Usual Treatment

- Assessment of pt via advanced trauma life support protocol with focus on spinal stabilization and immobilization on board and/or cervical collar to prevent further injury.
- Consider timely surgical intervention if appropriate; injury is typically irreversible after 48 h.
- Maintain adequate spinal cord perfusion via adequate fluid resuscitation and vasopressors, if needed.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CNS	Neurologic deficits, autonomic dysreflexia, altered mental status, neurogenic shock, spinal shock	Weakness, headache, hyperhidrosis	Thorough neuro exam	Review imaging
HEENT	Cervical cord injury, inability to tolerate secretions	Pain, choking, drooling, weakness	Neurologic exam, c-collar/halo vest	Early intubation with in-line stabilization, CT scan
RESP	PE, hypoventilation, pulm edema	SOB, tachypnea, hemoptysis	Decreased chest rise/ breath sounds, use of accessory respiratory muscles	CXR, ABG, PFTs
CV	Hypotension, conduction abnormalities, low baseline BP	Dizziness, nausea, fatigue	Orthostasis	ECG, invasive BP monitoring
GI	GI atony, GI bleeding/stress ulcer, ileus	Nausea, heartburn, melena, abdominal pain	Abdominal TTP, bloody stool, absent bowel sounds	NGT, GI prophylaxis
RENAL	Frequent UTI, renal dysfunction, lyte disturbance, priapism	Dysuria, hematuria	Urinary cath (often suprapubic)	BMP
HEME	Anemia, hypercoagulability	Dizziness, easy bleeding/bruising, h/o blood clot	Visible purpura, bruises, swollen/ tender limb(s)	VTE prophylaxis CBC, coagulation, US if warranted
MS	Bone fractures, decubitus ulcers, skeletal muscle spasticity/contractures	Chronic muscle spasms, areas of skin breakdown	Thorough MS exam	Imaging if necessary

Key References: Stevens RD, Bhardwaj A, Kirsch JR, et al.: Critical care and perioperative management in traumatic spinal cord injury, *J Neurosurg Anesthesiol* 15(3):215–229, 2003; Oropello JM, Mistry N, Ullman JS: Spinal injuries. In Hall JB, Schmidt GA, Kress JP, editors: *Principles of critical care,* ed 4, New York, 2015, McGraw-Hill.

Perioperative Implications

Preoperative Preparation

- Perform thorough neurologic examination.
- Ensure adequate volume resuscitation.
- Strongly consider RA if appropriate.

Monitoring

- Standard ASA monitors.
- May consider invasive hemodynamic monitoring, including arterial line and CVP if instability anticipated.
- Monitor temp closely, as hypothermia/hyperthermia may occur.

Airway

- Consider advanced airway techniques (i.e., fiberoptic intubation/video laryngoscopy) if neck extension is limited due to cervical injury or c-collar/ halo vest.

Preinduction/Induction

- Rapid sequence intubation, particularly in acute spinal cord injury, due to high risk of respiratory compromise, gastric atony/dilation, and aspiration.
- Be sure pt is adequately anesthetized prior to endotracheal intubation to avoid autonomic dysreflexia.
- Avoid significant hypotension.
- Pt may have increased resistance to NDMBs.
- Avoid succinylcholine, as it may result in severe bradycardia and hyperkalemia.
- Atropine should be administered if succinylcholine must be used.

Maintenance

- Keep MAP >80 mm Hg when possible for adequate perfusion to spinal cord.
- Maintain adequate analgesia to avoid autonomic dysreflexia.
- Have short-acting vasopressors and antihypertensives readily available.

Extubation

- Use caution.
- Depending on level lesion, and risk respiratory muscle paresis and/or paralysis, long-term mechanical ventilation may be indicated.

Adjuvants

- Consider body warmer and/or warmed IV fluids.
- Careful padding of pressure points/decubitus ulcers.
- NGT for decompression, given risk of gastric atony.

