

Takayasu Disease

Risk

- Worldwide incidence: 2.6 cases per million per y.
- Race with highest prevalence: Asian.
- Females 8–9 times more likely to be affected than males.

Perioperative Risks

- Severe uncontrolled Htn leading to end-organ dysfunction
- Stenosis of major blood vessels affecting regional circulation
- Difficulties in monitoring BP.
- Long-term corticosteroids.

Worry About

- Multiple occlusions of peripheral arteries, CHF, stroke, cardiac valve dysfunction, hypertensive episodes, intracranial hemorrhage, and iatrogenic adrenal suppression

Overview

- A rare systemic inflammatory large-vessel vasculitis primarily affecting the aorta and its main branches
- Initial “inflammatory phase” characterized by systemic illness with malaise, fever, weight loss, and fatigue
- Secondary “pulseless phase” characterized by vascular insufficiency from intimal narrowing of the vessels manifesting as arm or leg claudication, renal artery stenosis causing Htn, and neurologic manifestations due to decreased blood flow to the brain

Etiology

- Unknown; some evidence to support a genetic predisposition.
- Cell-mediated immune mechanisms are of primary importance.

- Panarteritic inflammatory infiltrates cause marked thickening of the affected artery. Segmental and patchy granulomatous inflammation leads to arterial stenosis, thrombosis, and aneurysms.
- Initial vascular lesions frequently occur in the left subclavian artery, with other branches and the aorta becoming affected as the disease progresses.

Usual Treatment

- Two components:
 - Controlling inflammatory process: Corticosteroids are the mainstay of therapy. Additional cytotoxic agents may be required to achieve remission and steroid taper.
 - Controlling Htn: Antihypertensive agents. Aggressive therapy is necessary to prevent complications. Low-dose aspirin may have therapeutic effect.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Uncontrolled Htn Aortic regurgitation Ischemic heart disease CHF Stenosis, thrombosis, or aneurysms of systemic and pulm vessels	Poor exercise tolerance, arm or leg claudication, angina, CHF symptoms, syncope, headaches, Hx of CVA	Chest exam for signs of CHF Absence of peripheral pulses BP with difference >10 mm Hg between the arms Arterial bruit Raynaud syndrome	LVH on ECG ECHO Angiography/MRA
RESP	Pulm Htn Ventilation-perfusion mismatch	Dyspnea		CXR ABG
RENAL	Renal artery stenosis		Uncontrolled BP Renal bruit	BUN/Cr Doppler US
ENDO	Cushingoid	Long term steroid use	Features of Cushing	Check blood sugar
CNS	CVA, intracranial hemorrhage, syncope, retinopathy	Headache Amaurosis fugax Stroke/TIA Seizures	Ophthalmic exam Carotid bruit Focal neurologic deficits	Angiography/MRA/CT
HEME	Anemia	Fatigue		FBS

Key References: Kathirvel S, Chavan S, Arya VK, et al: Anesthetic management of patients with Takayasu's arteritis: a case series and review, *Anesth Analg* 93(1):60–65, 2001; Keser G, Direskeneli H, Aksu K: Management of Takayasu arteritis: a systematic review, *Rheumatology* 53(5):793–801, 2014.

Perioperative Management

Preoperative Preparation

- Assess myocardial and volume status.
- Assess peripheral pulses.
- BP control.

Technique

- General anesthesia involving endotracheal intubation/extubation and inadequate depth may result in considerable BP fluctuation and may precipitate cerebral hemorrhage, rupture of aneurysms, and cardiac dysfunction.
- Regional techniques avoid the need for cerebral monitoring, although they may be associated with hypotension. Anticoagulation precludes. Epidural and spinal used successfully for cesarean section.

Monitoring

- Measure BP proximal to areas of arteritis. When weak or absent peripheral pulses, pulse oximetry,

automatic NIBP, and Doppler flow signals can be used to record blood pressure.

- Avoid invasive BP due to increased risk of vessel damage. Femoral may be preferred site.
- Consider cerebral monitoring if asleep and compromised carotid blood flow (e.g., transcranial Doppler, EEG, cerebral oximetry).
- ECG and urine output to assess adequacy of coronary and renal blood flow.

Airway

- Hyperextension of head during laryngoscopy may compromise cerebral blood flow.

Induction

- Avoid a hypertensive crisis during tracheal intubation. Regional anesthesia should proceed with cautious neuraxial dosing to minimize hypotension.

Maintenance

- Maintain BP, avoid tachycardia, and maintain peripheral perfusion. Avoid excessive hyperventilation due to effect on CBF.

Extubation

- Aim for prompt awakening to allow prompt evaluation of mental status.

Adjuncts

- If risk of adrenal suppression from long term steroids, consider need for supplemental periop dosing.
- Consider periop antibiotics if immunosuppressed.

Postoperative Period

- Continue CV, CNS, and renal monitoring. Control BP. Consider ICU/PACU overnight.
- Risk of infection and sepsis due to immunosuppression.

Tetanus

Risk

- A major public health problem in the developing world, but improving; responsible for 200,000–300,000 deaths/y in 2000 and only 60,000 in 2013, and the vast majority were neonatal deaths.
- Incidence in USA: 0.16 cases/million population (1998–2000).

- Highest incidence in USA is among the elderly (>60 y), persons of Hispanic ethnicity, older adults with diabetes, and parenteral drug users.

Perioperative Risks

- Difficult airway or intubation in the presence of masseter spasm, neck rigidity, or opisthotonus

- Autonomic instability with sudden fluctuations in BP, arrhythmias, cardiac failure, and cardiac arrest

Worry About

- Spasms of the laryngeal and respiratory muscles can be life-threatening as a result of airway obstruction or chest wall rigidity respectively, and may mandate urgent ET intubation.

Kirk Lalwani