Cardiomyopathy, Restrictive

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Risk

- About 5% of all primary cardiomyopathies.
- Idiopathic type is rare, and may be familial.
- 50% of pts with AL-type amyloidosis are affected.
- Endomyocardial fibrosis is endemic in Africa, Asia, and Central and South America.

Perioperative Risk

- Diastolic dysfunction and low cardiac output state.
- Right heart failure with ascites and congestive hepatomegaly.
- Left heart failure with pulmonary edema.
- Cardiac arrhythmias, especially atrial fibrillation, ventricular arrhythmias, and AV block.

Worry About

- Thromboembolic complications
- Valvular insufficiency
- Autonomic neuropathy causing hemodynamic instability
- Respiratory, renal, CNS, and airway manifestations of underlying disease.

Overview

- Heterogeneous group of diseases characterized by restrictive cardiac physiology and diastolic dysfunction.
- Cardiac amyloidosis is a disorder of extracellular deposition of proteinaceous material in the myocardium and other organs.
- Endomyocardial fibrosis is a restrictive obliterative cardiomyopathy associated with eosinophilia.
- Pathophysiology: Increased stiffness of the myocardium that leads to restrictive ventricular filling with elevated filling pressures and dilated atria.
- Left ventricular systolic function is usually normal.
- Cardiac valves may be affected by infiltrative conditions causing stenosis or regurgitation
- Cardiac amyloidosis has a poor prognosis, especially when LVH, reduced systolic function, and heart failure is present.

Etiology

- Primary restrictive cardiomyopathy includes idiopathic (unknown cause) and genetic causes (mutations of sarcomere proteins including troponin I and T).
- Secondary restrictive cardiomyopathy occurs as part of a multisystem disorder, which include infiltrative diseases (amyloidosis, sarcoidosis, Gaucher disease), storage diseases (hemochromatosis, Fabry disease, glycogen storage disease), autoimmune disease (scleroderma), endomyocardial disease (carcinoid, endomyocardial fibrosis), and as a sequelae of cancer therapy (radiation therapy, anthracycline).

Usual Treatment

- Loop diuretics reduce pulmonary and systemic congestion
- ACE inhibitors and angiotensin receptor blockers may counteract neurohormonal changes associated with heart failure
- Pts with AFIB require rate control with beta-blockers, electrical/pharmacologic cardioversion to restore normal sinus rhythm, and anticoagulation.
- Pacemaker may be required with high-grade A-V block.
- Heart transplantation may benefit pts with advanced disease.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Possible difficult airway and macroglossia secondary to amyloidosis Uveitis secondary to sarcoidosis Supraglottic granuloma secondary to sarcoidosis	Dyspnea Dysphagia Abnormal speech	Enlarged tongue Cervical lymphadenopathy Conjunctival/iris nodules secondary to sarcoidosis	Careful airway assessment Ophthalmologic evaluation
RESP	Cardiogenic pulmonary edema Pleural effusion Pulmonary hypertension Restrictive lung disease secondary to sarcoidosis	Dyspnea Cough Chest pain Hoarseness	Crackles Rhonchi Stridor	CXR CT chest Spirometry ABG
CV	Diastolic dysfunction Possible LV systolic impairment Bi-atrial enlargement Valvular abnormalities Conduction abnormalities and AV block Atrial/ventricular arrhythmia Small vessel disease secondary to amyloidosis	Dyspnea Orthopnea Edema Enlarged abdomen Syncope Angina	Irregular pulse Elevated JVP Kussmaul sign Hepatosplenomegaly	ECG ECHO Cardiac cath Cardiac MRI Endomyocardial biopsy BNP Holter monitor
RENAL	Nephrotic syndrome secondary to amyloidosis Nephrogenic DI Fanconi syndrome ESRD	Proteinuria Renal insufficiency	Edema Hypertension	BUN/Cr Urine analysis Renal Bx
NEURO	Autonomic/peripheral neuropathy secondary to amyloidosis Cranial neuropathy secondary to sarcoidosis Carpal tunnel syndrome secondary to sarcoidosis/amyloidosis	Paresthesias Weakness Incontinence Diarrhea	Orthostatic hypotension	Tilt table test
GI	Malabsorption (malnutrition) secondary to amyloidosis Ascites GI bleed Gastroparesis Pseudo-obstruction	Weight loss Diarrhea Nausea, vomiting Abdominal pain	Palpable liver and spleen Jaundice and icterus	LFT GI endoscopy and biopsy
HEME	Anemia Thrombocytopenia/thrombocytosis Eosinophilia Plasma cell dyscrasia/multiple myeloma	Bleeding Bruising	Purpura Pallor	CBC Bone marrow aspirate Serum protein electrophoresis Iron studies
ENDO/ METAB	DM in hemochromatosis Hypothyroidism/hypopituitarism secondary to sarcoidosis Electrolyte abnormalities			Basic metabolic panel Serum calcium Fasting blood sugar HbA1c Endocrine evaluation Thyroid function test
MS	Arthropathy/polyarthritis secondary to amyloidosis/sarcoidosis Compression fractures Myopathy secondary to amyloidosis	Bone pain Joint pain	Shoulder pad sign	Joint aspiration Imaging

Key References: Wexler RK, Elton T, Pleister A, et al.: Cardiomyopathy: an overview, *Am Fam Physician* 79:778–784, 2009; Fleisher LA, Fleischmann KE, Auerbach AD, et al.: 2014 ACC/AHA guideline on perioperative cardiovascular evaluation and management of patients undergoing noncardiac surgery: executive summary: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines, *Circulation* 130:2215–2245, 2014.

