- Laryngospasm on anesthetic induction and airway obstruction due to chronic URIs, chronic otitis media, and/or tongue becoming wedged in the cleft
- Difficult intraop oxygenation due to chronic aspiration syndrome
- Increased risk for transfusion if anemic due to poor ability to feed
- Intraop airway obstruction and extubation by Dingman gag
- Intraoperative dysrhythmias caused by surgical infiltration of epinephrine
- Postop airway obstruction by forgotten pharyngeal packs and severe lingual edema
- Undiagnosed associated congenital heart and renal diseases

Overview

- Congenital condition occurs by 7th-12th wk of intrauterine life and is multifactorial, but it can be associated with a single cause such as benzodiazepine usage.
- Cleft palate repair at 12–18 mo; cleft lip closed at 3 mo if also present; single to multiple stage methods employed dependent on type of defect(s).
- Usually not associated with severe blood loss.
- Postop airway obstruction may occur more frequently in prolonged procedures.
- A tongue stitch is often placed at end of surgery for management of possible airway obstruction, and it is removed the next day.

Usual Treatment

- If child is in otherwise good health, a palatoplasty is performed electively.
- All children with cleft palate should have repair by 18 mo to ensure:
  - Normal speech development
  - Appropriate social integration
  - Normal growth of maxilla

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Otitis media Clear rhinorrhea Difficult airway	Ear pain Snore, grunt	Temporomandibular exam Airway exam (micrognathia)	
CV	Associated congenital heart disease	SOB, cyanosis, poor growth	CV exam, club foot	ECG, ECHO
RESP	URI Aspiration	Cough, fever Congestion SOB, cyanosis	Chest exam Chest exam	CXR
GI	Impaired deglutition Malnutrition	Nasal regurgitation Poor growth		Observe feeding
HEME	Anemia	Malnutrition	Pallor	Hgb/Hct
RENAL	Associated congenital defects	UTI	Club feet	UA, BUN/Cr

Key References: Chiono J, Raux O, Bringuier S, et al.: Bilateral suprazygomatic maxillary nerve block for cleft palate repair in children, *Anesthesiology* 120(6):1362–1369, 2014; Steward DJ: Anesthesia for patients with cleft lip and palate, *Semin Anesth Periop Med Pain* 26(3):126–132, 2007.

Perioperative Implications

Preoperative Preparation

- Recognize possibility of multiple future procedures and attempt to minimize stress during induction. Consider oral premedication.

Anesthetic Technique

- GA, usually induced via a mask and using increasing concentrations of volatile agent in O_2, to avoid paralysis until airway is secured.
- Oral airway or gauze packing of cleft may help manual ventilation by preventing tongue from lodging in cleft.
- Intubation, often with RAE ETT secured to mandible, because access to airway may be severely limited.

Monitoring

- Precordial stethoscope, pulse oximeter, and noninvasive BP measurement.
- Maintain normocapnia if epinephrine injection is used.

Postoperative Considerations

- Significant risk for airway obstruction due to edema
- Often obligate mouth breathers
- Judicious use of opioids in a monitored setting; rectal acetaminophen is helpful, especially in combination with suprazygomatic maxillary nerve blocks

Anticipated Problems/Concerns

- Airway difficulty during induction and intubation, especially when associated with other facial anomalies
- Postop airway obstruction due to forgotten pharyngeal pack, severe lingual edema, or obligate mouth breathing

Coagulopathy, Factor IX Deficiency

Thomas M. McLoughlin Jr.

Risk

- Within USA, approximately 4000 persons are affected (20% of all hemophiliacs): incidence: 1:25,000–30,000 males; 75–100 are people born with the disease in USA each year.
- No racial prevalence.
- Highest prevalence overwhelmingly in males.

Perioperative Risks

- Increased risk of hemorrhagic complications from any procedure.
- Of affected individuals, 60% have severe disease (<1% normal circulating factor IX).
- Of carrier females, 10% have abnormal hemorrhage risk.

Worry About

- Excessive and/or uncontrollable hemorrhage
- Tendency for recurrent hemorrhage after initial control
- Expansive deep and soft-tissue hematomas
- Increased risk if hepatic dysfunction from prior plasma product transfusions

Overview

- Inherited; also called hemophilia B or Christmas disease.
- Very similar to hemophilia A (classic hemophilia), but with somewhat less severe bleeding frequency and severity.
- Hemarthrosis accounts for 75% of bleeding episodes. Chronic debilitating arthritis is a common development.
- Soft-tissue hematomas and hematuria also common.
- Intracranial hemorrhage is most common fatal complication.
- Disease severity proportional to circulating factor IX activity (<1% normal activity = severe disease; >5% = generally mild disease).
- Modern maintenance factor replacement treatment results in normal life expectancy.

Etiology

- Sex-linked recessive disorder.
- 70% of cases inherited; 30% result from spontaneous mutation.
- Acquired factor IX deficiency associated with liver disease.
- Adult levels may not be reached in healthy newborns until 6 mo of age.

Usual Treatment

- Restoration of circulating factor IX activity; biological half-life is 18–24 h.
- Plasma-derived pooled factor IX concentrates (AlphaNine SD, Mononine).
- Recombinant factor IX concentrates (BeneFIX [Pfizer], Rixubix [Baxter], and Ixinity [Emergent]) along with an extended half-life (approximately 86 h) recombinant product (Alprolix [Biogen]).
- Of patients in USA, >75% use recombinant products for maintenance therapy.
- In vivo effect of recombinant factor IX products is less than that of plasma-derived products.
- Rarely (3–5% of pts), acquired alloantibodies to administered factor IX substantially complicate treatment.
- Prothrombin complex concentrates and FFP are alternatives for life-threatening hemorrhage if concentrates unavailable.

