Assessment Points

Type	Effect	Assessment by Hx	PE	Test
MEN 1	Parathyroid hyperplasia (assoc nephrolithiasis) Pancreatic tumors (insulinoma, glucagonoma), gastrinoma Ant pituitary tumor (prolactinoma, GH tumor, ACTH/Cushing)	Family Hx of endocrine tumors Fatigue, muscle weakness, flank pain, renal stones, Hx pathologic fractures Diaphoresis, palpitation, abdominal pain Diarrhea, reflux, dyspepsia Headache, visual changes	Htn Neck nodule Altered mental status Flank tenderness Tremor, mental status changes (hypoglycemia) Visual field defect Acromegaly (GH) Cushingoid habitus	NIBP and ECG Serum calcium Sestamibi scan, PTH level, neck CT, bone density, BUN/creatinine, pelvic x-ray Serum glucose, lytes, CT/MRI Endoscopic US Head CT/MRI metabolic panel, specific hormone level
MEN 2a AND 2b	Pheochromocytoma	Family Hx, episodic sweating, palpitations, anxiety, tremor	Htn (paroxysmal), arrhythmia	CT/MRI, NIBP ECG/consider ECHO, 24-h urine for catecholamines, metanephrines
	Medullary cancer of thyroid Parathyroid adenoma (see MEN I)	Can be asymptomatic Family Hx Hx urinary stones Symptoms of hypercalcemia	Thyroid mass Neck nodule	Calcitonin levels Serum calcium, serum PTH level, BUN/Cr Pelvic x-rays

Key References: Chen H, Sippel R, O'Dorisio MS, et al.: The North American Neuroendocrine Tumor Society consensus guidelines for the diagnosis and management of neuroendocrine tumors: pheochromocytoma, paraganglioma, and medullary thyroid cancer, *Pancreas* 39(6):775–783, 2010; Grant F: Anesthetic considerations in the multiple endocrine neoplasia syndromes, *Curr Opin Anaesthesiol* 18(3):345–354, 2005.

Perioperative Implications (Men 1)

Monitoring

- Parathyroid surgery: ECG signs of hypercalcemia (arrhythmias, prolonged PR, short QT), consider using EMG ETT for monitoring recurrent laryngeal nerve intraop. Unpredictable response to muscle relaxants with hypercalcemia, monitor the TOF. PTH levels; significant decrease expected post successful resection; monitor calcium level postop.
- Pituitary adenomas: Tight BP control; acromegalics may have impaired ulnar circulation to hand which increases risk morbidity from radial a-line; monitor urine output (risk for DI, SIADH)
- Insulinoma surgery: Requires tight, careful blood glucose control; increased risk hypoglycemia periop; arterial line
- Gastrinomas: Arterial line; pts at risk for labile BP

Airway

- Acromegaly: Increased risk of difficult mask airway and intubation; also increased incidence of sleep apnea; have difficult airway equipment ready
- Parathyroidectomy: Risk of surgical damage to recurrent laryngeal nerve, and vocal cord paresis periop (risk of hoarseness to stridor to complete airway obstruction if bilateral)

Maintenance

- Parathyroidectomy: Draw post-resection PTH levels to confirm removal of tumor
- Insulinomas and gastrinomas: Monitor volume status, glucose, and BP control
- Pituitary adenomas: Usually transsphenoidal approach; tight BP control; watch UO

Perioperative Implications (Men 2)

Monitoring

- Pheochromocytoma: Standard ASA monitors, arterial line, CVP, UO.
- Total thyroidectomy: Standard ASA monitors. Consider use of EMG ETT to monitor recurrent laryngeal nerve intraop. Postop PTH levels to check for adequate parathyroid function.
- Parathyroidectomy: See MEN 1 section.

Airway

- Thyroidectomy and parathyroidectomy: Review ENT preop evaluation, including ENT's fiber optic exam of larynx, CT/MRI scans, sestamibi localization scans for potential mass effects of tumor on airway; also note baseline vocal cord function. Communicate with surgeon for plan

Maintenance

- Pheochromocytoma: Tight BP control before and during resection (anesthetics, nipride, phentolamine, esmolol, calcium channel blockers, epidural infusions); after adrenal ligation, BP support with fluid boluses, prn pressors (NE, phenylephrine). Monitor glucose.
- Thyroidectomy: If using EMG ETT, avoid muscle relaxants.
- Parathyroidectomy: See MEN 1.

