

Monitoring

- Monitor for myocardial ischemia; can have CHF if volume overload and LV dysfunction present.
- Monitor blood sugar if on steroids.

Airway

- Due to impaired motility, swallowing dysfunction, and aspiration risk, many recommend regional anesthesia and only instrumentation of airway with awake fiberoptic techniques.
- Little information on the appropriate use of muscle relaxants. It is suspected that pts with dermatomyositis are sensitive to nondepolarizing muscle relaxants because of their diminished muscle mass. Muscle relaxants given with close monitoring.

Intraoperative Period

- If ventilatory status is marginal preop, then control ventilation. Airway protection and adequate ventilation are the two primary concerns.

Induction

- High incidence of swallowing and vocal cord dysfunction in these pts may lead to pooling of saliva in the pharynx and aspiration into the trachea.

Maintenance

- CV instability
- Narcotics used with caution to prevent any postop resp dysfunction

Extubation

- CV and pulm drive insufficiencies common with myopathies

Adjuvants

- No known adjuvant concerns except those that weaken muscular function.

Postoperative Period

- Respiratory insufficiency is the major postop complication. Due to the weakness of thoracic muscles,

pts may have a diminished cough reflex, leaving them vulnerable to atelectasis.

- Weakness of pharyngeal muscles may make pts more vulnerable to aspiration pneumonia.
- A titrated analgesic regime or regional block for pain relief has been used in most of the anecdotal case reports.

Anticipated Problems/Concerns

- Increased risk of infections such as digestive and respiratory infections

Dextrocardia

Glyn D. Williams

Risk

- Birth prevalence of approximately 1:10,000; equal sex distribution
- 25% have PCD, an autosomal recessive disorder

Perioperative Risks

- Increased risk of cardiac decompensation, pulm Htn, resp failure, airway obstruction, sepsis, raised intracranial pressure, and death
- Increased likelihood of emergent open-heart or abdominal surgery

Worry About

- Heterotaxy syndrome (approximately 40% have dextrocardia)
- PCD (approximately 50% have dextrocardia)
- Distinguish from dextroposition—right cardiac displacement by extracardiac causes (lung, diaphragm, pericardium abnormalities)

Overview

- Dextrocardia results from embryologic anomalies. The heart is positioned in the right hemithorax, with base and apex of heart pointing caudally and to the right.

- Mirror-image dextrocardia can be asymptomatic incidental finding.
- PCD: Associated with:
 - Middle ear infections.
 - Paranasal sinusitis.
 - Lung disease (bronchiectasis, pneumonia).
 - Infertility.
 - Hydrocephalus.
 - Retinitis pigmentosa.
 - Situs inversus totalis.
 - Heterotaxy.
- Heterotaxy (1:6000 live births) is failure of usual R-L asymmetry: Associated with:
 - Congenital heart disease (many variants).
 - Brain (e.g., encephalocele).
 - Skeletal (e.g., spine deformities).
 - Facial (e.g., micrognathia).
 - Resp (e.g., tracheoesophageal fistula, PCD).
 - Gut (e.g., duodenal atresia, volvulus).
 - Pancreatic and liver hypoplasia.
 - GU tract (e.g., renal agenesis).
 - Other (e.g., diaphragmatic hernia).

Etiology

- The human fetal heart develops from a primitive cardiac tube, with sinus venosus, atrium, ventricle, bulbus cordis, and truncus arteriosus connected in series. With growth, the tube loops right or left. Dextrocardia can occur with abnormal looping.
- Heterotaxy and PCD result from abnormal structure and function of motile cilia. The ventral node, a transient midline structure present in early fetal life, has specialized monocilia that generate unidirectional extraembryonic fluid flow, which initiates normal R-L asymmetry. Abnormal flow leads to heterotaxy.

Usual Treatment

- PCD: Supportive pulmonary therapies, sinus surgery
- Heterotaxy: Surgical repair of congenital heart disease and medical management of cardiac failure, arrhythmia, antibiotic prophylaxis if immunocompromised

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Difficult airway Aspiration risk	Snores Gags with feeding	Micrognathia, cleft palate Ear and sinus infection	CT sinuses CBC, differential
RESP	Pneumonia Bronchiectasis Poor secretion clearance	Dyspnea Chronic cough	Tachypnea Lung field consolidation Wheezing	CXR, CT scan PFTs, bronchoscopy
CV	One or two ventricle physiology Arrhythmia, heart failure Pulm Htn	Dyspnea Blue Syncope	Exercise intolerance Cardiac failure Cyanotic heart disease	ECG, Holter, CXR, TTE, MRI, CT, heart cath, SpO ₂
GI	Obstruction, short gut syndrome, GERD, biliary atresia, pancreatitis	N/V, pain, distension Heartburn Yellow	Acute abdomen Hypoactive bowel sounds Jaundice	Abdominal x-ray, LFT Lytes, amylase Endoscopy
RENAL	GU anomalies	Urinary infections	Posterior urethral valves Hypospadias	BUN/Cr, lytes, CBC, US, MRI
CNS	Neurologic anomalies ICP high, meningitis	Irritability, lethargy Headache, seizures	Meningismus Papilledema	Lumbar puncture, head CT, MRI
MS	Sternum, spine, limb defects	Difficulty walking	Skeletal exam	X-ray
IMMUNE	Immunocompromised (asplenia/polysplenia)	Recurrent infections	Signs of infection	Ultrasound, MRI Immunology work-up

Key References: Rapoport Y, Fox CJ, Khade P, et al.: Perioperative implications and management of dextrocardia, *J Anesth* 29(5):769–785, 2015; Lobo LJ, Zariwala MA, Noone PG: Primary ciliary dyskinesia, *QJM* 107(9):691–699, 2014.

