Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Soft tissue distortion of airway, loose teeth, mucosal ulceration	Stridor	Airway and dental evaluation	
RESP	Spontaneous pneumothorax, reactive airways, infiltrates, fibrosis, pulm Htn	Tachypnea, dyspnea, cough, smoking history		CXR, ABG, PFTs, CT with cysts or nodular infiltrate
GI	Ulceration, obstruction, hepatic dysfunction		Jaundice Hepatomegaly	Bilirubin, albumin AST, ALT, INR
CNS	Diabetes insipidus, neuropathy, exophthalmos	Polyuria, polydipsia	Neuro exam	Urine and serum Osm, lytes
HEME	Thrombocytopenia, anemia, leukopenia	Bruising or bleeding	Splenomegaly	CBC

Key References: Morimoto A, Oh Y, Shioda Y, et al.: Recent advances in Langerhans cell histiocytosis, *Pediatr Int* 56(4):451–461, 2014; Broscheit J, Eichelbroenner O, Greim C, et al.: Anesthetic management of a patient with histiocytosis X and pulmonary complications during Caesarean section, *Eur J Anaesthesiol* 21(11):919–921, 2004.

Perioperative Implications

Monitoring

- Routine
- Foley in pt with DI
- Arterial cath for ABG in those with pulm involvement, frequent Na checks in those with DI, as well as pulse pressure variation as a marker of volume status in those with DI

Preinduction/Induction

- Airway soft tissue or mandibular involvement may distort anatomy.
- Cervical vertebrae lesions may cause cervical instability.
- Ensure adequate preoxygenation, especially if there is significant pulm involvement.
- Usual precautions, depending on severity of organ involvement.

Maintenance

- For pts with DI, consider aqueous ADH infusion and isotonic crystalloid fluids.
- Stress dose steroids if pt has had steroid therapy.
- Usual precautions, depending on severity of organ involvement.

Extubation

- Awake extubation if anatomy is distorted and airway was difficult for mask ventilation or intubation.
- Severe pulm involvement may delay extubation.

Regional Anesthesia

- Follow ASRA precautions if thrombocytopenic or elevated INR.
- Use caution with interscalene and supraclavicular blocks in pts with pulm disease.

Postoperative Period

- May need continued stress dose steroid coverage for several days postop.
- Closely monitor oxygenation and ventilation when pulm disease present, and evaluate for pneumothorax.

Anticipated Problems/Concerns

- Organ dysfunction (hepatic, pulm, hematologic, hypothalamic, or bone).
- DI.
- Adrenal suppression due to chronic steroid therapy; may experience intraop hypotension without stress steroids.
- Severe pulm involvement may increase risk of pneumothorax and complicate extubation.

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Huntington Disease

David A. Wyler

Risk

- General prevalence: 5-7:100,000
- Highest prevalence in Caucasians of western European descent

Perioperative Risks

- Increased risk of respiratory complications secondary to bulbar muscle incoordination
- Autonomic dysfunction

Worry About

- Microaspiration, bronchospasm, chemical pneumonitis, and aspiration pneumonia
- Drug-drug interactions with anesthetic drugs and psychotropic medications
- Prolonged effects with succinylcholine
- Dysautonomia, gastroparesis, and fluctuating HR and BP

Overview

- Inherited progressive neurodegenerative disease of the CNS, primarily the basal ganglia.
- More common adult-onset variant begins in the fifth decade and leads to complete disability and death within 20 y.
- Heterogeneous presentation of dysregulation of motor coordination, cognitive decline, and psychiatric manifestations.
- Classically known for choreiform (repetitive, rapid, jerky, involuntary) movements from degeneration of GABAergic neurons of the basal ganglia specifically of the striatum (caudate and putamen).
- Chorea, early motor sign along spectrum; progresses to parkinsonian-like movements (bradykinesia, rigidity, and postural instability) late in the adult-onset disease.
- Worsening subcortical dementia (declining executive function and cognition without amnesia) and severe depression accompany disease progression.
- Juvenile variant presents with parkinsonian signs at onset, lacks choreiform movements, and has least favorable prognosis along spectrum.
- Skeletal muscle incoordination of the laryngeal and pharyngeal muscles leads to devastating respiratory sequelae and death.
- See also Parkinson Disease.

Etiology

- Autosomal dominant inheritance.
- Trinucleotide repeat expansion of CAG codon on the IT15 gene on chromosome 4 results in the overproduction and aggregation of the protein Huntingtin.
- Length of repeat correlates well with extent of Huntingtin production, disease severity, and age of onset.
- Huntingtin accumulates in the nuclei and cytoplasm of all CNS neurons; degeneration occurs most notably in vulnerable neurons of the caudate and putamen.
- Striatal cell death occurs by glutamate- and dopamine-induced excitotoxicity, oxidative stress, impaired energy metabolism, and apoptosis.

Usual Treatment

- No definitive cure; treatment is supportive, focusing on alleviation of symptoms.
- Early symptoms of chorea are treated with neuroleptics, dopamine-depleting medication. Surgical implantation of deep brain stimulators may be helpful.
- Gene-modifying therapy is currently under investigation.

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).