Adjuvants

- Pheochromocytomas: Require adequate preop treatment to control BP, HR, and restore blood volume (10–14 d alpha-adrenergic blockers [e.g., phenoxybenzamine, prazosin], hydration, then initiate beta-blockade)
- Hyperparathyroidism with symptomatic hypercalcemia: Preop hydration; diuresis with furosemide; consider biphosphonates, calcitonin, or glucocorticoids

Anticipated Problems/Concerns

- MEN 1: Parathyroidectomy: Postop hypocalcemia, recurrent laryngeal nerve damage/VC paresis, neck hematoma/airway compromise. Transsphenoidal pituitary adenoma resection: Hypopituitarism, SAIDH/DI. Acromegaly: Potential difficult airway. Pancreas tumors: Hyperglycemia/hypoglycemia. Gastrinoma/VIPoma: Labile BP
- MEN 2: Pheochromocytoma; malignant Htn and labile BP, increased risk of CVA, and MI

Multiple Myeloma

Ryan J. Kline | Gregory Bordelon | Alan David Kaye | Amit Prabhakar

Risk

- Represents 1.6% of all new cancer cases in USA; estimated 26,850 new cases in 2015.
- Estimated 11,240 deaths, or 1.9% of all cancer deaths in USA in 2015.
- Incidence: 7.5:100,000 white males; 4.5:100,000 white females; 15.1:100,000 black males; 11.2:100,000 black females; 7.9:100,000 all races male; 5.1:100,000 all races female (based on 2008 to 2012 data).
- Race: 1.1% of all malignancies in white population; 2.1% of all malignancies in black population.
- Male to female ratio: 3:2.
- Age: Median age 68 y in men, 70 y in females; most frequently diagnosed between 65 and 74 y (28.2%).
- Increased risk among those with MGUS.
- Fourteenth leading cause of cancer death.
- Survival: Median survival 3 y; 100% fatality rate; median age of death 75 y; 46.6% 5-y survival.

Perioperative Risks

- Pts typically anemic.
- Pathologic fractures occur with this disease; careful positioning and padding essential.
- Coagulopathy common with thrombocytopenia, thrombocytopathy, and decreased functional plasmatic coagulation factors.
- Renal failure is the most common cause of mortality; concern for anesthetics with renal elimination.
- Hypercalcemia common and can cause morbidity and mortality.
- Infection risk real, especially if pt has recently had a stem cell transplant.

Overview

- Part of a spectrum ranging from MGUS to plasma cell leukemia (malignancy of antibody forming cells).
- Also known as plasmacytosis, myelomatosis, or Kahler disease; classified within non-Hodgkin lymphomas.
- Proliferation of plasma cells results in functioning peripheral blood cells and leads clinically to
  - Impaired production of blood cells >pancytopenia (leucopenia anemia thrombocytopenia).
  - Formation of plasmacytoma (mass), leading to lytic lesions in bone.
  - Impaired immunity (humoral) >infections.
  - Increased plasma cells (antibody-forming cells) >amyloidosis (soft tissue, lungs, kidneys) and hyperviscosity.
- Presenting signs: High sedimentation rates, anemia, signs of coagulopathy.
- Renal failure from toxic immunoglobulin deposition in renal tubuli most common cause of mortality; 10% of pts develop amyloidosis.

Etiology

- Genetic instability: Translocation at 14q32 and/or deletion of chromosome 13, leading to either neoplastic plasmacytes producing either a monoclonal immunoglobulin (IgG, IgA, IgD) or isolated light chains (Bence Jones plasmacytoma)
- Environmental and occupational causes
- Radiation (increased incidence in survivors of the atomic bombing of Nagasaki)

Usual Treatment

- Alkylating chemotherapeutic agent
- Immunomodulatory drugs: thalidomide, lenalidomide, or pomalidomide
- Stem cell transplantation
  - Autologous
  - Allogenic
- Glucocorticoids
- Interferon alpha-2b
- Protease inhibitors
  - Bortezomib: inhibitor of 26S proteasome >inhibition of proteasome in myeloma
  - Carfilzomib: inhibitor of 20S proteasome >increase in polyubiquitinated proteins

