

Usual Treatment

- Cardiac surgery
- Vitamin D (cholecalciferol and calcitriol), calcium supplementation

- Parathyroid hormone therapy
- BMT or thymic grafts (complete DiGeorge syndrome)

- Irradiated transfusion products
- IV Ig therapy
- Antibiotic prophylaxis

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CNS	Language and motor developmental delay	Failure to meet milestones	Speech or language delay	Physical exam
HEENT	Palatal laryngotracheal anomalies Facial dysmorphism	Difficulty feeding Nasal regurgitation	Cleft palate Hypotonia Hypernasal speech, micrognathia	CT Barium swallow study
CV	Conotruncal cardiac defects(interrupted aortic arch, truncus arteriosus, TOF, ASD, VSD, vascular rings)	Failure to thrive Cyanosis Dyspnea	Cyanosis, heart murmur, dyspnea, dysphagia	ECHO, CT
RESP	Asthma (atopic)	Dyspnea	Wheezing	PFT
MS	Scoliosis Rheumatoid arthritis	Asymmetric spine, painful joints	Asymmetry of spine, joint inflammation	Radiographs
HEME	Hypoplastic/aplastic thymus Immunodeficiency Severe combined immunodeficiency Autoimmune disease	Recurrent URIs, otitis media, opportunistic infections Thyroiditis Rheumatoid arthritis Recurrent bleeding	Symptoms of PNA, otitis media, sinus infections or severe immunodeficiency Symptoms of hypothyroid/hyperthyroid Symmetric degenerative joint disease	CXR Ig levels: Increased IgE, decreased IgA, decreased CD3+ Decreased or increased TSH, T3, T4 X-ray of affected joints CBC: Decreased platelets
ENDO	Hypocalcemia	Stiffness or twitching	Tetany	Increased phosphorus, decreased Ca ²⁺ Decreased PTH

Key References: Seroogy CM: DiGeorge (22q11.2 deletion) syndrome: clinical features and diagnosis. Stiehm ER, TePas E, editors. Waltham, MA, 2015, UpToDate; Hauk PJ, Johnston RB, Liu AH, et al.: Immunodeficiency. In Hay WW Jr, Levin MJ, Detering RR, et al, editors: *Current diagnosis & treatment: pediatrics*, ed 22, New York, NY, 2013, McGraw-Hill.

Perioperative Management

Preoperative Considerations

- BMP, Ca²⁺, Phos, CBC, CD3+ count
- Type and cross irradiated blood products prn
- Review imaging and cardiac studies
- Reverse isolation precautions prn

Monitoring

- Standard ASA monitors.
- Arterial, central line prn.
- Consider preop calcitriol and intraop Ca²⁺, as well as phosphate; premedicating with calcitriol and calcium can prevent intraop hypocalcemia.

General Anesthesia

- All IV, arterial, and central access placed under sterile technique
- Anticipate difficult mask/intubation scenario.

Regional Anesthesia

- Difficult neuraxial anesthesia placement due to scoliosis.
- Caution in pts with thrombocytopenia.
- Consider increased risk for developing infection at site of injection.

Postoperative Period

- Poor wound healing.

- Increased infection risk.
- Continue to monitor lytes; stress can precipitate a hypocalcemic crisis.

Anticipated Problems/Concerns

- High infection risk
- Lyte imbalances
- Airway/facial anomalies necessitating FO or video laryngoscopy
- Cardiac defects with shunting lesions

Dilated Cardiomyopathy

Frank W. Dupont

Risk

- DCM is a largely irreversible form of heart muscle disease, with an estimated prevalence of 1:2500; it is the third most common cause of CHF and most frequent cause for heart transplantation.
- DCM leads to progressive CHF, ventricular and supraventricular arrhythmias, conduction system abnormalities, thromboembolism, and sudden or heart failure–related death.
- Marked limitation of exercise capacity is a reliable predictor of mortality.

Perioperative Risks

- Increased periop morbidity and mortality, particularly in high-risk surgery cases:
 - CHF exacerbation
 - Renal failure

- Systemic or pulm embolization from dislodged intracardiac thrombi

Worry About

- Autonomic instability
- Malignant tachyarrhythmias
- Worsening LV systolic and/or diastolic function, RV dysfunction

Overview

- Syndrome characterized by dilatation and impaired systolic function of left, right, or both ventricles with normal ventricular wall thickness
- LV systolic (decreased EF) and diastolic dysfunction (noncompliant ventricle), RV dysfunction; possibly pulm Htn and AV valvular regurgitation
- High risk of sudden cardiac death

Etiology

- Cause of idiopathic DCM remains unclear, but several pathophysiologic mechanisms have been implicated: genetic and familial factors, inflammatory and infectious factors, cytotoxicity, cell loss, and abnormalities in endogenous repair.

Usual Treatment

- Medical interventions primarily based on CHF treatment with diuretics, ACEI, ARB, vasodilators, and β-adrenergic receptor–blocking agents; anti-coagulants for thromboembolic prophylaxis; ICD implantation for management of tachyarrhythmias and CRT for dyssynchrony
- Surgical treatment for refractory end-stage CHF: LVAD placement, heart transplant

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Arrhythmias CHF Myocardial ischemia	Palpitations DOE Orthopnea PND Angina	Narrow pulse pressure, pulsus alternans Displaced PMI Systolic murmur (MR), S ₃ , S ₄ JVD, ascites, pedal edema	ECG, EPS ECHO Stress test Coronary angiography
RESP	Pulm edema	Dyspnea	Rales, wheezes	CXR ABG
GI	Hepatic congestion	Abdominal distension	Hepatomegaly	LFTs, PT, albumin
HEME	Coagulopathy	Bruising		PT/PTT
RENAL	Renal insufficiency	Oliguria		BUN/Cr, FEN _a
CNS	Cerebral infarcts	Stroke	Focal neurologic deficits	CT, MRI

Key References: Maron BJ, Towbin JA, Thiene G, et al.: Contemporary definitions and classification of the cardiomyopathies: an American Heart Association Scientific Statement from the Council on clinical cardiology, heart failure and transplantation committee; quality of care and outcomes research and functional genomics and translational biology interdisciplinary working groups; and Council on epidemiology and prevention, *Circulation* 113(14):1807–1816, 2006; Sumler ML, Andritsos MJ, Blank RS: Anesthetic management of the patient with dilated cardiomyopathy undergoing pulmonary resection surgery: a case-based discussion, *Semin Cardiothorac Vasc Anesth* 17(1):9–27, 2013.

