Worry About
- Difficult pt positioning secondary to sustained muscle contractions
- Difficult intubation as a result of poor extension of the cervical spine and diminished mouth opening

Overview
- ST is defined as twisting of the neck caused by involuntary muscle contractions.
- Idiopathic ST is a slowly progressive disease that manifests between the third and fifth decades. Idiopathic ST is likely caused by abn of the basal ganglia circuitry.
- Dystonia typically progresses over 3–5 y before it plateaus.
- Pain occurs in 75% of pts and contributes to disease disability.
- If ST occurs acutely, it is necessary to rule out causes related to trauma, medications (metoclopramide, haldol, phenothiazines), intracranial abnormalities (tumors, AVMs, hemorrhages), and neck pathology (retropharyngeal abscess).
- The sternocleidomastoid and trapezius muscles are most commonly involved, but extracervical dystonia may occur in 20% of pts.
- Jerking of the head and head tremors are common features.
- Head positioning determines the type of torticollis:
  - Rotational torticollis: Rotation of the head around the long axis of the neck.
  - Anterocollis: Head tilts forward with neck flexion.
  - Retrocollis: Head tilts backward with neck extension.
  - Laterocollis: Head tilts to one side with the ear pulled toward the shoulder.

Etiology
- A genetic component probably contributes to the development of ST since it is a familial pattern.
- Trauma, medications, and intracranial pathology can cause focal dystonic reactions such as torticollis.

Usual Treatment
- Chemical denervation with botulinum toxin is the mainstay of therapy. Botulinum toxin is injected into overactive muscles in the neck that are responsible for the dystonia. It usually takes a week to take effect and lasts up to 3 mo before a repeat injection must be given.
- Pharmacologic therapy with anticholinergics, benzodiazepines, Parkinson medications, and baclofen are used as adjuncts to botulinum toxin.
- Surgical options include mechanical denervation of affected muscles, deep brain stimulation, and intrathecal baclofen if spasticity is prominent.

Assessment Points

System	Effect	Assessment by Hx	PE
HEENT	Head deviation	Twisting, pulling sensation	Hypertrophic SCM and trapezius
RESP	Restrictive lung disease	Dyspnea	
GI	GERD	Food regurgitation; pain after meals	
CNS	Diplopia Difficulty with transfers Aspiration risk	Vision deficits Coughing with food Use of walker, cane, wheelchair	Abnormal eye movements. facial droop, depressed gag, tremor, spasticity of muscles, weakness

Key References: Mac TB, Girard F, McKenty S, et al.: A difficult airway is not more prevalent in patients suffering from spasmodic torticollis: a case series, *Can J Anaesth* 51(3):250–253, 2004; Patel S, Martino D: Cervical dystonia: from pathophysiology to pharmacotherapy, *Behav Neurol* 26(4):275–282, 2013.

Perioperative Implications
Preinduction/Induction/Maintenance
- Routine considerations.
- Consider the use of nondepolarizing muscle relaxants.
- NMB may have no effect on muscle contractures, which are permanently shortened muscles that result from structural muscle changes.
- Anticipate the use of fiberoptic intubation.

Preoperative Considerations
- Consider preop injections of Botox at least 1 wk prior to anesthesia.
- It is imperative to evaluate the range of cervical spine extension, maximal mouth opening, and integrity of the temporomandibular joint.

Monitoring
- Routine

General Anesthesia
- Propofol is likely to be safe with all dystonias.
- GA with thiopental, succinylcholine, atracurium, isoflurane, and fentanyl is thought to be safe in spasmodic torticollis.

Regional Anesthesia
- Limited reports but thought to be safe
- Postop period
- Risk of aspiration

Anticipated Problems/Concerns
- Anticipate difficult intubation secondary to fixed head turning from muscle contractures that do not respond to muscle relaxants.
- Be aware of cervical spine pathology that may result from prolonged torticollis.
- Neurologic conditions such as cranial nerve dysfunction and seizure disorders may accompany secondary ST caused by an underlying intracranial lesion.
- ST can cause head tremors, which should not be confused with hyperkinetic movement disorders.

Spinal Cord Injury

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Risk
- In USA, trauma is the number one cause.
- Approx 10,000–12,000 cases per y.
- Males (80%) primarily affected.
- Motor vehicle collisions, falls, violence (GSW), sports-related injuries, hematoma, and transverse myelitis most common.

Perioperative Risks
- Hypotension, particularly in acute injury resulting in neurogenic shock and concurrent hypovolemia
- Autonomic instability, which may result in severe cardiopulmonary compromise.

Worry About
- Autonomic dysreflexia (T6 or above, but as low as T12 in some; usually seen 1–6 mo after injury)
- Hypoventilation leading to acute respiratory compromise and chronic infections
- Irregular thermoregulation

Overview
- Injury to the spinal cord at level of the cervical or thoracic vertebrae resulting in loss of underlying sympathetic stimulation, unopposed vagal tone, and sensory and motor deficits.
- Cervical spine and thoracolumbar junction (T11–L2) most susceptible to injury.
- Lower lumbar regions also susceptible to injury, although they do not usually result in neurologic injury.
- Neurologic injuries rare from sacral or coccygeal fractures.
- Acute (<3 wk): Neurogenic shock, urinary/fecal retention, vagal predominance, and hyperesthesia.
- Chronic (>6 mo) injury: Hyperreflexia and spasticity.
- Upregulation of Ach receptors from immobilization results in increased resistance to NDMBs and increased potassium release with succinylcholine.

Etiology
- Direct mechanical injury usually from traumatic insult leading to hemorrhage, edema, and ultimately spinal cord ischemia.
- Release of inflammatory mediators and membrane-destabilizing enzymes leading to disruption of electrophysiologic pathways and tissue degeneration.
- Complete lesions result in total loss of sensory and motor function below level of injury.
- Incomplete lesions result in maintenance of some function below primary level of injury and some degree of recovery.
- Above T1 = quadriplegia, below T2 = paraplegia, usually.

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.