Postoperative Period

- Due to increased risk of hypoxia, pt should receive continuous supplemental oxygen and pulse oximetry monitoring, aggressive pulmonary toilet.
- Low threshold for reintubation, particularly in higher lesions.
- Increased risk of visceral pain, phantom pain, and muscle spasms.

Anticipated Problems/Concerns

- Autonomic dysreflexia; if left untreated, can lead to myocardial ischemia and potentially cardiac arrest.
- Make it a priority to minimize inciting factors.
- Pts are at increased risk of developing VTEs, which may lead to pulm embolism.
- Pt should be on VTE prophylaxis and GI ulcer prophylaxis if there is no contraindication.

Spinal Muscular Atrophy

Karim El Harchaoui

Risk

- Incidence: 7.8 -10:100,000 live births.
- Estimated panethnic disease frequency: ~1:11,000.

Perioperative Risks

- Airway: Intubation can be difficult.
- Pts may need postop respiratory support.
- Anesthetic risk varies significantly between the different types of SMA.

Worry About

- Pts may display increased sensitivity and prolonged effect of neuromuscular blockers.
- Postop respiratory depression could be catastrophic.
- Concomitant pulmonary disease and bulbar dysfunction.
- Gastroesophageal reflux.
- Prolonged effects of nondepolarizing neuromuscular blockers (even after reversal).
- Neuromuscular monitoring can be unreliable.

Overview

- SMA is an autosomal recessive neuromuscular disease.
- The disorder leads to degeneration of the anterior horn cells of the spinal cord, which causes muscle weakness.
- The rate of progression varies between pts and is classified in four categories:
  - SMA I (Werdnig-Hoffman disease): Onset of symptoms before 6 mo of age
  - SMA II (Dubowitz type): Onset between 6 and 18 mo
  - SMA III (Kugelberg Welander disease): Present later in childhood
  - SMA IV (adult type): Onset during middle or late age
- Prognosis for long-term survival depends on the type and ranges from neonatal death to onset of weak muscles in adulthood.
- Respiratory failure is the major cause of mortality.
- Scoliosis and chest wall muscle weakness may predispose to pulmonary dysfunction.
- Normal intellectual and emotional capacity.

Etiology

- Autosomal recessive inheritance can occur with deletions or mutations in the survivor motor neuron genes located on chr5q13.
- The loss of full-length SMN protein leads to degeneration of anterior spinal motor neurons and, in severe cases, degeneration of brainstem nuclei.
- Degeneration of spinal anterior neurons and brainstem nuclei correspond to a range of clinical characteristics, including global hypotonia, pulmonary insufficiency, and autonomic and bulbar dysfunction.

Usual Treatment

- There is no cure for SMA.
- Supportive treatment as required, including physiotherapy and orthopedic intervention, to prevent contractures and maximize respiratory function.
- Low threshold for antibiotic use during acute illnesses due to the risk of pneumonia.

Classification of Spinal Muscular Atrophy

Type	Age at Onset	Highest Motor Milestone Achieved	Lifespan Without Treatment	Symptoms	Affected Organ
Type I Werdnig-Hoffman disease	Birth–6 mo	Never sits unsupported	<2 y	Progressive muscle weakness, respiratory failure, hypotonia, reduced bulbar function	Muscular: Respiratory
Type II Dubowitz disease	6–12 mo	Sits independently, never stands or walks	70% reach adulthood	Progressive onset of proximal limb weakness in infancy Legs > arms Scoliosis Joint contractures	Muscular: Kyphoscoliosis Joint contractures
Type III Kugelberg-Welander disease	>18 mo	Stands and walks	Normal lifespan	Onset of proximal weakness during childhood Legs > arms Scoliosis Increased risk of fractures	Muscular: Joint problems
Type IV Adult SMA	>5 y to mostly >30 y	Normal	Normal	Onset of proximal leg weakness in adulthood	Muscular

Key References: Islander G: Anesthesia and spinal muscle atrophy, *Paediatr Anaesth* 23(9):804–816, 2013; Darras BT: Spinal muscular atrophies, *Pediatr Clin North Am* 62(3):743–766, 2015.