Perioperative Implications

Preoperative Preparation

- Assess myocardial function and signs of left and right heart failure.
- Evaluate volume status.
- Identify electrolyte abnormalities.
- Assess conduction disturbances and ensure recent pacemaker interrogation.
- Continue perioperative beta-blockade.

Monitoring

- Supplement standard ASA monitors with invasive intra-arterial blood pressure monitoring.
- Central venous access allows monitoring of right-sided pressures and administration of vasoactive and inotropic drugs.
- PA catheter may be useful for assessment of loading conditions and measuring cardiac output in pts with advanced disease.
- Intraop TEE monitoring may be considered when significant fluid shifts are expected and when valvular dysfunction or myocardial depression is present.

Airway

- Macroglossia and laryngeal tumors occur in pts with amyloidosis.
- Supraglottic lesions associated with sarcoidosis may complicate airway management.

Induction

- Maintain preload and adequate heart rate to ensure adequate cardiac output.
- Avoid medications that cause myocardial depression, bradycardia, or decrease venous return.
- Etomidate is suitable because of minimal impact on hemodynamics and myocardial function.
- Ketamine, despite intrinsic cardiodepressant properties, maintains SVR.
- Carefully titrate IV opioids such as fentanyl or sufentanil.
- Caution with neuraxial blockade because of reduced venous return and SVR.

Maintenance

- A balanced anesthesia technique with lower doses of inhaled volatile anesthetics supplemented with opioids can be used.
- TIVA may also be considered.
- Judicious fluid management guided by invasive pressure monitoring, urine output, and other indices of tissue perfusion.
- Maintain normal sinus rhythm with normal heart rate.
- Monitor for hypotension, especially with autonomic neuropathy.

Extubation

- Ensure hemodynamic stability, adequate respiratory drive, and complete neuromuscular reversal.

Postoperative Period

- Ensure adequate cardiac output and end-organ perfusion.
- Postop pain management with central neuraxial blockade entails risk of decreased venous return and reduced SVR.

Anticipated Problems/Concerns

- Management of postop right (hepatic congestion, peripheral edema) and left heart failure (pulm edema).
- Postop AV block and arrhythmias.

Carnitine Deficiency

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Risk

- Rare (1:40,000 in Japan)

Perioperative Risks

- Hypoglycemia triggered by fasting
- Massive rhabdomyolysis and cardiac arrest described following GA and succinylcholine. (The response may be confused with malignant hyperthermia.)
- No evidence that susceptibility to malignant hyperthermia is associated with the carnitine palmitoyltransferase enzyme system

Worry About

- Periop hypoglycemia: Avoid prolonged fasting; IV glucose should be administered
- Neurologic and cardiopulmonary status: Determine if a cardiomyopathy is present

Overview

- CACT and CPT2 are essential cofactors in enzymatic transport of long-chain fatty acids into mitochondria, in which they are oxidized.
- When carnitine is deficient, peripheral tissues cannot use fatty acids for energy production, and the liver cannot adequately make ketone bodies as an alternative substrate.
- The tissues become glucose dependent, and their metabolism exceeds the liver's capacity for glucose production.
- This glucose dependency can lead to severe liver failure (increased hepatic enzymes, lactic acidosis, and encephalopathy) and hypoketotic hypoglycemia.

Etiology

- CACT resulting from mutations in the *SLC22A5* gene, which leads to the production of defective OCTN2 carnitine transporters
- CPT deficiency resulting in impaired transfer of fatty acids into mitochondria
- CPT deficiency associated with rhabdomyolysis and higher incidence of renal insufficiency

Usual Treatment

- Dietary supplementation with L-carnitine and high-carbohydrate diet to prevent hypoglycemia

Assessment Points

System	Effect	Assessment by Hx	Test
CV	Cardiomyopathy		ECHO
HEPATIC	Hypoglycemia Hepatomegaly with fatty infiltration	Lethargy	Blood glucose Bilirubin Liver function tests
HEME	Coagulopathy	Bleeding	Hypoprothrombinemia
CNS	Encephalopathy	Vomiting, diarrhea	Hyperammonemia
RENAL	Renal insufficiency	Recurrent myoglobinuria	BUN/Cr

Key References: Lucas M, Hinojosa M, Rodriguez A, Garcia Guasch R: Anaesthesia in lipid myopathy, *Eur J Anaesthesiol* 17(7):461–462, 2000; Lilker S, Kasodekar S, Goldszmidt E: Anesthetic management of a parturient with carnitine palmitoyltransferase II deficiency, *Can J Anaesth* 53(5):482–486, 2006.

Perioperative Implications

Preoperative Preparation

- Continue daily carnitine therapy.
- Glucose infusion preop.
- Avoid protracted preop fasting.
- For emergency surgery while pt is in metabolic crisis, rehydrate; correct glucose, acid-base, and electrolyte imbalances; use IV carnitine if necessary; treat hypoprothrombinemia with FFP.

Monitoring

- Routine

Airway

- Best to avoid succinylcholine for intubation

Maintenance

- IV glucose infusion and frequent monitoring of serum glucose level.
- Muscle weakness may be present and requires careful titration of muscle relaxant dosing.

Extubation

- No unusual concerns

Adjuvants

- Consider antiemetic prophylaxis to speed resumption of oral intake.

Anticipated Problems/Concerns

- Periop hypoglycemia and metabolic acidosis/decompensation.
- In the presence of carnitine deficiency, propofol may theoretically result in mitochondrial dysfunction and cellular hypoxia.
- Increased risk of bupivacaine-induced cardiotoxicity may be seen. Double-check before giving bolus epidural injections.