Overview

- Systemic vasculitis of small, medium, and occasionally large arteries
- Characterized by necrotizing granulomatosis of upper and lower respiratory tracts in addition to glomerulonephritis

Etiology

- Autoimmune disorder of unknown etiology.
- Type II hypersensitivity reaction.
- May involve lack of alpha-1 antitrypsin.
- Antineutrophilic cytoplasmic antibodies are involved.
- Symptoms include
  - Upper airway involvement in 95% of pts, including paranasal sinus drainage and nasal mucosa ulceration.
  - Subglottic stenosis present in 9–16% of pts.
  - Pulm involvement manifests as cavitating granulomatous lesions.
  - Pulm arterial/venous vasculitis creates V/Q mismatch and pulm shunting.
  - Lower resp tract findings also may be present including cough, dyspnea, and hemoptysis.
  - CXR may reveal alveolar opacities, diffuse hazy opacities, nodules, and pleural opacities.
  - 77% of pts manifest with renal failure.
  - Eye involvement in 52% of pts including conjunctivitis, scleritis, keratitis, uveitis, and episcleritis.
  - Skin symptoms include papules, vesicles, purpura, ulcers, and nodules occurring in 40% of pts.
  - Nonspecific symptoms include night sweats, malaise, fatigue, arthralgias, anorexia, and weight loss.
- Diagnosis:
  - Biopsy of nasopharyngeal lesion preferred, showing necrotizing granulomatous vasculitis
  - Biopsy of kidney or lung showing segmental necrotizing glomerulonephritis with no immunoglobulin deposition
  - Elevated ESR, leukocytosis, normocytic anemia, and thrombocytosis

Usual Treatment

- Cyclophosphamide combined with oral glucocorticoid.
- Complete remission may take 1–2 y.
- 90% of pts achieve improvement, and 75% have remission.
- 50% of pts in remission have relapse.
- Morbidity from disease includes renal insufficiency, hearing loss, tracheal stenosis, and saddle nose deformity.
- Drug considerations:
  - Glucocorticoid side effects include diabetes, cataracts, osteoporosis, and Cushingoid features.
  - Cyclophosphamide side effects include cystitis, bladder cancer, myelodysplasia, and infertility.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	MI PVD	H/o ischemic heart disease		ECG
RESP	Destructive lesions of epiglottis, pharynx, or larynx V/Q mismatch and pulm shunting, destructive lesions pulm parenchyma		SOB, cough, hemoptysis, pleuritic CP, upper airway ulcerations	PFTs, CXR, ABG
RENAL	Glomerular destruction and tubular atrophy	Caution with drugs dependent on renal excretion		Renal function panel, renal biopsy, urinalysis
HEME	Bleeding propensity	CH/o cyclophosphamide or methotrexate treatment	Petechiae, bleeding gums	CBC, clotting studies
OPHTH	Vision loss, keratitis, scleritis, conjunctivitis, uveitis		Ophthalmic exam	Visual acuity test
ENT	Nasal mucosal ulceration or obstruction	Nasal discharge or drainage, epistaxis, hyposmia, epiphora	Upper airway exam	Nasal biopsy
NEURO	Peripheral neuropathy		Peripheral neuro sensory exam	
DERM	Ulceration distal arms/legs			Skin biopsy

Key References: Rookard P, Hechtman J, Baluch AR, et al.: Wegener's granulomatosis, *Middle East J Anaesthesiol* 20(1):21–29, 2009; Kahn AM, Elahi F, Hashmi SR, et al.: Wegener's granulomatosis: a rare, chronic, and multisystem disease, *Surgeon* 4(1):45–52, 2006.

Perioperative Implications

Preoperative Preparation

- Upper airway assessment to identify ulcerations or obstructing lesions, CXR, and PFTs.
- Screen for symptoms including cough, dyspnea, hemoptysis, or pleuritic chest pain.
- Consider RA when possible, but be aware that pts may have peripheral neuropathy and coagulation disorders that may add risk to the procedure.

