Adjuvants
- Careful consideration to increased bleeding risk if considering regional anesthesia.
- Must obtain bleeding time, coagulation studies, and platelet count prior to performing regional anesthesia.
- Parenteral opioids are acceptable.

Postoperative Period
- Extended postoperative monitoring for 24 h with pulse oximetry
- May require frequent suctioning and positioning especially with more severe CNS dysfunction
- Continue anticonvulsive therapy
- May require PRBC and/or FFP transfusion for continued bleeding

Anticipated Problems/Concerns
- Intraoperative hemorrhage with the potential for continued hemorrhage in the postop period
- Respiratory failure and pulmonary complications in pts with lung involvement
- Aspiration
- Seizures

Kartagener Syndrome

Nancy C. Wilkes

Risk
- KS, first described in 1933, is part of a larger family of diseases classified as PCD.
- The triad of KS consists of bronchiectasis, chronic sinusitis, and situs inversus; it has an incidence estimated at 1:15,000–40,000 births.
- The disease is likely underdiagnosed because a limited amount of centers have resources to provide an accurate diagnosis.
- No known predilection for race or gender.
- Symptoms more prevalent in children in the first decade of life.

Perioperative Risks
- Morbidity: Lung infection, pulm edema, atelectasis, sinusitis

Worry About
- Pulm function and anatomy.
- Airway obstruction due to ineffective clearance of secretions.
- Bronchiectasis, which can lead to cor pulmonale, amyloidosis, and pulm edema and is usually found in the middle or lower lobes in KS pts, as opposed to the upper lobes in cystic fibrosis pts.
- Chronic disease with variable onset.
- Chemical injury from aspiration in left lung, which is the larger lung in pts with KS.
- Unintended bronchial intubation with single-lumen ETT, resulting in nonventilation of right lung (in those with pulm inversion).
- Left-sided double-lumen tubes may occlude orifice of left upper lobe.
- Nasal catheters relatively contraindicated because of risk of paranasal sinusitis and ear infections.
- Increased susceptibility to overall infection due to impaired neutrophil chemotaxis.

Overview
- Complete situs inversus (including dextrocardia).
- PCD resulting in chronic respiratory tract infections, bronchiectasis, and sinusitis.
- Approximately half of patients with PCD have situs inversus and thus are classified as having KS.

Etiology
- Congenital defect in synthesis of various parts of cilia (dynein arms, radial spokes, nexin links, microtubules) that results in abnormal/dyskinetic ciliary movement.
- Ciliated epithelium covers most areas of the upper resp tract, including the nasal mucosa, paranasal sinuses, middle ear, eustachian tube, and pharynx. The lower resp tract contains ciliated epithelium from the trachea to the resp bronchioles.
- Autosomal recessive inheritance pattern; genetically heterogeneous with multiple chromosomes likely responsible for phenotype.

Usual Treatment
- Aerosol administration to reduce secretion viscosity
- Antimicrobial therapy for chronic resp tract infections and sinusitis
- Surgical intervention for persistent bronchiectasis
- Conventional and assisted airway clearance techniques (chest physiotherapy, PEP mask, forced oscillation techniques, exercise programs, and physical activity)
- Nasal steroid sprays
- Inhaled bronchodilators and anti-inflammatory medications to treat bronchospasm

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Dextrocardia		Right-sided heart tones	CXR, ECHO, ECG
RESP	Bronchiectasis Ciliary dyskinesia	Dyspnea Cough Halitosis	Decreased breath sounds, rhonchi, crackles, wheezes	CXR, bronchoscopy, spirometry, bronchography
IMMUNO	Chronic pansinusitis, nasal polyposis Bronchitis	Nasal drainage, morning sore throat Cough, mucus production	Frontal and maxillary tenderness Rhonchi	CT sinuses Sputum and tracheal aspirate for culture and Gram stain
	Pneumonia Otitis media	Cough, fever Earache, hearing loss	Rales, rhonchi Erythematous tympanic membrane	CXR, SpO_2 Audiometry, tympanotomy

Key References: Leigh MW, Pittman JE, Carson JL, et al.: Clinical and genetic aspects of primary ciliary dyskinesia/Kartagener syndrome, *Genet Med* 11(7):473–487, 2009; Mathew PJ, Sadera GS, Sharafuddin S, et al.: Anaesthetic considerations in Kartagener's syndrome–a case report, *Acta Anaesthesiol Scand* 48(4):518–520, 2004.

