

# 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<br>Aortic regurgitation<br>Ischemic heart disease<br>CHF<br>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<br>Absence of peripheral pulses<br>BP with difference >10 mm Hg between the arms<br>Arterial bruit<br>Raynaud syndrome | LVH on ECG<br>ECHO<br>Angiography/MRA |
| RESP   | Pulm Htn<br>Ventilation-perfusion mismatch   | Dyspnea   |  | CXR<br>ABG                            |
| RENAL  | Renal artery stenosis  |   | Uncontrolled BP<br>Renal bruit   | BUN/Cr<br>Doppler US                  |
| ENDO   | Cushingoid   | Long term steroid use   | Features of Cushing  | Check blood sugar                     |
| CNS    | CVA, intracranial hemorrhage, syncope, retinopathy   | Headache<br>Amaurosis fugax<br>Stroke/TIA<br>Seizures   | Ophthalmic exam<br>Carotid bruit<br>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

- Respiratory failure may require NM paralysis in addition to sedation for effective PPV in the presence of severe spasms.
- Autonomic instability: Tachycardia, bradycardia, Htn, hypotension, arrhythmias, cardiac failure, and repeated cardiac arrest.
- Pneumonia, sepsis, myoglobinuria, pulm embolism, bony fractures, and hyperthermia.
- Autonomic instability is a hallmark of the disease and may cause fatal cardiac arrest.
- Initial injury may be insignificant or unnoticed by the pt.
- Neonatal tetanus typically presents 6–8 d after birth with trismus and inability to feed.
- Tetanus may follow surgery (usually intraabdominal or on contaminated tissues), burns, gangrene, dog bites, chronic infection, parenteral drug use, dental infection, abortion, and childbirth.

### Usual Treatment

- Neutralize circulating toxin with IV human antitetanus globulin.
- Eradication of the organism by wound care, surgical debridement, and antimicrobial therapy.
- High-dose metronidazole or penicillin G (erythromycin if penicillin allergy) therapy IV for 10 d is effective at eradicating spores and bacilli.
- Control muscle spasms by sedation, other muscle relaxants, and NM paralysis.
- Magnesium may control spasms and autonomic disturbances in mild cases, but has no beneficial effect on mortality compared to diazepam, which is considered the standard of treatment.
- Supportive therapy, including ventilatory support, treatment of autonomic instability, nutritional support, prophylaxis of DVT, and prevention of nosocomial infection, particularly ventilator-associated pneumonia.

### Overview

- Infection of penetrating wounds or devitalized tissue with spores of anaerobic gram-positive bacillus *Clostridium tetani*; enters the CNS via peripheral nerves and spreads via retrograde intraneuronal transport to disable inhibitory pathways in the spinal cord and brain (glycine and GABA).
- CNS disinhibition characteristically begins with spasms of the masseter muscles (“risus sardonius,” lockjaw) and progresses to involve rest of the body, including spasms of respiratory muscles (“respiratory convulsions”) that cause glottic spasm, airway obstruction, hypoxia, and respiratory failure.

### Etiology

- Infection of penetrating wound or devitalized tissue by spores of anaerobic, gram-positive bacillus *Clostridium tetani*; they proliferate and produce a potent exotoxin, tetanospasmin.
- Tetanospasmin is taken up by motor nerve endings and spreads to other neurons in skeletal muscle, the spinal cord, and brain, where it principally inactivates inhibitory interneurons in glycinergic and gamma-aminobutyric acid pathways.

