Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Htn (LVH with diastolic dysfunction, MI/CHF if long standing), dysrhythmias	Angina, poor exercise tolerance, palpitations	Two-flight walk, ability to lie flat, chest auscultation, peripheral edema check	ECG, ECHO, CXR
RENAL	Decreased serum K^+, nephropathy	Constipation, fatigue, muscle weakness/pain		K^+, BUN/Cr

Key References: Hansson J: Liddle's syndrome: review of the clinical disorder and its molecular genetic basis, *Endocrinologist* 10(4):229–236, 2000; Hayes NE, Aslani A, McCaul CL: Anaesthetic management of a patient with Liddle's syndrome for emergency caesarean hysterectomy, *Int J Obstet Anesth* 20(2):178–180, 2011.

Perioperative Implications

Preoperative Preparation

- Treatment of Htn; ideally normalization of <140/90 mm Hg would occur prior.
- Assessment of cardiac function (ECG, CXR, possible ECHO).
- Assessment of renal function (specifically BUN/Cr).
- Assessment of electrolyte balance (specifically K^+) and associated clinical symptoms.

Monitoring

- Continuous ECG to monitor myocardial ischemia and hypokalemic dysrhythmias

Airway

- No airway changes expected

Induction

- Labile hemodynamics in pts with longstanding Htn requires careful titration of induction medications.
- Caution using drugs dependent on renal excretion.

Maintenance

- Higher mean arterial pressure goals tend to be required.
- Monitor fluid balance; renal insufficiency is possible, and untreated pts are volume-overloaded.
- Possible prolongation of NMB due to hypokalemia.
- Lyte monitoring/replacement if symptomatic prior or ECG changes are present; hyperventilation and nasogastric suctioning can worsen hypokalemia.

Extubation

- Pts are prone to excessive tachycardia and Htn; exclude typical causes such as pain, agitation, hypoxia, and hypercarbia before treating.

Postoperative Period

- Adjuvant Htn therapy often required
- Lyte monitoring

Anticipated Problems/Concerns

- Untreated cerebrovascular, CV, and renal disease

Lipidemias

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Risk

- Prevalence in USA: 13.1% in people >20 y.
- Prevalence highest among Hispanics.
- Cigarette smoking is a risk factor.
- Incidence highest among men ≥45 y and women ≥55 y.
- Htn is a risk factor.
- Low HDL (<40 mg/dL) is a risk factor.
- Family Hx of premature CHD in first degree relative (male <55 y or female <65 y) is a risk factor.

Perioperative Risks

- Pancreatitis with hypertriglyceridemia
- Stroke and transient ischemic attacks
- Myocardial ischemia, infarction, CHF

Worry About

- Angina of increasing frequency or severity and new-onset angina
- Peripheral atherosclerosis
- Worsening or new-onset CHF
- TIAs

Overview

- Hypertriglyceridemia, hypercholesterolemia, lipodystrophy: Köbberling-Dunnigan syndrome (familial lipodystrophy of limbs and trunk, autosomal dominant) may lead to macrosomia; familial generalized lipodystrophy (Berardinelli-Seip syndrome: autosomal recessive) leads to macrosomia.
- Hypolipidemia: LDL deficiency (autosomal recessive abetalipoproteinemia, autosomal dominant familial hypobetalipoproteinemia); normotriglyceridemic abetalipoproteinemia (LDL absent); autosomal recessive Tangier disease (severe deficiency of HDL); secondary to cancer, myeloproliferative disorders, liver failure familial hypoalphalipoproteinemia (HDL deficiency).

Etiology

- Autosomal dominant or recessive inheritance
- Secondary to systemic illness (i.e., primary hypothyroidism, nephrotic syndrome, and extrahepatic obstruction of bile)

Usual Treatment

- Cholestyramine and colestipol inhibit absorption of bile acids derived from cholesterol.
- Neomycin blocks cholesterol absorption.
- Diet and exercise.
- Thyroid hormone clears LDL.
- Fish oils (omega-3 fatty acids) reduce triglyceride levels.
- Nicotinic acid inhibits VLDL and LDL production; also an HDL-raising drug.
- Fibric acids clofibrate and gemfibrozil to increase catabolism of triglyceride-rich lipoproteins.
- Niacin/statin combination therapy promotes optimal lipid values for several at-risk pt populations.
- Statins inhibit HMG CoA reductase; these are the mainstay of lipid-lowering therapy, reducing risk for ASCVD.

Assessment Points

System	Effect	Assessment by Hx	PE	System
HEENT	Tangier disease		Lobulated, bright orange-yellow tonsils Hepatosplenomegaly Peripheral neuropathy (in 50% of pts)	Lipoprotein profile
CV	Myocardial ischemia and infarction Left ventricular dysfunction	Angina or its equivalents Dyspnea, edema, exercise intolerance, MI	Displaced PMI S_3 S_4	ECG, CXR, stress testing, ECHO, coronary angiography
RESP	CHF	Dyspnea, orthopnea, cough	Rales and rhonchi	CXR
RENAL	Impaired renal perfusion	Nighttime urinary frequency		BUN, Cr
CNS	Cerebrovascular atherosclerosis	TIAs	Carotid bruit	Carotid US and angiography

Key References: Stone NJ, Robinson JG, Lichtenstein AH, et al.: 2013 ACC/AHA guideline on the treatment of blood cholesterol to reduce atherosclerotic cardiovascular risk in adults: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines, *J Am Coll Cardiol,* 63(25 pt B):2889–2934, 2014; Hindler K, Shaw AD, Samuels J, et al.: Improved postoperative outcomes associated with preoperative statin therapy, *Anesthesiology,* 105(6):1260–1272, 2006.

