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.

- ECG with lead II and V5 monitoring to assess for ST segment changes.
- Consider attaching defibrillator/cardioversion pads if Hx of significant arrhythmias.

Airway
- Consider rapid sequence intubation for risk of aspiration.
- Assess for difficult airway secondary to friable oral and pharyngeal mucosal surfaces.

Preinduction and Induction
- If compromised cardiac status, induction with minimal alterations in afterload and preload.
- If known severe CAD, maintain afterload to preserve coronary perfusion pressure.
- If clinically significant pleural effusions, expect loss of functional residual capacity and faster desaturation.

Maintenance
- Avoid fluid overload for pts with depressed cardiac function.
- Avoid hypotension in pts with significant coronary disease.
- In acute phase, highly febrile pts may have a greater anesthetic need and insensible fluid loss.

Extubation
- Period with greatest myocardial oxygen consumption and vigilance for cardiac decompensation

Postoperative Period
- Continued ECG for myocardial ischemia or signs of heart failure.
- Consider perioperative troponin trending if concern for subclinical myocardial ischemia based on exam or ECG findings.

Anticipated Problems/Concerns
- If acute myocardial ischemia from coronary thromboembolic event, consider cardiac cath with angioplasty and stenting.
- If CHF develops in the setting of fluid overload or evidence of myocardial ischemia, consider transport to ICU for continued monitoring and treatment.

Klippel-Feil Syndrome

Ronald S. Litman | Lee A. Fleisher

Risk
- Incidence estimated at 1:40,000 live births (but milder cases go unrecognized).
- Slight female predilection (63%).

Perioperative Risks
- Cervical spine instability and cardiopulmonary complications.
- Often occurs in association with other clinical syndromes (e.g., fetal alcohol, Goldenhar).

Worry About
- Exacerbation of cervical spine instability during airway maneuvers, endotracheal intubation, and subsequent positioning.

Overview
- Congenital abnormality consisting of the following triad of findings: Fusion of two or more cervical vertebrae, low posterior hairline, cervical immobility.
- Type 1: Extensive fusion of many cervical vertebrae; type 2: Fusion at only one or two cervical interspaces; type 3: Fusion in the cervical spine and in the lower lumbar spine.
- Severity ranges from mild (often not recognized until late in life) to severe (recognized at birth because of obvious deformity).
- Careful preop assessment of cervical spine anatomy and degree of instability.
- Review of systems for other congenital abnormalities: Renal dysfunction (64%), scoliosis (60%), deafness (30%), Sprengel scapular deformity (25–35%), congenital heart disease (4.2–14%), mental deficiency, pulmonary disability, and cleft lip and palate.

Etiology
- Unknown

Usual Treatment
- Symptomatic; depends on organ system involvement

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Head and neck immobility		Decreased ROM of cervical spine, low posterior hairline, webbed neck, facial asymmetry, cleft palate, torticollis, vocal cord dysfunction	Flexion/extension radiographs of cervical spine Consider MRI of cervical spine
CV	Bradyarrhythmias and AV conduction pathway abn (due to CNS malformations) Cardiac defects (most commonly VSD)	Syncope	Murmurs	ECG, ECHO
RESP	Central alveolar hypoventilation, pulmonary agenesis or hypoplasia, restrictive lung disease (due to severe scoliosis)	Sleep apnea, snoring, difficulty breathing		ABG CXR (if symptomatic)
RENAL	Urinary tract abn, renal agenesis, ureteral duplication			BUN, Cr if indicated, renal US
CNS	Hindbrain abnormality (e.g., syringomyelia, Arnold-Chiari malformation) Mental retardation, deafness, strabismus	Peripheral neurologic dysfunction (e.g., weakness, paresthesias, paraplegia, quadriplegia)	Neurologic exam	
MS	Scoliosis, Sprengel deformity (scapular elevation), hypermobility of cervical spine, spondylosis/decreased mobility of cervical spine		Exam of spine and shoulders	X-rays if indicated

Key References: Stallmer ML, Vanaharam V, Mashour GA: Congenital cervical spine fusion and airway management: a case series of Klippel-Feil syndrome, *J Clin Anesth* 20(6):447–451, 2008; Hase Y, Kamekura N, Fujisawa T, et al: Repeated anesthetic management for a patient with Klippel-Feil syndrome, *Anesth Prog* 61(3):103–106, 2014.

Perioperative Implications

Preoperative Preparation
- Careful and complete evaluation of cervical spine anatomy and instability and of other major organ system abnormalities

Monitoring
- Depends on pt's physical condition

Airway
- If indicated, awake intubation using maneuvers to stabilize cervical spine; complete immobility with use of fiberoptic intubating bronchoscope ideal

Preinduction/Induction
- Depends on pt's physical condition

Maintenance
- Careful positioning of head and neck with maintenance in neutral position

Extubation
- Depends on extent of cervical spine pathology and respiratory compromise

Adjuvants
- No special considerations

Anticipated Problems/Concerns
- Exacerbation of preexisting cervical spine instability leading to neurologic deterioration