- 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

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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