Perioperative Implications

Preinduction
- Consider omitting anticholinergics and cough suppressants from preanesthetic medication.
- Chest physiotherapy, bronchodilators, and incentive spirometry are often beneficial.
- Treat underlying pulm infections.
- Immunize against influenza A and pneumococcal organisms.

Monitoring
- In dextrocardia, position of ECG leads should be the mirror image of normal, as should that of paddles of external defibrillation, cardioversion, and pacing.
- Because the great vessels and thoracic duct are likely to be reversed, consider cannulation of the internal jugular vein from the left.
- Pulm artery cath should be oriented in anticipation of a clockwise direction of migration.
- Pregnant pts with KS should be positioned in right uterine displacement rather than left.

Induction and General Anesthesia
- Emphasize aseptic technique secondary to abnormal neutrophil chemotaxis.
- Aim for nontraumatic airway manipulation to avoid possible infection.
- Humidify inspired gases.
- Inhalation injury usually occurs in left lung, which is also larger lung.
- Bronchial intubation with a single-lumen ETT usually involves left side.
- Right bronchial suctioning will be more difficult to perform with nonangulated suction cath.
- Left-sided double-lumen tubes may occlude orifice of left upper lobe.
- When lung isolation is needed, consider tracheal intubation first with a bronchial blocker in the appropriate bronchus.
- If a double-lumen tube is required, consider inserting a left-sided tube with the bronchial tube on the right; the endobronchial stylet and the upper part of tube must be bent 180 degrees from original orientation prior to insertion such that the normal curvature of the oropharynx is still followed. The same principles apply to use of a right-sided tube.
- Extubation of the trachea should occur as soon as possible after the patient meets common extubation criteria.

Regional Anesthesia
- Use regional or local anesthetic techniques when possible to avoid airway manipulation and complications and to preserve resp muscle function intraop and postop.

Postoperative Period
- Consider nonnarcotic analgesia and/or epidural analgesia for postop pain.
- Avoid excessive sedation and encourage early ambulation to aid in clearance of airway secretions
- Chest physiotherapy, bronchodilators, and incentive spirometry may be beneficial.
- Oral airway preferred over nasal airway secondary to increased risk of sinusitis.

Anticipated Problems/Concerns
- Lung infection is common as result of ciliary dyskinesia.
- Fluid overload can precipitate cor pulmonale and pulm edema.
- Avoid nasal cath and/or airways to minimize chances of paranasal sinusitis.

Kawasaki Disease

Kumaran Senthil | Todd J. Kilbaugh

Risk
- Incidence in USA: 20 hospitalizations per 100,000 children
- Most commonly in pts <3 y of age
- Asian and Pacific Islanders have a higher rate, implying an unknown genetic effect
- Infants <6 mo and children >5 y have a higher risk of developing coronary artery lesions

Perioperative Risks
- Risk of myocarditis, valvular disease, pericardial effusions, and arrhythmias during the acute phase, defined as within 2 wk of fever onset.
- Coronary artery thromboembolic events from coronary artery aneurysm, stenosis, or obliteration can develop subacutely, usually within 6 wk of fever onset, and can become chronic concerns.

