Postoperative Period

- Common setting for unrecognized vitamin K deficient coagulopathy, given inadequate oral intake and aggressive antibiotic therapy.

Anticipated Problems/Concerns

- Oral vitamin K is often ineffective therapy in pts with GI disease or cholestatic disease.
- IV vitamin K should be administered in a diluent such as 0.9% isotonic sodium chloride and administered at a rate no faster than 1 mg/min to reduce the risk of an adverse reaction.
- FFP will only temporize VKDB unless a supplemental source of vitamin K is provided.
- Prolonged PT/INR related to liver disease often will not correct with vitamin K supplementation.

Von Hippel-Lindau Disease

David Hallsworth

Risk

- Rare; approximate incidence is 1:36,000.
- Usually occurs in young adults with complex multiple manifestations.

Perioperative Risks

- Pts with cerebral hemangioblastoma have a 23% incidence of VHLD; assess other systems carefully.

Worry About

- Space-occupying central nervous tumors (retinal and cerebellar hemangioblastomas in 60% of pts).
- Pheochromocytoma (7–20% pts) may be undiagnosed.
- Pregnancy and childbirth may dramatically change disease progression and symptom expression; multidisciplinary involvement essential.

Overview

- VHLD is a complex multisystem disorder, and pts frequently require anesthesia for surgical treatment of tumors and embolizations.
- Most common causes of death are renal cell carcinoma or complications from cerebral hemangioblastomas.

Etiology

- Autosomal dominant with variable expression, due to mutation of a tumor suppressor gene on chromosome 3p25–p26.
- The most common lesions are hemangioblastomas (benign vascular tumors) involving the retina, cerebellum, brainstem, spinal cord, adrenal glands, and kidneys. VHLD is also associated with pheochromocytoma, renal and pancreatic tumors, endolymphatic sac tumors of the middle ear, and papillary tumors of the broad ligament and epididymis.
- Type I pts are less likely to develop pheochromocytoma than type II.

Usual Treatment

- Surveillance and surgical management of tumors with or without radiotherapy.
- Manage active tumors and/or complications of treatment (e.g., pheochromocytoma, diabetes, steroid insufficiency, renal insufficiency).

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Retinal hemangioblastomas Glaucoma Hearing loss	Visual loss, blindness	Ocular microscope and pressure testing	Fluorescein angiography
CV	Erythrocytosis	History of venous thromboembolism		Full blood count
RESP	Cystic lung tumor	Chest pain, hemoptysis		CXR, CT
GI	Pancreatic cysts	Abdominal discomfort		US/CT
RENAL	Renal tumors and/or previous nephrectomy			Blood lytes and renal function tests Renal US/CT
ENDO	Pheochromocytoma Adrenal insufficiency due to adrenal resection Diabetes (due to previous pituitary surgery)		Complications of diabetes BP	Urinary catecholamines Plasma metanephrines/normetanephrines, adrenal imaging
CNS	Cerebellar hemangioblastoma Spinal cord hemangioblastoma	Headache, nausea, visual disturbance, motor and sensory deficit	Neurologic examination Ocular signs of raised ICP	CT brain MRI spine
PNS	Nerve root lesions are very rare			
MS	Limb weakness due to CNS tumors		Neurologic examination	

Key References: Plon SE, Jonasch E: Clinical features, diagnosis, and management of von Hippel-Lindau disease. In *UpToDate*, Atkins MB, Firth HV, Perrone RD, et al, editors: *UpToDate*, Waltham, MA (Accessed June 20, 2016); Hallsworth D, Thompson J, Wilkinson D, et al: Intracranial pressure monitoring and caesarean section in a patient with von Hippel-Lindau disease and symptomatic cerebellar hemangioblastomas. *Int J Obstet Anesth* 24(1):73–77, 2015.

Perioperative Implications

Monitoring

- Full invasive monitoring if pheochromocytoma is present or suspected.
- Consider ICP pressure monitoring if pt is symptomatic.

Induction

- Spinal and epidural anesthesia are relatively contraindicated if CNS/spinal tumors are present; discuss with neurosurgeons.

Maintenance

- TIVA has theoretical advantages on cerebral circulation and ICP if cerebral tumors present.

Adjuvants

- Intraop control of blood sugar

Postoperative Period

- May require HDU if complex comorbidity

Anticipated Problems/Concerns

- Surgery for one problem often complicated by other manifestations of the disease