Assessment Points

System	Effect	Assessment Hx	PE	Test
GI				LFTs if hepatitis Hx
HEME	Coagulopathy	Dental extractions, menses, lacerations, epistaxis	Ecchymoses, hematomas	Prolonged PTT; PT and platelet count usually normal
RENAL	Hematuria; eventual clot formation can obstruct collecting system	Discolored urine		BUN/Cr, urine dipstick or microscopic exam
CNS	Intracranial hemorrhage	Headache	Neurologic exam	
PNS	Discrete peripheral neuropathies	Hx of compressive hematoma	Sensory and motor exam	
MS	Hemarthrosis, chronic arthritis	Painful, warm joints	Decreased ROM	X-rays usually not necessary

Key References: Franchini M: Current management of hemophilia B: recommendations, complications and emerging issues, *Expert Rev Hematol* 7:573–581, 2014; Mensah PK, Gooding R: Surgery in patients with inherited bleeding disorders, *Anaesthesia* 70(Suppl 1):112–120, 2015.

Perioperative Implications

Preoperative Preparation

- Collaborate with consulting hematologist.
- Schedule surgery early in wk to allow optimal postop laboratory support of the assessment of hemostasis; if multiple procedures are contemplated in near future, schedule simultaneously.
- Assess preop factor IX activity; determine goal as guided by magnitude of hemostatic challenge (15–30% factor IX activity for minor lacerations/hematomas; 40–60% for hemarthrosis or major hemorrhage, 50–100% for periop coverage or life-threatening bleeding).
- Units of factor IX needed (plasma-derived) = (Weight in kg) (fractional increase in factor IX activity desired); once-daily dosing is sufficient for maintenance.
- Units of factor IX needed (recombinant) = (Weight in kg) (fractional increase in factor IX activity desired) (reciprocal of observed potency for product). BeneFIX demonstrates 0.8 IU/dL observed activity per administered unit; Rixubis demonstrates 0.9 IU/dL activity per administered unit; Alprolix demonstrates 1 IU/dL activity per unit.

Monitoring

- Confirm expected increase in factor IX activity after preop dose but before incision.

Airway

- Laryngoscopy to avoid tissue trauma; consider mask ventilation.
- Avoid blind oral instrumentation.
- Nasotracheal route is best avoided.

Maintenance

- Consider tourniquets and local cooling to minimize blood loss.

Extubation

- Avoid coughing on endotracheal tube.
- Caution with oropharyngeal suction; best done under direct vision.

Adjuvants

- Regional anesthesia not absolutely contraindicated, but consider with caution; successful brachial plexus blockade at the axilla has been described; no epidural hematoma from neuraxial technique reported when diagnosis of hemophilia B known in advance.
- Postop factor IX activity requirements following major surgery are 75-100% POD 0-3; 60-80% POD 4-6; and 40-60% POD 7-14.

Anticipated Problems/Concerns

- Excessive periop blood loss and hematoma formation
- Potential for delayed or recurrent bleeding after initial control
- Increased likelihood of infectious blood-borne disease (HIV, hepatitis), mostly in pts treated with plasma replacement products before the early 1990s

Coarctation of the Aorta

Thomas M. Chalifoux | Edmund H. Jooste

Risk

- Sixth most-common congenital heart defect: 4:10,000 live births
- Recognized in 5–8% of pts with CHD

Perioperative Risks

- Perioperative mortality: 1% when associated with no other cardiac anomalies in neonates, 10% when associated with a VSD, and 50% when associated with HLHS; children and adults: Less than 0.5%
- Postop risk of paraplegia: 0.5–1.5% (even lower risk if younger than 1 y of age)

Worry About

- Closure of the ductus arteriosus in neonates and infants, which can lead to acute LV failure and hypoperfusion distal to coarctation.
- Maintain adequate perfusion to the lower portion of the body during cross-clamping of the aorta to provide adequate perfusion to spinal cord and abdominal vital organs.
- Intraop systemic Htn proximal to the aortic cross-clamp.
- Acute hypotension and metabolic acidosis on release of aortic cross-clamp.
- Postop systemic Htn.

Overview

- Congenital narrowing of the aorta at or near the ductus arteriosus or ligamentum arteriosum, causing a hemodynamically significant pressure gradient
- Commonly associated defects in neonates and infants: Bicuspid aortic valve, mitral valve anomalies, PDA, aortic hypoplasia, VSD, AV canal defects, d-TGA, and single ventricle variants
- Usually an isolated defect in older children and adults
- Lifelong surveillance needed after repair

Etiology

- Several theories: Abnormal flow patterns in the developing fetal heart, which may cause decreased aortic flow resulting in aortic hypoplasia; ectopic ductal tissue in the aorta; or a combination of both
- Possibly a component of trisomy 13, trisomy 18, deletion of chr 22q11, Turner syndrome, Kabuki syndrome, or Takayasu arteritis

Usual Treatment

- Surgical repair for initial management, using several techniques, including subclavian flap aortoplasty, resection and end-to-end anastomosis, and prosthetic patch augmentation; left thoracotomy (common) and cross-clamp time should be minimized to 20 min, but repair of associated defects may require sternotomy and CPB with or without DHCA.
- Transcatheter balloon angioplasty used for initial management of native coarctation in older infants and young children and for management of recoarctation, which may include endovascular stent placement; also stent procedure of choice in older children and adults. Children with stents may require stent dilation as the child grows.