.
- Protein S is a cofactor of protein C.
- Stimulates fibrinolysis possibly by neutralizing plasminogen activator inhibitors
- Deficiency causes hyperthrombotic state
- During SIRS and sepsis, there is decreased synthesis of protein C.

Etiology

- Inherited: Autosomal dominant with variable expressivity
- Homozygotes develop life-threatening visceral vessel thrombosis or purpura fulminans (massive cutaneous necrosis) in early neonatal period.

- 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 Angina Peripheral arterial disease	Hx of MI, angina 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 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