

Sarcoidosis

Risk

- Risk varies; ≤ 1 –80:100,000, with highest incidence in Sweden; in USA, occurs in 30/100,000.
- Presents in pts ages 20–40 y in USA.
- More common in African Americans than whites in USA.
- Females at greater risk than males.

Perioperative Risks

- Severity depends on degree of airway, lung, cardiac, and CNS involvement.

Worry About

- Airway granulomas distorting and obstructing anatomy risking obstruction with sedation and making intubation potentially difficult

- Degree of lung involvement and pulm fibrosis
- Cardiac involvement, heart block, arrhythmia, and CHF
- CNS involvement

Overview

- Multisystem granulomatous disorder with widespread noncaseating epithelioid cell granulomas.
- Lung most frequently affected organ.
- Airway abnormality secondary to granulomas.
- Local organ distortion can result in symptoms.
- In mononuclear inflammatory cells, T-helper cells + mononuclear phagocytes lead to the formation of granulomas.

Etiology

- Unknown disease due to exaggerated cellular immune response involving mononuclear phagocytes and T lymphocytes

Usual Treatment

- Steroids: Oral prednisone (inhaled steroids not shown to be consistently effective)
- NSAIDs: Salicylates
- Chloroquine or hydroxychloroquine for mucocutaneous sarcoidosis
- If steroids ineffective, methotrexate or immunosuppressive agents

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Involvement of nares, polyps with distorted anatomy; larynx granulomas, epiglottitis, arytenoid involvement	Dyspnea Breathing difficulty	Nasal stuffiness, wheezing, hoarseness, stridor Can see vocal cord palsy or paralysis	Laryngoscopy
CV	Heart block or arrhythmia Cor pulmonale secondary to RV enlargement	Palpitations	Arrhythmia Rales	ECG
RESP	Pulm granulomas, airway obstruction Bilateral hilar lymphadenopathy (eggshell calcifications of hilar nodes); pulm fibrosis; interstitial disease	Dyspnea Wheezing, cough	Dry rales Wheezes	CXR PFTs (decreased vital and diffusing capacities) ABG CT if airway obstruction considered an issue
GI	Liver involvement			Increased LFTs, increased alkaline phosphatase
ENDO	DI	Thirst		
RENAL	Increased Ca^{2+} resorption leading to nephrocalcinosis and other renal issues			BUN/Cr
CNS	Nerve involvement DI	Space-occupying lesions Seizures Psychiatric examination	Focal nerve deficits	

Key References: Sanders D, Rowland R, Howell T: Sarcoidosis and anaesthesia, *BJA Education* 16(5):173–177, 2015; Iannuzzi MC, Rybicki BA, Teirstein AS: Sarcoidosis, *N Engl J Med* 357(21):2153–2165, 2007.

Perioperative Implications

Preoperative Preparation

- Adequate steroid coverage.
- For pulm and airway, determine if airway obstruction exists and degree of pulm involvement. Evaluate for Hx of SOB and dyspnea.

Obtain CXR and consider PFTs and preop ABG based on symptoms and Hx. If airway obstruction is suspected, obtain CT to better define issues.

Airway

- Distortion or obstruction secondary to granulomas
- Hypoxia secondary to lung disease

Monitoring

- Observe for heart block.
- Arrhythmia.

Anticipated Problems/Concerns

- Airway problems secondary to distorted anatomy
- Pulm problems secondary to lung involvement

Sarcoma

Risk

- Malignant bone tumors. Incidence is 1:100,000; 3000 new cases/y in USA, with bimodal age distribution (first peak during adolescence, second peak in older adulthood).
- Soft tissue sarcomas: Incidence is 1:100,000 for ages <20 y and 7:100,000 for ages ≥ 20 y; 12,000 new cases/y in USA; mean age at diagnosis is 58 y.
- Prevalence equal in both genders and all races except Ewing sarcoma (high Caucasian predominance).
- Overall: 15% of cancers in children age <20 y.

Perioperative Risks

- Morbidity and mortality related to surgical procedure.
- Metastatic vital organ involvement, especially lungs and liver.
- Mass effect, direct compression of organs and vascular structures.

Worry About

- Adriamycin-induced cardiotoxicity (global LV hypokinesia)
- Mitomycin-induced acute pulm toxicity, pulm fibrosis, ARDS with increased FiO_2
- Immunosuppression, hemorrhagic cystitis, renal failure induced by chemotherapeutic agents

Overview

- Heterogeneous group of malignant tumors of connective tissue derived from the embryonic mesoderm.
- Two most common sarcoma forms: Malignant bone tumors and soft tissue sarcomas.
- Malignant bone tumors such as osteosarcomas and Ewing sarcomas can be found throughout bones and cartilage.
- Soft tissue sarcomas often arise from muscles, joints, fat, nerves, deep skin tissues, and blood vessels.
- Can spread aggressively by local invasion and early hematogenous spread, especially to lungs.