Treatment of Complications

- Bone disease-related pain: Opioid preparations, immediate- and extended-release formulations, lidoderm patches, diclofenac topical products; radiation (refractory pain and cord compression), surgical intervention
- Anemia: Iron, B_{12}, folate, erythropoietin, transfusion
- Infection: Vaccination against *Streptococcus pneumoniae*, *Haemophilus influenzae*, H1N1, seasonal flu; antibiotics; IV immune globulin
- Hypercalcemia: IV fluid and corticoid steroid, bisphosphonates (if unresponsive to hydration), calcitonin, furosemide
- Renal failure: Treatment of dehydration, hypercalcemia, and hyperuricemia; chemotherapy (e.g., vincristine, doxorubicin); alkaline diuresis; trial of plasma exchange in acute evolving renal failure; hyperviscosity syndrome; exchange of plasma (plasmapheresis)

Assessment Points

System	Clinical Manifestations	Signs and Symptoms	Anesthetic Implication
MS	Bone pain Pathologic fracture	Usually lumbar 95% more than one side	Positioning to prevent fracture
HEME	Bleeding and bruising Coagulopathy Normochromic normocytic anemia Capillary fragility	Secondary to thrombocytopenia Absorption of clotting factor Weakness Purpura Dark circles (raccoon-like) around eye, secondary to prolonged Valsalva	Availability of FFP and plts Increased transfusion requirements, ventilator management
METAB	Hypercalcemia Infection Hyperviscosity	Confusion, somnolence, constipation, nausea, thirst, bone pain Secondary to humoral immunity of normality Epistaxis Visual disturbance Carpal tunnel Headache Somnolence, bruisability	Increased fluid requirements, maintenance of adequate urine output Antibiotic coverage Preoperative: plasmapheresis, increased fluid requirement intraop Temperature maintenance to prevent microvascular sludging
CNS/PNS	Spinal cord compression Meningitis Carpal tunnel Peripheral neuropathies Stroke (hyperviscosity)	Signs of weakness and numbness of extremities	Positioning of pt Diligent use of muscle relaxants Avoidance of depolarizing muscle relaxants
RENAL	Renal insufficient/failure	Secondary to direct tubular injury Amyloidosis Involvement by plasmacytoma	Adequate hydration
RESP	Pneumonia Respiratory insufficiency	Secondary to rib fracture	Extubation problems Pneumothorax intraop
HEENT	Amyloidosis	Macroglossia Skin lesions of lips	Airway problems

Key References: Kyle RA, Rajkumar SV: Multiple myeloma, *N Engl J Med* 351(18):1860–1873, 2004; Palumbo A, Gay F: How to treat elderly patients with multiple myeloma: combination of therapy or sequencing, *Hematology Am Soc Hematol Educ Program* 566–577, 2009, http://dx.doi.org/10.1182/asheducation-2009.1.566.

Perioperative Implications

Preoperative Preparation

- Recombinant erythropoietin increases Hgb and decreases transfusion requirement
- Antibiotics and gammaglobulin prophylaxis

Airway

- May be difficult due to macroglossia

Maintenance

- Regional anesthesia is contraindicated due to bony lesions, coagulopathy, and neurologic deficit.
- Unpredictable pharmacokinetic of protein-bound drugs.

Postoperative Period

- Continue adequate hydration.
- Aggressive pulmonary toilet.
- Treat specific complication (refer to Treatment of Complications section).

Anticipated Problems/Concerns

- Careful positioning to prevent fractures

Multiple Organ Dysfunction Syndrome

Zerlina Wong | Jesse M. Raiten

Risk

- Most common cause of death for pts in ICU
- Incidence 11-40% of adult ICU pts
- Risk factors: (1) Severe illness at time of ICU admission; (2) severe sepsis or infection at time of ICU admission; (3) old age
- Associated with trauma, sepsis, shock, male sex, African American race, chronic health conditions, malnutrition, use of immunosuppressants

Perioperative Risks

- Labile hemodynamics
- Difficulty with oxygenation and ventilation
- Malnutrition
- Altered drug metabolism

Worry About

- Volume status
- Drug metabolism
- Antibiotic selection
- Difficulty cross-matching blood products
- Transfusion reactions

Overview

- MODS is a dynamic process; clinical course and causes are highly variable.