Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Ulceration, oral lesions	Dysphagia, pain	Airway assessment	
CV	Rare: Pericardial effusion, conduction defects, murmurs, CHF Mediastinal mass	Dyspnea, fatigue	Narrow pulse pressure, pericardial friction rub, cardiomegaly	CXR, CT scan, ECG, ECHO
GI	Hepatosplenomegaly hypoalbuminemia	Loss of appetite Weight loss	Hepatosplenomegaly	Albumin
HEME	Anemia Leukostasis Thrombocytopenia	Weakness, easy fatigue,	Pallor Ecchymoses Petechiae, easy bruising, nose-bleeds	CBC Bone marrow aspirate results
RENAL	Renal failure from tumor lysis syndrome (acute loss of tumor)	Decreased UO	Decreased UO	BUN/Cr, hyperkalemia ↑ phosphate, ↑ or ↓ Ca^{2+}
CNS	Cranial nerve infiltration (very rare), meningeal leukemia (less common in adults), vincristine neuropathy	Cranial nerve palsies, clouding of mental status, peripheral neuropathy	Weakness	EMG
MS	Infiltration of bony cortex and periosteum, synovial membranes	Bone pain	Bone swelling	X-ray CT scan

Key References: Bryan JC, Jabbour EJ: Management of relapsed/refractory acute myeloid leukemia in the elderly: current strategies and developments, *Drugs Aging* 32(8):623–637, 2015; Latham GJ: Anesthetic considerations for the pediatric oncology patient—part 2: systems-based approach to anesthesia, *Paediatr Anaesth* 20(5):396–420, 2010.

Perioperative Implications

Preoperative Preparation

- Assess volume status, CBC, electrolytes, renal function, N/V, diarrhea, and oral mucositis.
- Review sign and symptoms and imaging reports of mediastinal mass for compromised airway.
- Airway assessment: Oral mucosal ulceration, edema, fibrosis, and neck mobility.
- Neutropenia precaution: Periop isolation, aseptic technique for safe port access, avoidance of per rectum medication and probe.

Monitoring

- Routine

Airway

- Signs of dysphagia, ulcerations, and airway bleeding from chemotherapy and candidiasis.
- Oral leukemia lesions can occur prior to or during therapy.
- Anterior mediastinal mass: Risk of difficult intubation, ventilation, and compromise hemodynamic. Fiber optic intubation, inhalation induction, and rigid bronchoscopy may be required; lateral or prone position may help with ventilation. Avoid muscle relaxants.
- Chronic radiation: Potential for difficult airway; high-dose or chronic radiation to oral cavity, head, and neck cause fibrosis and stiffness of soft tissue resulting in limited mouth opening and neck extension.
- Post radiation changes: Affecting airway mucosal fibrosis, subglottic edema, and supraglottic and subglottic narrowing or stenosis may complicate the airway management.
- Leukemic infiltration of tonsils and adenoids, retropharyngeal lymph nodes, and cervical lymphadenopathy may cause airway obstruction and difficulty in intubation and ventilation.

Induction

- Brief heparinization and thrombocytopenia may influence choice of local, spinal, or epidural
- MAC or deep sedation with propofol and remifentanil infusion preferred over propofol alone
- GA may require in younger pediatric age with special equipment and monitoring in various remote anesthesia locations, such as radiation therapy suite

Maintenance/Extubation

- Routine inhalation technique or TIVA
- Low FIO_2 for during and after bleomycin therapy
- Prophylaxis for N/V
- Multimodal pain management

Anticipated Problems/Concerns

- Risk of infection; aseptic technique with placement of all lines.
- Dexamethasone may precipitate tumor lysis syndrome; anesthesia provider should communicate with oncology team before using dexamethasone for N/V prophylaxis in high-risk pts.
- Caution required for subclinical cardiomyopathy, pericardial effusion, airway management, and pulmonary dysfunction after chemotherapy and radiation therapy.
- Use of epidural blood patch for treatment of PDPH is controversial; may increase risk for infectious complication and CNS leukemia spread.
- Blood products: CMV depleted, irradiated blood, platelets; careful with doses and GVHD.
- ICU admission: Cardiorespiratory failure, pleural effusion, pneumonia, sepsis, compromised airway, GVHD following HSCT, pulmonary complications, and multiple organ failure.

Liddle Syndrome

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Risk

- Extremely rare but described in a variety of populations
- True incidence and prevalence unknown

Perioperative Risks

- Chronically untreated Htn

Worry About

- Undiagnosed cerebrovascular, CV, and/or renal disease secondary to chronic Htn
- Worsening of hypokalemia with hyperventilation and nasogastric suctioning
- Hypokalemia-induced dysrhythmias and potentiation of NMB

Overview

- Monogenic AD gain-of-function mutation in the ENaC resulting in early onset Htn, hypokalemia, and metabolic alkalosis with suppressed plasma renin activity; resembles primary aldosteronism but aldosterone excretion is markedly suppressed (also known as "pseudoaldosteronism").
- Htn results from volume expansion due to increased Na^+ reabsorption via the constitutively active ENaC; urinary secretion of K^+ and H^+ occurs to balance out movement of electrical charges, resulting in a hypokalemic metabolic alkalosis.
- Presentation is variable.
  - Htn may not be early in onset or severe.
  - Hypokalemia may be absent.
  - Family history is not reliable, as spontaneous mutations have been reported.

Etiology

- ENaC is a membrane-bound ion channel located on the apical membrane of the principal cell in the distal tubule, which is selectively permeable to sodium ions; their activity is normally regulated by aldosterone.
- ENaC is composed of three subunits: α, β, and γ.
- NEDD4, a ubiquitin ligase enzyme, negatively modulates ENaC via ubiquitination.
- Gene mutations resulting in deletions or alterations of the carboxy-terminus of the β or γ subunits (located on chromosome 16p) make NEDD4 binding impossible; this prevents channel degradation and removal, resulting in the constitutive activity of ENaC.

Usual Treatment

- Amiloride or triamterene therapy (direct ENaC inhibitors) with a low-sodium diet.
- Kidney transplantation is curative.
- Htn and hypokalemia are not responsive to mineralocorticoid antagonists (e.g., spironolactone) because the increased activity of ENaC is not mediated by aldosterone.

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.