Intraoperative Considerations

- Upper airway considerations should include careful physical inspection using laryngoscopy for ulcers of the palate, pharynx, or epiglottis. Care should be taken during intubation to avoid bleeding or displacement of brittle tissue. May consider regional anesthesia to avoid airway manipulation.
- Respiratory considerations include increased dead space and V-Q mismatch due to pulmonary artery and vein vasculitis. Bronchial obstruction and destruction may occur; thus frequent suctioning may be required. Monitoring of ABG ensures adequate oxygenation.
- Cardiovascular considerations include increased risk of MI due to not only peripheral but also coronary vasculitis as well. Avoid situations of increased preload, afterload, heart rate, or coronary spasm.
- Pts on corticosteroid therapy should be given 100 mg hydrocortisone prior to surgery to avoid Addisonian hypotensive crisis.
- Renal considerations include avoidance of anesthetics that require renal excretion such as morphine, meperidine, diazepam, midazolam, vecuronium, pancuronium, and nitroprusside.
- Cyclophosphamide inhibits pseudocholinesterase, which prolongs the activity of succinylcholine, thus warranting consideration when determining paralytic drug choice.

Postoperative Period

- Close observation of the upper airway following extubation should be performed, as edematous granulation tissue from intubation is possible.

Wilms Tumor

Peter J. Davis

Risk

- Most common malignant renal tumor in childhood.
- Accounts for 6% of all childhood malignancies.
- 5-7.8 cases per million children <15 y old in the USA.
- Prevalence: Males equal to females.
- Peak age is 1–3 y.
- 5% bilateral.
- Relapse-free survival rate at 2 y: 90%.
- Pts with favorable staging have an 80–90% chance of cure. Pts with metastasis have 50% long-term survival.
- Overexpression of HER-2 oncoprotein is a good predictor of survival.
- Along with hepatoblastoma, more common in Beckwith-Wiedemann syndrome.

Perioperative Risks

- Increased intraabdominal pressure
- Immunocompromised
- Tumor extension into renal vein, IVC, and heart
- Some treated with chemotherapy prior to surgery
- Associated Htn
- Acquired Von Willebrand syndrome, 10%

Worry About

- Anomalies:
  - Aniridia 1%, hemihypertrophy 2%
  - Neurofibromatosis
  - Beckwith-Wiedemann syndrome
  - GU abnormalities, horseshoe-shaped kidney, cryptorchidism, gonadal dysgenesis, hypospadias, duplication of collecting systems
- Metastatic disease: Lymph nodes, lung, liver, brain

Overview

- Most common abdominal tumor of childhood; prognosis related to staging.
- Because of the location of the tumor, blood loss can be significant.
- Tumor is also associated with other congenital abnormalities, which may affect anesthetic and/or surgical management.
- Tumor extension into IVC and heart carries increased morbidity and mortality.

Etiology

- Embryonal neoplasm
- No consistent chromosome abn, although abnormalities in chromosomes 1 and 11 are common.
- Three genes associated with Wilms:
  - 11p13 interstitial deletion associated with Wilms-aniridia-growth retardation
  - 11p15.5 deletion associated with Beckwith-Wiedemann syndrome
  - Third locus not determined to be associated with familial Wilms

Usual Treatment

- Chemotherapy (with vincristine, actinomycin D, and adriamycin).
- Radiotherapy.
- Surgical removal of tumor: If tumor bilateral, surgery has focused on nephron-sparing procedures. (Procedure including biopsy followed by chemotherapy and delayed definitive resection.)
- Open or laparoscopic procedure.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Beckwith-Wiedemann syndrome	Obstructive airway secondary to large tongue	Direct exam	Blood glucose levels
CV	Htn Tumor extension into heart	Asymptomatic	Htn	ECG CT abdomen US renal vein/IVC Cardiac ECHO
RESP	Resp compromise	Abdominal distentions Metastatic disease Tumor embolization	Increased RR Hypoxemia	Pulse oximetry CT abdomen US renal vein/IVC Possible cardiac ECHO
GI	Gastric reflux	Increased intraop pressure Hx of reflux	Abdominal distention	Review CT scan
HEME	Von Willebrand syndrome	Unusual bleeding		Bleeding time Ristocetin platelet aggregation