Etiology

- Genetic factors, high-dose radiation, carcinogens (dibenzanthracene, methylcholanthrene), and some viral infections (Moloney sarcoma virus, human herpes virus 8) may predispose pts to sarcoma.
- von Recklinghausen's disease: 10–12% develop neurofibrosarcomas.
- Li-Fraumeni syndrome: Strong association with sarcomas.
- Retinoblastoma survivors have much higher incidence of osteosarcomas.
- Pager's disease: 0.9% develop osteosarcoma.
- Kaposi sarcoma strongly associated with human herpes virus 8 in immunocompromised pts.

Usual Treatment

- Neoadjuvant chemotherapy with antineoplastic agents
- Wide surgical resection
- Radiation

Assessment Points				
System	Effect	Assessment by Hx	PE	Test
CV	Atrial myxoma (ball-valve effect)	Sx of CHF, pulm edema	Rales S ₃	CXR ECHO CT angio
	Vena cava obstruction	RV failure, CV collapse	Possible caput medusae, venous engorgement, edema	
	SVC syndrome	Head swelling, airway edema, increased ICP	Venous congestion of head and neck	CT angio
RESP	Pulm embolus	Dyspnea		PE protocol CT
GI	Gastroparesis Bowel obstruction Hepatic metastases Sarcoma of ampulla of Vater	Early satiety Vomiting Obstructive jaundice Hepatic dysfunction	Abdominal distention Jaundice	X-ray of abdomen EGD, bilirubin levels
HEME	Hypercoagulable Pancytopenia due to chemotherapy Anemia due to GI hemorrhage	Opportunistic infections Bruising, bleeding	Gross rectal bleeding	PT/PTT Leukocyte and plt counts CBC Stool guaiac
RENAL	Compression of ureters by retroperitoneal tumor	Sx renal failure		BUN/Cr Renal US or CT
CNS	CN compression	Various symptoms Dysphagia Loss of sensation, motor function	Neurologic exam	EMG Head CT
MS	Bone sarcomas Limb loss	Hypercalcemia	Chvostek's sign	Blood Ca ²⁺

Key References: Burningham Z, Hashibe M, Spector L, et al.: The epidemiology of sarcoma, *Clin Sarcoma Res* 2(1):14, 2012; Makela J, Kiviniemi H, Laitinen S: Prognostic factors predicting survival in the treatment of retroperitoneal sarcoma, *Eur J Surg Oncol* 26(6):552–555, 2000.

Perioperative Implications

Preoperative Preparation

- Use aspiration precautions for pts with gastroparesis, including prokinetics and NG decompression.
- Assess end-organ impairment (especially heart and lungs) secondary to chemotherapeutic agents.

Monitoring

- Use arterial line and CVP or PA cath for resection of large tumors.

Airway

- Risk of aspiration with large abdominal mass or brainstem compression.
- Possibility of difficult airway with SVC syndrome.

Induction

- Hemodynamic instability with cardiac involvement or caval compression.

Maintenance

- Potential blood loss

Extubation

- Awake if at risk for aspiration due to GI or CNS involvement.

Adjuvants

- Altered pharmacokinetics with hepatic or renal involvement.

Postoperative Period

- Pulm embolism, coagulopathy

Anticipated Problems/Concerns

- Adverse effects of chemotherapeutic agents
- Respiratory compromise due to pulm metastases
- Mass effect and/or organ compression and functional impairment
- Effects of prolonged anesthesia
- In prolonged abdominal cases, hypothermia, complications of massive transfusion

Scheie Syndrome (Mucopolysaccharidosis Type IS)

Arun K. Gupta

Risk

- Incidence: 1:500,000
- Inherited as autosomal recessive

Perioperative Risks

- Difficult intubation
- Positioning difficulty
- Cardiac problems

Worry About

- Macroglossia leading to difficult intubation
- Valvular heart disease, cardiomyopathy
- Stiff joints

- Spastic paresis
- Poor IV access

Overview

- Described by American ophthalmologist Harold Glendon Scheie in 1962.
- Mildest form of mucopolysaccharidosis type (1) and a rare lysosomal storage disease.
- Pts are of normal height and do not show intellectual deficiency.
- Corneal clouding and glaucoma.
- Coarse facial features.
- Normal life expectancy, stiff joints, and aortic regurgitation.

Etiology

- Caused by mutations in genes that control production of enzyme alpha-L-iduronidase (IDUA).
- The gene known as IDUA causes deficiency of enzymes and results in buildup of undigested mucopolysaccharides in the cells.
- Diagnosis occurs by elevated GAG in urine and demonstration of IDUA deficiency in leukocytes.

Usual Treatment

- Enzyme replacement
- Hematopoietic stem cell transplant
- Bone marrow transplant and cord blood transplant