Perioperative Implications**Perioperative Preparation**

- Congenital heart disease: Consult experts.
- PCD: Optimize pulmonary status.
- Immunocompromised: Choice of antibiotics.
- Consider reprogramming AICD/pacemaker.

Monitoring

- ECG, defibrillator: Reverse positions
- Heterotaxy: TEE, invasive hemodynamic monitoring
- Central lines: May have abnormal anatomy

Airway

- Possible difficult mask, difficult intubation.
- Avoid nasal intubation (sinusitis).

Induction

- Congenital heart disease concerns

Maintenance

- Humidify gases (PCD)
- Congenital heart disease concerns

Extubation

- Assess cardiopulmonary reserve.

Adjuvants

- Nitric oxide if pulm Htn

Postoperative Period

- Consider ICU if PCD or heterotaxy.

Anticipated Problems/Concerns

- Dextrocardia provides periop challenges ranging from minor (unusual ECG finding) to severe (life-threatening crisis). Pts with PCD may require additional pulmonary support during the periop period. Heterotaxy pts have high periop risk. Input from appropriate experts is recommended.

Diabetes, Type I (Insulin-Dependent)

Michael F. Roizen

Risk

- Incidence in USA: 1.25 million.

Perioperative Risks

- Risk of requiring a CABG is increased 5–10 times in presence of ESRD, CHF, or autonomic neuropathy; without these conditions, the risk is 1-1½ times that of a normal person.

Worry About

- Autonomic neuropathy, gastroparesis, and sudden postop death.
- Painless myocardial ischemia.
- Atlantooccipital joint immobility.
- Tight glucose control might be indicated in pregnant pts and those difficult to wean from bypass (in ECC as well as in the case of predictable global or focal CNS ischemia).

Overview

- Endocrinopathy assoc with ESRD or ophthalmic, myocardial, and neuropathic disease

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Possible atlantooccipital dislocation secondary to abnormal collagen glycosylation	Pain	Neck ROM, "prayer sign"	Usually not needed; x-rays of neck in extension
CV	Angiopathy, LV dysfunction (increased 4–10 times with Htn) Ischemic PVD	Poor exercise tolerance Angina, CHF symptoms	Two-flight climb Chest exam for signs of CHF BP lying and standing	ECG, CXR, coronary calcium score if indicated
RESP	Decreased lung elastance; decreased FEV ₁ ; decreased FVC	Poor exercise tolerance		Generally not needed
GI	Gastroparesis	Early satiety		
RENAL	Nephropathy, especially with Htn	N/V, impotence; orthostatic symptoms Nonprotein foods		BUN/Cr
ENDO	Decreased insulin from islets			FBS, lytes
CNS	Autonomic dysfunction secondary to neuropathy	Early satiety, impotence, N/V, orthostatic symptoms		HRV on ECG BP change on standing
PNS	Stocking-glove neuropathy leading to infections		PNS exam, especially if regional anesthesia is planned	
MS	Impaired joint mobility secondary to nonenzymatic glycosylation of collagen	Joint mobility	Decreased ROM of joints	

Key References: Daneman D: Type 1 diabetes. *Lancet* 367(9513):847–858, 2006; Preiser JC, Devos P, Ruiz-Santana S, et al.: A prospective randomised multi-centre controlled trial on tight glucose control by intensive insulin therapy in adult intensive care units: the Glucontrol study. *Intensive Care Med* 35(10):1738–1748, 2009.

Perioperative Implications**Preoperative Preparation**

- Metoclopramide (10 mg/70 kg) in pts with gastroparesis.
- Assess myocardial and volume status.

Monitoring

- Myocardial ischemia can indicate CHF if volume overload and LV dysfunction are present.
- Blood sugar.

Airway

- Atlantooccipital dislocation possible; see HEENT. Do prayer sign test; pt may have gastroparesis.

Induction

- Osmotic diuresis can lead to hypovolemia; ANS and CV dysfunction cause fluctuations in BP and HR.

Maintenance

- CV instability; volume status is key to avoiding renal and myocardial dysfunction with surgery; checking

RR variation (HRV) to determine autonomic insufficiency likelihood still not widespread.

Extubation

- CV and pulm drive insufficiencies common with neuropathies.

Adjuvants

- Rx for tight control
- Regional: Diabetic nerves may be more prone to edema, especially if epinephrine has been used.