### Assessment Points

| System | Effect  | Assessment by Hx            | PE   | Test         |
|--------|---|-----------------------------|--|--------------|
| HEENT  | Laryngospasm and glottic obstruction  | Dyspnea, noisy breathing    | Stridor, retractions of accessory muscles, limitation of mouth opening and ROM of neck         |              |
| CNS    | Generalized or localized muscle rigidity and spasms                         | Dysphagia, drooling, spasms | Opisthotonus, trismus, “risus sardonius,” onset of spasms with minimal stimuli, bony fractures |              |
| CV     | Cardiac failure, myocarditis, arrhythmias, Htn, hypotension, cardiac arrest | SOB, palpitations           | Episodic fluctuations in BP, heart rate; arrhythmias, signs of cardiac failure                 | ECG, ECHO    |
| RESP   | Hypoventilation, apnea, respiratory failure, pneumonia                      | Dyspnea                     | Hypoventilation, limited chest excursions, decreased breath sounds, rhonchi, cyanosis          | ABG, CXR     |
| RENAL  | Rhabdomyolysis  | Pink or red urine           | Hematuria  | US, serum CK |

**Key References:** Rodrigo C, Fernando D, Rajapakse S: Pharmacological management of tetanus: an evidence-based review, *Crit Care* 18(2):217, 2014; Rodrigo C, Samarakoon L, Fernando SD, et al: A meta-analysis of magnesium for tetanus. *Anaesthesia* 67(12):1370–1374, 2012.

### Perioperative Implications

#### Preinduction/Induction/Maintenance

- Adequate sedation with benzodiazepines to control spasms; muscle relaxants may be necessary.
- Minimize environmental stimuli.
- Difficult airway or intubation: Consider fiberoptic intubation.
- Avoid pancuronium and desflurane (sympathetic stimulation).
- Resistance to multiple nondepolarizing agents has been described.

#### Monitoring

- ECG for dysrhythmias
- Echocardiography (CV decompensation)
- Arterial line for continuous BP measurement and arterial blood gas measurement
- NM monitoring with nerve stimulator

#### General Anesthesia

- Magnesium sulfate may be useful in controlling spasms, decreasing autonomic instability, and decreasing the requirements for sedative drugs.
- Watch for S-T segment and T-wave changes that may indicate toxic myocarditis.
- Hypotension and bradycardia may be indicative of brainstem involvement and a poor prognosis.
- Elective tracheostomy recommended for long-term ventilator support and pulm toilet.
- Consider pulm embolism in the event of sudden decompensation during anesthesia.
- Maintain alkaline diuresis in the event of myoglobinuria.

#### Regional Anesthesia

- Consider adding epidural anesthesia for autonomic hyperreactivity.

#### Postoperative Period

- Endotracheal intubation or tracheostomy is needed for assisted ventilation on ICU with sedation and NMBs.
- Benzodiazepines, magnesium sulfate, opioids, clonidine, and intrathecal baclofen may help control spasms; magnesium also decreases autonomic instability and the need for sedation.
- Nutritional support via enteral or parenteral feeding.
- DVT prophylaxis to prevent pulm embolism.

#### Anticipated Problems/Concerns

- Sudden CV instability or cardiac arrest may occur.
- Propranolol, labetalol, and phentolamine are assoc with increased risk of cardiac arrest.
- Mortality in US averages about 10%, rising to 50% in pts >60 y of age.
- Abnormal neurologic findings may persist for up to 2 y following recovery.

## Tetralogy of Fallot

Sarah Deverman

### Risk

- Occurs in 4–5:10,000 live births (1:2,000–2,500)
- Most common cyanotic CHD (10% of all CHDs)
- Occurs equally in males and females

### Perioperative Risks

- If unrepaired, tet spells can lead to RVH, RV failure, and death (50% in first year of life).
- Mortality after TOF repair: 5–8% in first 2 y postrepair (if uncomplicated anatomy).

- Increased mortality if coexisting PA hypoplasia, atresia, or major AP collaterals.

### Worry About

- Increased R-to-L shunt from decreased SVR or increased PVR
- Crying and agitation leading to tet spell leading to more hypoxemia, hypercarbia, acidosis
- Air bubbles in IV tubing
- Polycythemia and assoc thrombocytopenia

- RV failure after inadequate or late repair
- Arrhythmias following repair

### Overview

- Anatomy:
  - RVOT obstruction: Infundibular narrowing, pulm stenosis, PA hypoplasia, pulm atresia.
  - VSD: Large, unrestricted.
  - Overriding aorta.
  - RV hypertrophy.