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.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
GENERAL	Failure to thrive	Poor feeding	Poor growth	Growth chart
NEURO	Intracranial aneurysm (child and adult)			
HEENT	Upper-body Htn (rare in neonate <5 d old)	Epistaxis Headache		Four extremity BP measurement
CV (general)			Systolic pressure and pulse gradient between upper and lower extremities (may not be present with PDA)	ECHO, ECG, CXR, MRI/MRA, cardiac cath with angiography
CV (neonate/infant)	CHF	Poor feeding	Tachypnea, cyanosis, hepatomegaly, metabolic acidosis	ABG, CXR
CV (child/adult)	Development of collateral circulation			CXR showing rib notching (a late finding)
PULM	CHF (neonate and infant)		Resp failure	CXR, ABG
RENAL	Renal failure secondary to poor perfusion (neonate and infant)			Lytes, BUN, creatinine, urine output and analysis
MS	Poor peripheral perfusion Spinal cord compression by dilated anterior spinal artery or branch compressing a nerve root	Claudication, lower extremity pain, paresthesia, muscle weakness	Diminished or absent femoral pulses	

Key References: Kenny D, Hijazi ZM: Coarctation of the aorta: from fetal life to adulthood, *Cardiol J* 18(5):487–495, 2011; Landsman IS, Davis PJ: Aortic coarctation: anesthetic considerations, *Semin Cardiothorac Vasc Anesth* 5(1):91–97, 2001.

Perioperative Implications

Preoperative Preparation and Induction

- Neonate/infant: Maintain PDA with PGE_1. PDA closure can lead to CHF, upper-body Htn, and lower-body hypoperfusion and shock.
- The presence of a VSD leads to significant left-to-right shunting and a further steal of the systemic blood flow. Do not decrease PVR further by hyperventilation or the use of 100% O_2.
- Right lateral decubitus position used for left thoracotomy. Good padding is important.
- Regular ETT used for neonates and infants, but consider bronchial blocker or double lumen ETT in older children and adults.

Monitoring

- Standard monitors, pulse oximeter × 2 (right upper and either lower extremity), and urinary cath.
- Right upper-extremity arterial cath (radial, ulnar, or axillary) or lower-extremity arterial cath if pressure gradient is high or a combination of arterial and NIBP monitoring in the RUE and a lower extremity.
- Central venous access required for infusion of vasoactive medications.
- SSEPs may be used to motor spinal cord perfusion during aortic cross-clamping (particularly if aortic gradient is high or there is little collateral circulation) in older children.

Maintenance

- To prevent spinal cord ischemia, passively cool to core temp 34–35° C, maintain normocapnia, and keep distal mean arterial pressure >40 mm Hg.
- Control Htn with titratable agents: Inhalation agent, sodium nitroprusside, esmolol, and nicardipine.
- If mean arterial pressure <40 mm Hg or there is significant change in the SSEP signal with aortic cross-clamp application, institute left heart bypass.
- Be prepared to treat a sudden drop in BP and acidosis following aortic cross-clamp release with fluids and sodium bicarbonate.

Postoperative

- Neonates and infants with CHF remain intubated and ventilated until condition improves.
- Children and adults may usually be extubated in the OR.
- Pain management: Opioids, dexmedetomidine, intercostal nerve block by surgeon, paravertebral cath, and epidural cath (must consider risk of epidural hematoma).

Anticipated Problems/Concerns

- Paraplegia likely secondary to spinal cord ischemia, particularly if clamp time >30 min
- Postcoarctectomy syndrome: Severe abdominal pain with tenderness, Htn, fever, vomiting, ileus, melena, and leukocytosis (occurs 2–3 d postop)
- Pulm Htn in neonates and infants with CoA and VSD (Rx: NO and milrinone)
- Stridor/partial airway obstruction at extubation secondary to recurrent laryngeal nerve injury
- Ventilatory compromise at extubation secondary to phrenic nerve injury causing hemidiaphragmatic paralysis
- Intraop and postop bleeding
- Aortic aneurysm, dissection, and rupture
- Neurologic symptoms from subclavian steal secondary to reduced perfusion of the left arm after subclavian flap angioplasty
- Chylothorax from thoracic duct injury
- Recoarctation (late complication)

Cogan Syndrome

Michael Carrigan | Jeffrey R. Kirsch

Risk

- Extremely rare: approximately 250 reported cases in the literature
- Mean presentation 30–40 y; however, cases in children and elderly reported
- No predilection for gender, race, or ethnicity
- Possible association with IBD

Perioperative Risks

- Hemorrhage
- Thrombosis and organ/limb ischemia
- Adrenal insufficiency and immunosuppression due to chronic treatment
- Postop N/V with vestibuloauditory dysfunction

Worry About

- Activity state of disease and hemorrhage/extension of pathologic vasculitis
- Coexisting vasculitis affecting cerebral, cardiac, mesenteric, and renal perfusion
- Sepsis with immunosuppression

Overview

- Heterogeneous presentation of nonsyphilitic interstitial keratitis and vestibuloauditory symptoms within 2 y of each other; note an atypical version allows exceptions to these criteria
- 10–15% of pts develop large cell vasculitis, usually aortitis
- Coronary involvement: often asymptomatic
- Typically sudden severe bilateral hearing loss; distinct from unilateral Meniere disease; deafness develops in ~50% of pts.
- Recurrent flares for majority of pts
- Mean long-term survival: 20+ y after diagnosis

Etiology

- No definitive cause, but an autoimmune process is suspected; often preceded by a viral prodrome.
- Proposed mechanisms include antibodies to an inner ear peptide, Cogan peptide, and HSP70.
- Rheumatoid factor and ANA are not consistently associated with diagnosis, but a small percent of pts are ANCA+.
- Approximately 50% have a history of daily smoking, and approximately 33% have or develop IBD.

Usual Treatment

- Topical steroids and mydriatic agents for isolated anterior chamber disease
- Systemic immunosuppressives for posterior chamber, inner ear, and vasculitis