Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Difficult airway	C-spine trauma/surgery H/O; difficult intubation	↓ C-spine ROM ↓ Mouth opening	Airway exam
CV	Orthostatic hypotension Baseline relative hypotension (15–20 mm Hg)	H/O dizziness when going from supine to upright position	↓ BP, orthostasis, tachycardia, bradycardia, AFIB	Orthostatic BPs ECG
RESP	Decreased resp volumes, atelectasis, pneumonia, hypoxemia Impaired cough reflex	SOB Difficulty w/secretions	Tachypnea Cyanosis Decreased/unequal BS	CXR ABG evaluation
GI	Full stomach status due to GI atonicity	Complaints of reflux		
RENAL	UTI, renal stone disease, renal failure	Flank pain	Chronic Foley catheter	UA and BUN/Cr
CNS	Bowel and bladder dysfunction Chronic and central pain states Altered MS (if severe head trauma)	Incontinence Chronic opioid therapy Adjuvant pain meds	Hyperreflexic below level of transection Babinski sign positive	Hyperalgesia Allodynia
PNS	Insensate below level of transection Pain at level of transection	Skin-color changes	Flushing/piloerection above Dry, pale skin below	
MS	Paralysis, muscular atrophy below Sacral decubiti	Paraplegia or quadriplegia	Muscle atrophy Sacral decubiti	

Key References: Krassioukov A: A systematic review of the management of autonomic dysreflexia after spinal cord injury, *Arch Phys Med Rehabil* 90:682–695, 2009; Liu N: Iatrogenic urological triggers of autonomic dysreflexia: a systematic review, *Spinal Cord* 53(7):500–509, 2015.

Perioperative Implications

Preoperative Preparation

- Nifedipine can be used for prophylaxis; given 30 min before procedure, likely to trigger AH.
- Attention to CV and pulm function, volume status, and airway exam.

Monitoring

- Consider preinduction invasive monitoring (arterial and CVP/PA catheters) if volume changes are expected and in setting of poor cardiac reserve (high lesions) and renal insufficiency.

Airway

- Be prepared for fiberoptic intubation.

Induction

- Use nondepolarizing muscle blockers when relaxation is necessary.
- IV nicardipine can be used to treat AD.
- Succinylcholine can cause severe K^+ release and hyperkalemia in chronic lesions.
- Consider nitroprusside before induction.

Maintenance

- GA with volatile agent superior to nitrous-narcotic technique for prevention/treatment of AD.

Regional Anesthesia

- Anesthetic technique of choice when possible.
- Spinal anesthesia highly effective in preventing AD precipitated by surgery.
- Ensure careful assessment of level of spinal blockade in SCI pts due to sensory deficits below injury: avoid unnecessarily high or inadequate blocks.
- Epidural anesthesia effective in preventing AD in laboring pts.

Extubation

- May be difficult due to resp insufficiency in pts with high-level spinal lesions

Adjuvants

- Muscle relaxants required in abdominal surgery due to diffuse increase in muscle tone

Postoperative Period

- AD can occur postop in setting of unrecognized or untreated distended bladder or rectum.
- Consider intracerebral hemorrhage protocol in the setting of unexplained delayed emergence with increased BP.

Becker Disease

Pikulkaew Dachsangvorn

Risk

- Prevalence is approximately 1:50,000

Perioperative Risks

- Myotonia

Worry About

- Myotonic episode leading to a difficult to ventilate/intubate situation

Overview

- Genetic disease that results in muscle membrane hyperexcitability and delayed relaxation
- Recessively inherited form of MC
- Initial symptoms start around 4–12 y of age, with generalized myotonia and moderate to pronounced muscular hypertrophy from chronically increased muscle activity
- Signs include muscle stiffness after voluntary contraction that improves with repetitive movement ("warm-up" phenomenon) and worsens after prolonged rest
- Many experience transient weakness (<1 min) upon initiating movement; history of clumsiness, dropping objects, impaired postural control, or uncontrolled falling upon standing
- Rarely, can have atrophy in the forearms and painful muscle cramps
- Most have normal life expectancy without significant handicap
- Aggravating factors: dietary insufficiencies, sleep deprivation, prolonged physical activity, and emotional stress
- Menstruation, pregnancy, and hypothyroidism may alleviate or worsen symptoms in some individuals
- No involvement in smooth and cardiac muscles, no extramuscular manifestations
- It is important to differentiate this from myotonia with dystrophy, which is a multisystem disorder
- Diagnosis:
  - Characteristic symptoms (described previously)
  - "Percussion myotonia": reflex hammer produces obvious dimpling or fasciculation in prominent muscles, such as thenar eminence or thighs, that lingers for several seconds
  - Objective evidence: electromyography
  - Molecular genetic testing is commercially available, although not sensitive for less common mutations

Etiology

- Impaired functioning of skeletal muscle ClC-1
- Skeletal muscle chloride channels serve to stabilize membrane potential at the resting level; impaired ClC-1 leads to sarcolemmal excitability and delayed muscle relaxation
- More than 120 mutations have been described; most mutations are unique to individual families or isolated cases

Usual Treatment

- Most pts prefer to minimize their symptoms by avoiding triggers.
- Pharmacologic therapies include quinidine and quinine, which are effective and well tolerated in low-dose, short-term use; however, continued administrations can lead to toxicity affecting vision, hearing, gastrointestinal, central nervous systems, and possibly causing death.
- Other drugs with variable success include procaine, tocainide, mexiletine, carbamazepine, and phenytoin, by use-dependent blockade of voltage-gated sodium channels.