Key References: Whyte SD, Mark Ansermino J: Anesthetic considerations in the management of Wilms' tumor, *Paediatr Anaesth* 16(5):504–513, 2006; Green DM: The evolution of treatment for Wilms tumor, *J Pediatr Surg* 48(1):14–19, 2013.

Perioperative Implications

Preoperative Preparation

- Htn-controlled.
- R/O renal vein and/or IVC tumor involvement.
- Evaluate for bleeding disorder.
- Evaluate cardiovascular function if prior chemotherapy Rx.

Monitoring

- Arterial cath may be indicated.
- CVP cath may be needed. especially if IVC and tumor extend mid-heart.
- For preexisting hematuria, Foley cath can aid in fluid balance.
- IV catheters above diaphragm; large-bore cath preferable.
- $ETCO_2$ to rule out air and/or tumor embolus.

Airway

- May be a problem if Beckwith-Wiedemann syndrome is present.

Preinduction/Induction

- Age-appropriate use of sedation.
- Rapid-sequence if increased intraabdominal pressure.
- Regional anesthesia; epidural or paravertebral block for postop pain.
- Preexisting chemotherapy may have cardiac depressant effect.
- IV access above diaphragm.

Maintenance

- Requires a prolonged procedure.
- Avoid N_2O.
- Maintain temperature.
- Increased third space fluid requirements.
- Procedure may be associated with large blood loss.
- Pulm function may be compromised, secondary to metastasis and/or tumor embolization, abdominal distention, and/or surgical traction.

Extubation

- Expected if temp maintained and pt hemodynamically stable.

Postoperative Period

- Administer pain control:
  - Multimodal anesthetic
  - RA (epidural or paravertebral)
- Third space fluid requirements.
- Htn may still be present.

Anticipated Problems/Concerns

- Risk of tumor and/or air embolus: If tumor extends into renal vein, IVC may have to be cross-clamped, the IVC opened, and the tumor removed.
- Intraop blood loss can be extensive.
- Periop implications.

Wilson Disease

Cobin D. Soelberg

Risk

- Incidence: 1:30,000.
- Slightly more common among Eastern European Jewish populations.
- Children and young adults tend to present with nonspecific GI symptoms.
- Adults tend to present with neurologic symptoms.

Perioperative Risks

- Increased risk of liver failure, kidney failure, and cardiac complications.
- 6–12% of all pts require liver transplantation.

Overview

- Presentation can vary widely. Hepatic symptoms tend to present prior to neurologic symptoms.
- Often nonspecific symptoms such as abdominal pain, nausea, and vomiting may occur. Rarely presents with acute liver failure.
- More commonly presents with elevated transaminases, hepatomegaly or hepatosplenomegaly, or mild jaundice.
- Can be diagnosed by presence of Kayser-Fleischer rings and low serum ceruloplasmin levels. In the absence of Kayser-Fleischer rings, diagnosis is more difficult, relying on free copper and liver copper concentrations.

Etiology

- Autosomal recessive.
- In the liver, copper is not passed to ceruloplasmin and so the liver has an excess of copper. Once this excess exceeds the ability of the liver to hold it, it is released in its free form into the blood. It then accumulates in tissues and causes damage.
- Copper balance is regulated through excretion of bile.
- Copper accummulation leads to hepatic cirrhosis
- Lenticular degeneration.

Usual Treatment

- D-penicillamine, an oral chelating agent
- Trientine in pts with adverse reactions to penicillamine
- Liver transplantation in rare cases of fulminant liver failure