

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 palmitoyl-transferase 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; Likker 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.