Perioperative Implications

Preoperative Preparation

- Consider cardiology consultation to optimize pt's cardiac condition.

Monitoring

- ECG with ST-segment analysis.
- Arterial line dependent on invasiveness of surgery.
- Consider PA cath if anticipation of large fluid shifts in moderate- to high-risk surgery.
- TEE is the monitor of choice for the assessment of biventricular function and AV valve regurgitation in invasive surgical cases.

Airway

- None

Preinduction and Induction

- Anesthetic principles are based on afterload reduction, preload conservation, and prevention of tachycardia and myocardial depression.

Maintenance

- Higher doses of volatile anesthetic agents are often poorly tolerated; thus a narcotic-based anesthetic with low-dose volatile agents and/or benzodiazepine supplementation may be preferable.
- Fluid management should be conservative to prevent fluid overload and acute CHF exacerbation.
- Inotropic support may be necessary.

Extubation

- Beware of tachycardia and Htn and treat proactively.

Postoperative Period

- Consider ICU admission and mechanical ventilation if major intraop fluid shifts have occurred.

Adjuvants

- Regional anesthesia techniques are not contraindicated in the absence of coagulopathy and provided that hypotension is prevented.
- ICD management precautions should be taken if applicable.

- DCM predisposes to decreased blood flow to liver and kidney, which prolongs action of many drugs; also increased volume of distribution requires drug dose adjustments.

Anticipated Problems/Concerns

- CHF exacerbation, hemodynamic instability, tachyarrhythmias

Diphtheria

Pierre Moine

Risk

- Approximately 0.001 cases per 100,000 population in USA since 1980 (<5 cases a year).
- Endemic in developing countries.
- Still common in countries where mass immunization programs are not enforced.
- After political changes in Eastern Europe and Central Asia at the end of the 20th century, a resurgence in many vaccine-preventable diseases, including diphtheria, was reported across these countries.
- Risk factors for diphtheria outbreaks: older age (they are not up to date with booster immunization against diphtheria), lack of vaccination, alcoholism, low socioeconomic status, crowded living conditions, and Native American background.

Perioperative Risks

- Early (days after exposure): Respiratory compromise; respiratory arrest; airway obstruction and hemorrhage; conduction abnormalities, dysrhythmia, cardiogenic shock, CHF, myocarditis; shock, coma, and death
- Late (2–6 wk): Myocarditis and polyneuritis

Worry About

- Respiratory diphtheria early toxic manifestations: neck edema, pharyngitis, large pseudomembranes,

massive swelling of the tonsils, bull-neck diphtheria (with massive edema of the submandibular and paratracheal region and foul breath, thick speech, and stridor), hoarseness, and difficulty breathing are associated with severe advanced disease/poor prognosis and with a significant early risk of total airway obstruction.

- Late toxic manifestations of diphtheria: polyneuropathy (resembles Guillain-Barré syndrome) and myocarditis (cardiac arrhythmias, conduction abnormalities, or CHF).
- Other complications: Septic arthritis, pneumonia, renal failure, endocarditis, encephalitis, cerebral infarction, and pulmonary embolism.
- Fatal pseudomembranous diphtheria typically occurs in pts with nonprotective antibody titers and in unimmunized pts. Death occurs in 5–10% of respiratory cases. Risk factors for death include bull-neck diphtheria, myocarditis with ventricular tachycardia, atrial fibrillation or complete heart block, an age of >60 y or <6 mo, alcoholism, extensive pseudomembrane elongation, and laryngeal, tracheal, or bronchial involvement, and delayed antitoxin administration.

Overview

- Diphtheria is caused by superficial infection of the respiratory tract or skin with toxin-producing strains of *Corynebacterium diphtheriae*. The pathogens

multiply locally and produce diphtheria toxin. This results in necrosis of the mucosal cells and production of a thick, gray pseudomembrane containing fibrin, epithelial cells, bacteria, and neutrophils. Diffusion of toxin in the circulation causes toxic neurologic and myocardial complications.

- The major risk factor for *C. diphtheriae* infection continues to be travel to an endemic country (Indian subcontinent, Africa, or South East Asia).
- Prompt consideration of diphtheria: Severe pharyngitis, difficulty swallowing, respiratory compromise, or signs of systemic disease, including myocarditis or generalized weakness, and presence of a pharyngeal pseudomembrane or an extensive exudate.
- Respiratory diphtheria: Sore throat with low-grade fever and a strongly adherent pseudomembrane of the tonsils, pharynx, or nose. Occasionally weakness, dysphagia, headache, and voice change. The diphtheritic pseudomembrane is gray or whitish, sharply demarcated and tightly adherent to the underlying tissues. Respiratory diphtheria can progress to a swollen so-called bull neck, and the pseudomembrane can progress to cause airway obstruction. Attempts to dislodge the membrane may cause bleeding. Respiratory diphtheria remains the most common clinical presentation.
- Systemic toxin-mediated neurologic and cardiac toxicity of diphtheria: Neuritis and polyneuropathy