Perioperative Implications

Preoperative Preparation
- Assess for CAD and peripheral vascular disease.
- Beta-blockers and nitrates given periop as tolerated.
- Statins have been associated with improved postop outcomes.

Monitoring
- Consider pulm artery catheter, transesophageal ECHO in the presence of large fluid shifts, history of ischemia, and high-risk surgery.

Airway
- Pts may have large head and neck and be overweight, making intubation difficult.

Maintenance
- Avoid hypothermia and anemia.
- Monitor for ischemia and cardiac failure.
- Insulin increases activity of lipoprotein lipase and releases FFAs.
- Sympathetic stimulation, stress, and catecholamines release FFAs.
- Spinal or epidural anesthesia and beta-blockers reduce FFA levels.
- Heparin releases two triglyceride hydrolases: lipoprotein lipase inhibited by protamine, and hepatic lipase resistant to protamine.

Extubation
- During noncardiac surgery, this may be time of greatest risk for ischemia.

Adjuvants
- Depend on lipid-drug binding and end-organ disease

Postoperative Period
- High incidence of ischemia, tachycardia, and MI for several days after noncardiac surgery.
- Treat pain, hemodynamic, and biochemical abnormalities.

Anticipated Problems/Concerns
- Concerns are related to atherosclerotic disease.

Long QT Syndrome

Regina Linganna | Lee A. Fleisher

Risk
- Prevalence of cLQTS: Approximately 1:5000 live births.
- Incidence of cLQTS: 1 in 10,000.
- 60–70% of those diagnosed are females.
- Males under 10 y of age have the highest mortality.
- Pts usually present in childhood with a cardiac event.

Perioperative Risks
- Torsades de pointes
- Sudden cardiac death

Worry About
- Sympathetic stimulation with laryngoscopy, pain, etc.
- Electrolyte abnormalities: hypokalemia, hypocalcemia, and hypomagnesemia

Overview
- cLQTS is diagnosed when the corrected QT interval is >500 ms in the absence of other causes
- Jervell and Lange-Nielsen syndrome is cLQTS associated with deafness; Romano-Ward syndrome is cLQTS without deafness
- aLTQS is most commonly drug induced or caused by an electrolyte abnormality
- Pathophysiology: Arrhythmogenic prolongation of the QT interval caused by mutated genes encoding the cardiac myocyte ion channels

Etiology
- Most common gene mutations: LQT1, LQT2, and LQT3.
- aLTQS primarily prolongs the QT interval by blockade of the rapid delayed I_{Kr}, encoded by *HERG*.
- Drug-induced: succinylcholine, ketamine, atropine, quinolone and macrolide antibiotics, dexmedetomidine, and ondansetron.

Usual Treatment
- Beta-blockade is the first line treatment
- In pts who are symptomatic despite beta-blockade, AICD implantation may be considered

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Torsades de pointes Ventricular fibrillation Sudden cardiac death	Convulsions, syncope	Tachycardia Tachycardia	ECG ECG
CNS	Syncope	Loss of consciousness	Neurologic exam	
METAB	Electrolyte abnormalities			Electrolyte panel Ca^{2+}, Mg^{2+}, K^{+}

Key References: Havakuk O, Viskin S: A tale of 2 diseases: the history of long-QT syndrome and Brugada syndrome, *J Am Coll Cardiol* 67(1):100–108, 2016; Owczuk R, Wujtewicz, Zienciuk-Krajka E, et al.: The influence of anesthesia on cardiac repolarization, *Minerva Anesthesiol* 78(4):483–495, 2012.

Perioperative Implications

Preoperative Preparation
- Elicit family history of sudden cardiac death or congenital deafness.
- 12-lead ECG.
- Ensure maintenance of beta-blockade.
- Ensure availability of defibrillator.
- Avoidance of spinal anesthesia superior to the level of T10 due to the increase in sympathetic tone of the unanesthetized fibers.

Monitoring
- Standard ASA monitors
- Adequate IV access for resuscitation should pt convert to lethal arrhythmia

Airway
- Pt needs to be deeply anesthetized before manipulation of the airway to reduce sympathetic discharge with laryngoscopy.

Preinduction/Induction
- Adequate anxiolysis prior to entering the OR to reduce sympathetic discharge associated with preop anxiety

Maintenance
- Multimodal analgesia for adequate intraop and postop pain control
- Avoidance of hypothermia and associated shivering
- Avoidance of hyperthermia to reduce sympathetic discharge associated with fever
- Avoidance of medications that further prolong the QT duration
- Avoidance of hypokalemia, hypomagnesemia, and hypocalcemia

Extubation
- Consider deep extubation of these pts to reduce sympathetic discharge with emergence.

Postoperative Period
- Continue standard ASA monitors.
- Adequate pain control.

Anticipated Problems/Concerns
- Conversion to lethal arrhythmia secondary to electrolyte abnormality, sympathetic stimulation, or medications that prolong the QT duration