- Benzodiazepine (midazolam) may be used to treat preop anxiety.
- Ketamine, a sympathetic stimulant, should be avoided.
- Serum cholinesterase activity may be decreased in pts on phenelzine, so the dose of succinylcholine may need to be reduced.
- The addition of epinephrine to local anesthetic solutions should probably be avoided.
- If hypotension develops, direct-acting drugs, such as phenylephrine, are preferred. The dose should also be decreased to minimize the likelihood of an exaggerated hypertensive response.

Anticipated Problems/Concerns

- In periop period, general rule is to try to continue antidepressant therapy.
- Be aware of potential interactions between anesthetic agents and antidepressants.
- Pts should be monitored for signs of serotonin syndrome.

Acknowledgment

I wish to thank Dr. Ian Yuan for his work on this chapter in the earlier edition of this book.

Dermatomyositis

Michael F. Roizen | Jeffrey D. Roizen

Risk

- Prevalence in USA: 3000–10,000.
- Group, demographics with highest prevalence include females, 2:1 relative to men, with a peak onset between 30–60 y of age.

Perioperative Risks

- Increased risk of respiratory failure and infections postop

Worry About

- Most case reports absolutely avoid depolarizing muscle relaxants and are careful with medications that have effects on muscle strength.
- Monitor muscle relaxant dosing and recovery.
- Valvular heart disease and cardiomyopathy: Cardiac muscle, though not severely involved, shows changes similar to skeletal muscles. Clinical manifestations are rare.

Overview

- Relatively rare diffuse connective tissue disorder of uncertain etiology characterized by idiopathic inflammatory myopathy with muscle involvement and weakness muscle and connective tissue involvement of skin and other organs. Valvular heart disease increases the risk of periop adverse cardiac events.
- Diagnosis is based on the clinical picture of muscle weakness and skin rash, myelography, raised serum CPK levels, and muscle biopsy.
- Respiratory system: Aspiration pneumonia may occur due to weakness of the muscle involved in swallowing. Progressive weakness of the intercostal and diaphragmatic muscles may result in respiratory insufficiency. Lung involvement may occur from the connective tissue disorder itself, which results in patchy infiltrates throughout both lungs, interstitial pneumonia, or fibrosis. Carcinoma of the bronchus or lung parenchyma is associated.

Etiology

- The lead theory regarding dermatomyositis involves a genetic predisposition to viral or immune destruction of muscles by viruses or other infectious agents. Dermatomyositis is considered a connective tissue diseases in the same category as lupus erythematosus or systemic sclerosis.

Usual Treatment

- Prednisone to control weakness and pain
- Various agents to control rash and calcinosis in skin

Assessment Points

System	Effect	Assessment by Hx	PE	Test
DERM	Characteristic rash Raynaud phenomenon	Treatment with diltiazem or colchicine to reduce calcinosis Hydroxychloroquine may reduce the photosensitive rash Raynaud phenomenon may present.	Classic purple rash on eyelids and over bony prominences Children's skin can become thick and hard; rash appears on the back, knuckles, chest, shoulders, neck, and face.	Biopsy in past—look at results
HEENT	Possible regurgitation and swallowing difficulties	Symptoms of regurgitation	Test of swallowing with water	Usually not needed, neck x-rays in extension; GI swallow for motility
CV	Valvular heart disease Cardiac muscle, though not severely involved, shows changes similar to skeletal muscles. Clinical manifestations are rare, but heart failure and conduction defects reported.	Poor exercise tolerance Angina CHF symptoms	Two-flight walk Chest exam for signs of CHF BP lying and standing	ECG, ECHO for valvular disease
RESP	Decreased lung elastance; decreased FEV; decreased FVC Aspiration pneumonia due to weakness of the muscle involved in swallowing Potential progressive weakness of the intercostal and diaphragmatic muscles results in respiratory insufficiency.	Poor exercise tolerance		Generally not needed
GI	Esophageal motility disorders, gastroparesis, GI ulcers and infections	Early satiety		
RENAL	Nephropathy, if treatment for many years			BUN/Cr
ENDO	Insulin resistance from high dose prednisone treatment			FBS, lytes
CNS	Fatigue and weakness	Early satiety, impotence, N/V, orthostatic symptoms		Changes related to degree of type 2 diabetes from therapy
PNS	Proximal muscle weakness	Shoulder-girdle weakness	PNS exam, esp. if regional planned, which is recommended by most case reports	Abnormal muscle biopsy and MRI of proximal muscles
MS	Impaired mobility and strength	Muscle strength	Weakness, inability to get out of chair by self, decreased ROM of joints	Elevated muscle enzyme levels

Key References: Gunusen I, Karaman S, Nemli S, Firat V: Anesthetic management for cesarean delivery in a pregnant woman with polymyositis: a case report and review of literature, *Cases J* 2:9107, 2009; Shrestha GS, Aryal D: Anaesthetic management of a patient with dermatomyositis and valvular heart disease, *Kathmandu Univ Med J* 10(38):100–102, 2012.

Perioperative Implications

- May exhibit signs of some paraneoplastic disorder (e.g., polyneuropathy, subacute cerebellar degeneration, multifocal neuroencephalopathy, myasthenic syndrome).
- If on steroids for treatment, may benefit from periop steroids.
- If given cytotoxic drugs, hematologic status needs examination.

Preoperative Preparation

- Administer metoclopramide (10 mg/70 kg) in pts with esophageal motility problem or gastroparesis.
- Assess myocardial and volume status.

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.