

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 PGE₁. 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% O₂.
- 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

- Typically high-dose steroids (1 mg/kg per d prednisone for 2–4 wk) until hearing improves, and then taper over 3–6 mo
- MTX, cyclophosphamide, azathioprine, leflunomide, tacrolimus, and rituximab all with case reports of effectiveness, usually reserved for severe organ or life-threatening presentations
- Surgical repair or bypass of diseased segments: favorable only when activity of the disease is under control with medical treatment
- Cochlear implant use: Very successful

Assessment Points				
System	Effect	Assessment by Hx	PE	Test
HEENT	Inner ear dysfunction Interstitial keratitis	Dizziness, tinnitus Photophobia, redness, tearing, blurry vision, and oscillopsia	Slit lamp exam	Calorics and audiogram with sensorineural hearing loss
RESP	Pulmonary embolism Pneumonia (due to immunosuppression)	Dyspnea Cough	Tachypnea Lung field consolidation Wheezing	CXR, V/Q scan, CT scan Bronchoscopy BAL ABG
CV	Aortitis Coronary arteritis Limb ischemia AI, MR	Chest/back/abdominal pain Dyspnea Orthopnea	Tachycardia Hypotension Disparate limb BP Heart murmur	ECG, TTE CT scan Limb duplex scan Angiography
GI	Mesenteric ischemia	Post prandial abdominal pain N/V	Abdominal tenderness Splenomegaly	CT scan Angiography Abdominal US
CNS	Intracranial manifestations of vasculitis	Weakness Numbness Falling Incoordination Difficulty speaking	Gait instability Dysmetria Functional neurologic deficits	Head CT and MRI to R/O tumor/stroke Carotid duplex US
HEME	Pancytopenia	Easy bleeding and bruising Fatigue Fever	Petechiae Rash Pallor Lymphadenopathy	CBC and differential Reticulocyte count Peripheral smear
METAB/ ENDO	Iatrogenic Cushing syndrome	Poor wound healing Skin changes Body habits changes Emotional/psychiatric changes	Striae Buffalo hump Skin wounds Moon facies Htn Hirsutism	Electrolytes HgbA _{1c}
RENAL	Glomerulonephritis	Hematuria Oliguria Headache Edema	Htn Peripheral edema	BMP, albumin, UA

Key References: Singer O: Cogan and Behçet syndromes, *Rheum Dis Clin North Am* 41(1):75–91, 2015; Gluth M, Baratz K, Matteson E, Driscoll CL: Cogan syndrome: a retrospective review of 60 patients throughout a half century, *Mayo Clin Proc* 81(4):483–488, 2006.

Perioperative Implications

Preoperative Preparation

- Assess disease activity state and screen for concomitant vasculitic processes.
- Ensure adequacy of blood products and IV access.
- Severe neutropenia may warrant prophylactic antimicrobial therapy and reassessment of timing risk/benefit.
- Concomitant steroid therapy and necessity of stress doses should be considered.

Monitoring

- Consider awake arterial line in appropriate limb as indicated.

- Consider CVP, TEE, or PA cath as indicated for disease burden and procedure.
- Consider BIS if cerebral circulation is affected.

Airway

- Use caution with edematous airway mucosa.

Preinduction/Induction

- Tailor afterload and preload management to cardiac function and concomitantly affected organs including cerebral, renal, and mesenteric beds.
- Avoid hypotension in concomitantly affected organs.

Maintenance

- Judicious blood pressure management to preserve diseased organ bed perfusion

Extubation

- Avoid Htn with vascular repairs and aneurysmal burden.

Postoperative Period

- Continue monitoring of hemodynamics.
- Maintain vigilance for hemorrhage.
- Immunosuppressed patients have an increased susceptibility to infection.
- Watch for signs of adrenal insufficiency.

Anticipated Problems/Concerns

- Maintain a low threshold to evaluate the occult disease burden of other organ systems not identified preop.

Complement Deficiency

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Risk

- C1 esterase inhibitor–deficiency incidence: 1:50,000–150,000 of the general population.
- Symptoms onset and diagnosis occur approximately at 20 y, and by 30 y approximately 98% of pts have symptoms.
- C2 deficiency incidence: <0.1% of the general

- Male versus female ratio: 1:6.
- Higher incidence (6%) in pts with autoimmune disease (see Immune Suppression).
- Incidence in pts with Hx of *Neisseria meningitidis*: 15%.
- C3 and C5–C8 deficiencies have increased risk for infections.

Perioperative Risks

- Possible life-threatening airway compromise
- Increased risk of postop infection, particularly if the deficiency affects the early complement components
- Risk for inflammatory complications (e.g., glomerulonephritis, vasculitis)