Worry About
- Acute coronary syndrome in pts with history of KD with coronary artery pathology
- Diminished left ventricular ejection fraction in 20% of cases during acute phase
- Oral mucous membrane inflammation during acute phase
- Vomiting and abdominal pain with risk of aspiration
- Aseptic meningitis from KD or as a potential side effect from IVIG therapy

Overview
- Acute febrile illness characterized by medium vessel vasculitis that mimics an infectious process, with the potential risk of myocarditis, pericardial effusions, and arrhythmias in the acute phase and development of coronary artery aneurysms subacutely.
- KD is clinically diagnosed by at minimum of 5 days of high fever and with at least four of the following criteria, or fewer if coronary artery lesions are present:
  - Swelling of hands and feet or redness of palms and soles.
  - Polymorphous rash.
  - Bilateral limbic sparing conjunctival injection.
  - Strawberry tongue, cracked lips, or erythematous oropharynx.
  - Cervical adenopathy with greatest node ≥1.5 cm in diameter.
- Atypical KD does not meet all of the clinical criteria for a complete diagnosis but meets some, with additional lab findings, such as elevated liver enzymes, decreased albumin, anemia, or sterile pyuria.
  - Atypical KD is more common in infants and older children and therefore is associated with greater risk for development of coronary artery lesions.
- Treated with high-dose IVIG and aspirin, preferably before day 10 of fever to limit coronary pathology.
- Risk of developing coronary artery lesions is approximately 3–5% of those treated with IVIG and 25% of those untreated.

Etiology
- No known etiology.
- Theories include viral illness, toxin-mediated process, or infectious trigger leading to vasculitis in predisposed pts.

Usual Treatment
- High-dose IVIG and high-dose aspirin within 10 d of illness to minimize risk of coronary artery lesions.
- If fevers recur, most clinicians consider another course of IVIG after 48 h from initial administration.
- Japanese data supports use of corticosteroids, but no supporting evidence in USA populations.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Conjunctival Injection Oral/mucosal inflammation Cervical adenopathy	Pain	Conjunctivitis, but sparing the limbus Strawberry tongue, cracked lips	
RESP	Pleural effusions Pulmonary nodules or interstitial infiltrates	Dyspnea	Tachypnea, dullness to percussion Dry crackles	CXR, US CT scan if other imaging is insufficient
CV	Congestive heart failure via myocarditis, pericardial effusions Valvular abnormalities, arrhythmias Coronary artery abnormalities Medium vessel noncoronary aneurysms	Exertional dyspnea, chest pain, abdominal pain Exertional chest pain	Tachycardia, S_3, distant heart sounds, hepatomegaly Tachycardia, systolic or diastolic murmur Palpable pulsatile masses	ECG, CXR, ECHO ECG, Holter monitoring, echocardiography ECHO, coronary angiography US
GI	Hydrops of the gallbladder	N/V, diarrhea, jaundice	RUQ tenderness, scleral icterus, hepatomegaly	RUQ ultrasound, LFT, GGT, bilirubin
CNS	Aseptic meningitis Anterior uveitis	Irritability, lethargy, headache Visual changes	Meningismus, photophobia, phonophobia	Lumbar puncture Slit lamp evaluation
HEME	Anemia Thrombocytosis (after first wk)	Fatigue	Pallor	CBC, reticulocyte count
DERM	Polymorphous exanthem		Starts as perineal desquamation, progresses to diffuse, erythematous, maculopapular	

Key References: Son MF, Newburger JW: Kawasaki disease, *Pediatr Rev* 34(4):151–162, 2013; Morrison JE, Anderson M, Chan KC, et al.: A 15-year review of children with Kawasaki's Syndrome having general anesthesia or deep sedation, *Paediatr Anaesth* 15(12):1053–1058, 2005.

Perioperative Implications

Preoperative Preparation
- If no diagnosis of undiagnosed protracted fever with rash, consider diagnosis of KD and potentially delay nonemergent cases until adequate cardiac evaluation.
- Assess cardiac status.
- In acute phase, concern for myocarditis and pericardial effusion with potential for development of acute heart failure.
- For pts with history of KD, assess coronary status and determine risk of developing myocardial ischemia.
- Assess need for rapid sequence intubation if active nausea or vomiting or risk for aspiration.

Monitoring
- Arterial line if indicated; insert with ultrasound guidance to evaluate for possible noncoronary arterial aneurysms.