- Directed pharmacologic approach to increase chloride conductance of skeletal muscle includes taurine and clofibric acid; however, the effect is modest.
- Other interventions include optimizing pt's emotional state and relaxation techniques. Some subjects have shown improvements with alcohol use. Exercises that improve flexibility and decrease muscle strains can be helpful.
- Gene therapy to introduce a functional copy of the normal gene has been considered; however, this may not be effective in disorders caused by a dominant-negative mechanism.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
No effects on CNS, CV, RESP, GI, GU, ENDO, HEME/ID systems because BD only affects skeletal muscle membranes.				
HEENT	Blepharospasm (myotonia of the eyelids)			
MS	Delayed muscle relaxation that is resistant to NDMR and DMR	Obtain full Hx regarding symptoms and aggravating factors	Upper-extremity atrophy Muscular hypertrophy in other areas of the body Reflex hammer test	EMG

Key References: Bandschapp O, Iaizzo PA: Pathophysiologic and anesthetic considerations for patients with myotonia congenital or periodic paralyses, *Pediatr Anesth* 23(9):824–833, 2013; Dunø M, Colding-Jørgensen E: Myotonia congenita. In Pagon RA, editor: *GeneReviews*. Available at <http://www.ncbi.nlm.nih.gov/books/NBK1355/>. (Accessed 23.02.16.)

Perioperative Implications

Preoperative Preparation

- Keep pt normothermic throughout pre-/intra-/and postop periods because shivering can trigger myotonic episode.
- DMR, NDMR, and regional anesthesia are ineffective in minimizing myotonic contractions because the defect lies within the muscle membrane.
- Regional anesthesia with peripheral nerve stimulation in combination with fentanyl and midazolam sedation for shoulder surgery has been used successfully without complication.

Monitoring

- Neuromuscular monitoring is mandatory.
- Core temperature monitoring is recommended.

Airway

- Can be difficult to ventilate/intubate if myotonia was elicited during induction/extubation.

Preinduction/Induction

- Consider administering IV lidocaine prior to propofol induction as pain associated with propofol injection can lead myotonia.
- Avoid depolarizing muscle relaxant (succinylcholine), as it has been shown to provoke severe generalized muscle stiffness, including masseter spasm and decerebrate posturing, making intubation and ventilation difficult to impossible.
- Response to NDMR appears to be normal; however, consider reducing the dose of NDMR in pts with associated muscle wasting.

Maintenance

- Consider short-acting NDMR and allow pt to recover fully from muscle relaxant without reversal because anticholinesterase can precipitate myotonia.
- Currently, there are no data on reversal of rocuronium by sugammadex for MC pts.

Extubation

- Consider avoiding anticholinesterase use.
- Avoid coughing on extubation.

Postoperative Period

- Continue to maintain normothermia and adequate pain control.

Anticipated Problems/Concerns

- No association with malignant hyperthermia as previously suggested.
- In rare cases, epinephrine or selective beta-adrenergic agonists in high doses may aggravate myotonia. Beta-antagonist propranolol has also been reported to worsen myotonia.

Beckwith-Wiedemann Syndrome

Arlyne K. Thung | Lee A. Fleisher

Risk

- 1 per 13,700 individuals.
- No gender predilection, although with monozygotic twins it is seen more in females than males.
- Conceptions from IVF have a 3–5 times increased risk of BWS.

Perioperative Risks

- Acute airway obstruction; difficult mask ventilation and intubation secondary to macroglossia
- Hypoglycemia due to islet cell hyperplasia and hyperinsulinemia
- Cardiac malformations

Worry About

- Persistent hypoglycemia, which may cause CNS damage; therefore intraop infusion of a glucose-containing solution and frequent glucose checks are required.
- Difficult airway management.

Overview

- Commonly known for the triad of EMG.
- Other clinical features include anterior earlobe creases, posterior helical pits, facial nevus flammeus, hemihyperplasia, renal anomalies, embryonal tumors, cardiac malformations, and hypoglycemia.
- 7.5% estimated risk for embryonal tumor development, which occurs in the first 10 y of life. Most common tumors are Wilms tumor and hepatoblastoma but may also include rhabdomyosarcoma, adrenocortical carcinoma, and neuroblastoma.
- Cardiac involvement often limited to mild cardiomegaly, although other cardiac defects have been reported (atrial and ventricular septal defects, tetralogy of Fallot, hypoplastic left ventricle, cardiomyopathy, cardiac tumors, and valvular disease).
- Hypoglycemia due to islet cell hyperplasia and hyperinsulinemia occurs in 50% of BWS pts, is often responsive to medical therapy, and usually regresses during the first 4 mo of life. Persistent hypoglycemia refractory to medical management may require pancreatectomy.

Etiology

- Clinically and genetically heterogeneous.
- May be genetically transmitted (15%) or occur sporadically (85%).
- Variety of mutations in chromosome 11p15.5 region.
- Mutation near gene for IGF-II.

Usual Treatment

- Prenatal detection of polyhydramnios, omphalocele, placentomegaly, macrosomia, macroglossia, and renal anomalies on fetal US may prompt genetic testing and counseling if BWS is suspected.
- Screening for hypoglycemia in the first few days of life if BWS is suspected. Surgical intervention if hypoglycemia persists despite medical management.
- Surgical repair of omphalocele.
- Possible reduction of macroglossia in the first year of life to avoid complications of airway obstruction, feeding, and speech difficulties.
- Infants with hypoglycemia and severe oral intolerance due to macroglossia may require gastrostomy tube placement as a temporizing measure until regular feeds become possible after glossal resection.
- Orthopedic follow-up to monitor leg-length discrepancies due to hemihyperplasia.
- Tumor surveillance (abdominal US, alpha-fetoprotein).
- Surgical resection of operative tumors.