Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Extraocular eye movements smooth on pursuit and jerky saccade; excessive drooling, dysarthria		Test muscles controlling extraocular movements and word/sentence formation	Swallow evaluation
CV	Fluctuating HR and BP	Review med list for CNS active meds and possible drug-drug interactions with anesthetic drugs	Compare HR/BP from supine to seated and standing	ECG
RESP	Aspiration pneumonia	Complaints of fever and chills	Chest auscultation and percussion	CXR, CBC
GI	Gastroparesis secondary to skeletal muscle incoordination and autonomic dysregulation	Dysphagia and early satiety		Swallow evaluation
RENAL	Possibly dehydrated	Recent poor PO intake	Dry mucous membranes	BUN/CR ratio ≥20; FNa <1
ENDO	Poor nutrition status	Poor PO intake frequently requiring feeding tube	Cachexia	Albumin, prealbumin
CNS	Choreiform motor movements, depression, and dementia	Erratic behavior; decreased independence with ADL; gait disturbance	Cognitive impairment and poor executive function on MMSE; erratic movements on extremity motor exam	CT or MRI to grade severity by extent of caudate atrophy
PNS	Dysautonomia	Symptoms of orthostatic hypotension or sexual dysfunction		
MS	Skeletal muscle incoordination primarily limbs and bulbar secondary to to striatal dysregulation			

Key References: Kivela JE, Sprung J, Southorn PA, et al.: Anesthetic management of patients with Huntington disease, *Anesth Analg* 110(2):515–523, 2010; Roos RA: Huntington's disease: a clinical review, *Orphanet J Rare Dis* 5:40, 2010.

Perioperative Implications

Preoperative Preparation

- Glycopyrrolate (0.2 mg/70 kg) in pts with excessive drooling
- Metoclopramide (10 mg/70 kg) with caution in pts with gastroparesis and dysphagia as this may worsen EPS

Monitoring

- Fluctuating HR; the pt can have bradyarrhythmias and tachyarrhythmias.
- End-tidal CO_2 waveform to be watched for steep upstroke, signifying bronchospasm and microaspiration.
- Pulse oximetry to evaluate for hypoxemia due to bronchospasm or V/Q mismatch (pneumonia or pneumonitis).
- Monitoring may be technically difficult owing to pt's erratic movement.

Airway

- Avoid awake procedures if airway is assessed as difficult; also avoid in advanced disease if ideal surgical field conditions are required.
- Pt may have sialorrhea.

Induction

- Consider RSI for aspiration risk; rocuronium 1 mg/kg is preferred over succinylcholine, which multiple studies link with a fluoride-resistant variant of pseudocholinesterase deficiency.

Maintenance

- Drug-drug interactions can occur between anesthetics and psychotropic drugs, such as hypotension with parkinsonian drugs (L-dopa and bromocriptine). Avoid sympathomimetics with MAO inhibitors. Dysrhythmias and orthostatic hypotension can occur with TCAs.

Extubation

- Corticobulbar dysfunction theoretically increases aspiration risk while anesthetics are lingering.

Adjuvants

- Sedative hypnotics (e.g., BZDs) help reduce EPS but increase drug-drug interactions with home psychotropic medications. They can also potentiate hypotension and may prolong emergence.

Postoperative Period

- Monitor for respiratory complications, fluctuations of HR and BP; expect behavioral challenges in advanced disease.

Anticipated Problems/Concerns

- Poor bulbar muscle coordination may increase risk for aspiration events and a subsequent respiratory complication, namely pneumonia.
- Drug-drug interactions between anesthetics and home psychotropic prescriptions may lead to hypotension, dysrhythmias, neuroleptic malignant syndrome, or worsening EPS.
- Succinylcholine should be avoided, as cases of pseudocholinesterase deficiency and prolonged muscle paralysis are reported.