Perioperative Implications

Preoperative Preparation

- Preop pulm evaluation and pulm function testing.
- Evaluate intubation conditions.
- Start air-stacking techniques preop.
- Make a preop and postop plan. Pt may require postop ventilator support.

Monitoring

- When nondepolarizing muscle relaxants are used, the effect should be monitored carefully both clinically and with a monitor of neuromuscular transmission and muscular contraction.
- Consider ABG.

Airway

- Difficult intubation can occur due to limited mobility of the cervical spine and reduced mouth opening.
- Pt may present with artificial ventilation (NIV).
- Awake fiberoptic intubation could be the technique for intubation in pts with restricted neck movements.

Preinduction/Induction

- No specific anesthetic drug is recommended.
- Laryngeal mask may be appropriate in superficial surgery.
- Peripheral neural blockade may be useful.
- Avoid succinylcholine due to the risk of inducing rhabdomyolysis and hyperkalemia.
- Nondepolarizing muscle relaxants are suitable but should be titrated carefully since sensitivity to these drugs appears to vary.
- Approach when choosing anesthetic techniques and agents:
  - Minimize modifications of chest wall dynamics due to residual muscle relaxants effect or high level of neural axis blockade.
  - Avoid excessive depression of central respiratory drive.

Maintenance

- Both TIVA and inhalation agents may be used.
- Pts with SMA are not at increased risk for malignant hyperthermia.
- Short-acting opioids are suitable for intraop use.
- Continuous infusion of local anesthetic solutions via peripheral nerve block cath should be considered as safer alternatives to systemic opioids.
- Wound infiltration anesthesia is recommended whenever possible.

Extubation

- Muscle strength must be evaluated before extubation, not only with train-of-four stimulation but also clinically.
- Reverse neuromuscular blockade with sugammadex.

Postoperative Period

- Pts with SMA I need postop ventilator support.
- Some pts with SMA II and III will require respiratory support during acute illness or in advanced disease; NIV for bridging from intubation to spontaneous breathing.
- Use oxygen with caution because too much oxygen can mask hypoventilation due to muscle weakness.
- Postop pain management must be individualized and multimodal. Acetaminophen and ibuprofen are useful.

Anticipated Problems/Concerns

- Opioid-induced respiratory depression is dangerous in SMA pts with weak muscles. Careful monitoring is mandatory.
- The major concern related to the response from anesthesia is prolonged impairment of neuromuscular function and suppression of central respiratory drive, which can compromise the limited pulm reserve leading to acute respiratory failure.
- Neuraxial anesthesia can be difficult or unreliable due to altered spine anatomy (severe scoliosis).

Stevens-Johnson Syndrome

Lauren M. Nakazawa | Lee A. Fleisher

Risk

- Incidence of SJS and TEN, a more severe variant of SJS, is 2–7 cases per million per y.
- Incidence around 100 times higher in the HIV-positive population.
- More common in women.
- Affects all age groups.

Perioperative Risks

- High risk for infection
- Hypovolemia
- Cutaneous, mucosal, and ocular injury
- Respiratory failure requiring mechanical ventilation in around 25% of pts

Worry About

- Sepsis and septic shock
- Fluid and lyte imbalances
- Development of multiorgan failure
- Disease recurrence if culprit drug is readministered

Overview

- Severe cutaneous reaction with epidermal necrosis and detachment in conjunction with mucosal and conjunctival involvement.
- SJS and TEN fall along a disease continuum. SJS is less severe, involving <10% total BSA. TEN involves >30% BSA, and SJS-TEN overlap involves 10–30%.
- Clinical presentation:
  - Prodrome: Fever, flu-like symptoms (malaise, myalgia, arthralgia), skin pain/tenderness, oral pain, photophobia, and conjunctival burning can be early signs of mucosal involvement.
  - Cutaneous lesions: Diffuse erythema or erythematous macules starting on trunk and face and developing central necrosis and bullae formation with eventual sloughing off of epidermis and exposed dermis.
  - Mucosal involvement in 90% of pts.