Hydrocephalus

Joseph R. Tobin | Timothy E. Smith

Risk

- Found in newborns and children with anatomic CNS abnormalities (including myelomeningocele)
- Head trauma and intracranial hemorrhage (prematurity, SAH, other causes)
- CNS tumors
- Meningitis
- Recurrent VP shunt malfunction

Perioperative Risks

- Cerebral ischemia and neurologic sequelae
- Impaired airway reflexes,LOC, gastric emptying
- Cardiorespiratory arrest

Worry About

- Intracranial Htn
- Persistent N/V
- Bradycardia
- Decreased LOC

Overview

- Excess accumulation of CSF due to obstruction in normal CSF flow pattern from ventricular system to cortical surface (obstructive hydrocephalus), or from impaired reabsorption of CSF at arachnoid villi (communicating hydrocephalus).
- Slow progressive hydrocephalus can be well tolerated for weeks, with slowly worsening symptoms (headache, nausea, papilledema).
- Acute hydrocephalus results in acute symptoms and may be life-threatening, owing to herniation of brain with catastrophic ischemic injury (bradycardia, Htn, depressed LOC, depressed airway reflexes and resp drive, and gastric atony).

Etiology

- Congenital: Anatomic abnormalities, including aqueductal stenosis, Arnold-Chiari malformation, Dandy-Walker syndrome
- Posthemorrhagic/posttraumatic: Intraventricular hemorrhage (newborns or adults) with blood clot in ventricular system
- Neoplastic: Brain tumor obstructing normal CSF flow
- Postinflammatory: Meningitis, abscess, meningoencephalitis, intracranial hemorrhage

Usual Treatment

- Surgical correction of underlying cause or CSF diversion procedures (ventriculoperitoneal, ventriculoatrial, or lumboperitoneal shunts).

- Glucocorticoids are used acutely to diminish edema associated with neoplasm or abscess and may diminish associated intracranial Htn.
- Acetazolamide to diminish CSF production and reduce intracranial Htn.
- Furosemide to acutely decrease cerebrovascular volume.
- Mannitol to decrease ICP.

Assessment Points

System	Effect	Assessment by Hx	Physical Examination	Test
CV	Bradycardia, Htn	Late signs	Pulse, BP	
RESP	Impaired respiratory drive and airway reflexes		Cranial nerve exam, stridor, swallowing abnormality	Pulse oximetry
GI	N/V, aspiration, abnormal feeding	Hx of progression of N/V		
CNS	Depressed LOC, increased ICP, headache	Timing of onset	Arousability and neurologic exam, tense fontanel, inferior eye deviation	CT scan

Key References: Bober J, Rochlin J, Marneni S: Ventriculoperitoneal shunt complications in children: an evidence-based approach to emergency department management, *Pediatr Emerg Med Pract* 13(2):1–22, 2016; Christian EA, Melamed EF, Peck E, Krieger MD, McComb JG: Surgical management of hydrocephalus secondary to intraventricular hemorrhage in the preterm infant, *J Neurosurg Pediatr* 17(3):278–284, 2016.

Perioperative Implications

Preoperative Preparation

- Assess urgency of presentation. Catastrophic increased ICP requires emergent intubation and hyperventilation. In young infants, direct neurosurgical needle puncture of a proximal lateral ventricle or previously inserted shunt may diminish ICP sufficiently to avoid a catastrophe.
- Secure IV access if possible, and consider acetazolamide 10 mg/kg IV or furosemide 1 mg/kg IV.

Monitoring

- LOC
- Routine

Airway

- Head up 10-20 degrees and midline may diminish ICP
- Aspiration risk due to gastric atony

Preinduction/Induction

- Sedatives usually are not indicated so that resp compromise or sedation does not increase ICP. Minimal sedation or use of local anesthetic can secure IV access without causing increased ICP due to pain, crying, or struggling.
- Rapid-sequence IV induction is preferred (because of aspiration risk), unless in doubt of airway anatomy.
- Debate over use of succinylcholine versus rapid-onset nondepolarizing muscle relaxant (rocuronium). Thiopental, propofol, or etomidate IV agents preferred; avoid ketamine.
- Mask induction may increase ICP by increasing cerebral blood volume. Once fontanelles are closed, the brain is limited to a closed space within the cranium; prior to that time (<18 mo), the brain has some room to expand. Sevoflurane may be the preferable agent for inhalation induction (well tolerated and minimal effects on cerebrovascular tone). Isoflurane and desflurane are associated with coughing and are not recommended for induction.
- Lidocaine 1–1.5 mg/kg IV may be useful adjunct to minimize increase in ICP due to laryngoscopy and endotracheal intubation.

Maintenance

- Volatile anesthetic (most commonly sevoflurane or isoflurane) <1 MAC, N_2O 0–70% (debatable) and opioid (i.e., fentanyl 2–5 μg/kg or equivalent).
- Maintain normothermia, cardiac output. Hyperventilation may be acutely helpful until CSF is diverted and ICP reduced.
- Normal saline at restricted or maintenance rate. Glucose support should only be administered for infants; avoid hyperglycemia.

Extubation

- Ensure return of airway reflexes, LOC, and resp drive.
- Failure of achieving above criteria may require CT scan and/or ICU monitoring.

Postoperative Period

- Usually unremarkable; depressed LOC is concern for periop ischemic insult or hemorrhage.
- EBL should be minimal.

Adjuvants

- Lidocaine, mannitol, furosemide, and spontaneous hyperventilation by pt

Anticipated Problems/Concerns

- Immediate postop neurologic exam should demonstrate improvement. If not improved, urgent CT scan and secure airway must be maintained. Postop ICU admission not required unless impaired neurologic status continues.

Hyperaldosteronism, Primary

Marc Chikhani | Jonathan G. Hardman

Risk

- Responsible for up to 20% of moderate to severe systemic arterial Htn.
- End organ damage from long-standing Htn (e.g., chronic kidney disease, cardiomyopathy).
- Abnormal glucose tolerance in up to 50% of pts with hyperaldosteronism.

Perioperative Risks

- Hypernatremia and hypervolemia with high total body sodium.
- Htn may be refractory to treatment, with increased risk of cardiovascular complications, including malignant hypertensive crisis.
- Hypokalemia and hypomagnesaemia with low intracellular potassium and magnesium may cause cardiac arrhythmia and general muscle weakness.

Worry About

- Hypertensive response to intubation or surgical incision
- Hypokalemia and associated muscle weakness or potential for arrhythmia
- Metabolic alkalosis

Overview

- Also known as Conn syndrome; described by Jerome W. Conn, University of Michigan, in 1955.
- Characterized by Htn, hypernatremia, hypokalemia, metabolic alkalosis, and low plasma renin level.
- Classically caused by a unilateral aldosterone producing adrenal adenoma.
- Primary hyperaldosteronism is a renin-independent and incompletely suppressible over secretion of the mineralocorticoid aldosterone secreted from the zona glomerulosa of the adrenal cortex.
- Aldosterone acts on the mineralocorticoid receptor in the distal convoluted tubule of the nephron and the collecting ducts to enhance sodium and water reabsorption, at the expense of potassium. Excess loss of potassium leads to loss of hydrogen ions to maintain electroneutrality.
- Usually aldosterone secretion is controlled by the renin-angiotensin feedback system in response to thirst, hypovolemia, reduced renal juxtaglomerular apparatus perfusion pressure, and reduced tubular sodium concentration.
- Aldosterone promotes restoration of circulating volume by correcting water and sodium depletion.
- Dx is by combination of clinical suspicion of persistent Htn, hypernatremia and spontaneous hypokalemia, and metabolic alkalosis in the absence of diuretics. Dx is confirmed by measuring the plasma aldosterone to renin ratio—a value over 35 ng/dL per ng/mL/h has sensitivity of 100% and specificity of 92%. Post test specificity can be improved by measuring post-sodium infusion aldosterone. Values above 7 ng/dL showed specificity of 100%.

Etiology

- 60–70% idiopathic hyperaldosteronism or bilateral idiopathic adrenal hyperplasia
- 30–40% unilateral aldosterone-producing adrenal adenoma (first described by Conn in 1955)
- Uncommon causes:
  - Unilateral adrenal hyperplasia
  - Familial hyperaldosteronism
  - Aldosterone producing adrenocortical carcinoma and ectopic aldosterone secreting tumors

Usual Treatment

- Aldosterone antagonists such as spironolactone (up to 300 mg/d) or the newer drug eplerenone (up to 100 mg/d).
- Both spironolactone and eplerenone are mineralocorticoid receptor antagonists and therefore have a potassium-sparing diuretic effect. Eplerenone is more selective for the mineralocorticoid receptor than spironolactone and has fewer glucocorticoid or androgen receptor antagonist side effects.
- Htn may be refractory to treatment with aldosterone antagonist alone and may require formal