- Mortality from SJS is around 5–10% and increases to 30% or more for TEN.
- Mortality primarily from sepsis, respiratory failure, and multiorgan dysfunction.
  - Prognosis worse with advanced age and greater BSA involvement.
  - Prognostic scoring system, called SCORTEN, can estimate pt survival.

Etiology

- Leading causes of disease are medications, followed by infections
- Medications most commonly implicated include allopurinol, anticonvulsants (lamotrigine, phenytoin, carbamazepine, phenobarbital), sulfonamide antibiotics, and oxicam NSAIDs.
- Reactions to medications occur in early treatment, typically occurring within the first 2 mo of initiation.
- Infectious etiologies: *Mycoplasma pneumoniae*, cytomegalovirus.
- Pathogenesis not completely understood; keratinocyte apoptosis attributed to cytotoxic T cells and natural killer cells through release of cytokines and cytotoxic proteins (granulysin, Fas-ligand, perforin, TNF-alpha).

Usual Treatment

- Depending on severity of disease and pt comorbidities, consider transfer to burn unit or ICU.
- If medication is suspected trigger of disease, attempt to identify and discontinue culprit drug.
- Similar to pts with major burns, treatment mainly consists of supportive care:
  - Wound care and eye care.
  - Pain management.
  - Fluid resuscitation, thermoregulation, and correction of electrolyte imbalances.
  - Nutritional support.
  - Monitoring for and treatment of superinfections.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Conjunctivitis, corneal ulceration Stomatitis, mucositis, pharyngeal erosions	Eye pain, photophobia Oral pain, odynophagia, impaired oral intake	Purulent discharge, corneal ulceration Oral/mucosal friability	Obtain baseline ophthalmologic exam
RESP	Erosions of trachea and bronchi Respiratory failure (pulm edema, pneumonia, infiltrates)	Dyspnea, cough, hemoptysis, hypoxemia	Tachypnea, pulm. consolidation, rales	CXR, CT scan ABG, bronchoscopy
CV	Hypovolemia Sepsis and septic shock	Dizziness, decreased urine output Lethargy, confusion	Tachycardia, hypotension, oliguria Fever	BP, CBC/BMP, lactate, ECG Blood culture—bacteremia (especially with *Staphylococcus aureus, Pseudomonas aeruginosa*)
GU	Urethritis Genital erosions	Difficulty voiding, dysuria Genital pain	Urinary retention Vulvar/vaginal bullae	Bladder scan, UA/culture Early gynecologic exam
HEME	Anemia Leukocytosis	Fatigue	Pallor, tachycardia Fever	CBC, differential Blood culture, CXR, urine culture, thorough dermatologic exam
METAB	Electrolyte abnormalities Insulin resistance Hypoalbuminemia			CMP Albumin

Key References: Rabito SF, Sultana S, Konefal TS, et al: Anesthetic management of toxic epidermal necrolysis: report of three adult cases, *J Clin Anesth* 13(2):133–137, 2001; Saeed H, Mantagos IS, Chodosh J: Complications of Stevens-Johnson syndrome beyond the eye and skin, *Burns* 42(1):20–27, 2016.

Perioperative Implications

Preoperative Preparation

- Correct preexisting electrolyte imbalances.
- Ensure adequate fluid resuscitation, as increased water loss from exfoliated skin can occur.
- Anticipate challenges with vascular access.
- Aim to minimize further cutaneous injury by placing soft foam or gel padding on OR table.
- Maintenance of normothermia is a challenge; transport pts to and from the OR with warm blankets and increase ambient OR temperature.

Airway

- Minimize upper airway instrumentation.
- Use extreme caution with friable oral and pharyngeal mucosal surfaces.
- Avoid nasal airway.

Preinduction/Induction

- Lubricate face and face mask prior to preoxygenation, and apply face mask with gentle pressure.
- Skin trauma can occur from tape, blood pressure cuffs, tourniquets, and adhesives (ECG leads, securing IV catheters).
- Use soft padding under blood pressure cuffs, nonadhesive pulse oximeters, and limit use of tape to secure IV or intra-arterial catheters. (Consider suturing in place or using gauze wrap.)
- Administer prophylactic antibiotics appropriate to surgical procedure; routine systemic antibiotic therapy not recommended in SJS or TEN unless there is evidence of superinfection.
- Similar to burn injured pts, SJS/TEN pts at risk for hyperkalemia if given succinylcholine.
- Meticulous ocular care and lubrication with eye drops or ointments.
- Anticipate difficulty securing and stabilizing endotracheal tube.

Maintenance

- Maintain normothermia; pts prone to heat loss from epidermal loss.
- Minimize conductive, convective, and evaporative heat loss by maintaining warm OR temp, using warming blankets, fluid warmers, and wrapping extremities with thermal insulation.
- Respiratory mucosal sloughing leading to tracheal or bronchial obstruction can occur and be life threatening; consider fiberoptic scope in the OR to aspirate bronchial casts and assess airway involvement.
- Monitor for adequate fluid resuscitation.

Extubation

- Use care with oropharyngeal suctioning to avoid further mucosal damage.
- Decision to extubate or not should be based on degree of airway involvement and intraop course.

Postoperative Period

- Increased susceptibility to infection
- Pain management

Anticipated Problems/Concerns

- Morbidity is worse in pts with tracheal or bronchial epithelial involvement.
- For pts that survive, long-term sequelae are common and primarily involve the skin, eyes, oral cavity, and teeth. Pulm complications occur, in addition to genital and urinary symptoms in female pts.

Sturge-Weber Syndrome

Puneet Khanna | Renu Sinha

Risk

- Incidence: 1 in 5000.
- Prevalence: No racial or sex prediction; sporadically occurring neurocutaneous syndrome.

Perioperative Risks

- Increased risk of seizures, neurologic deficits, bleeding due to presence of angiomas involving the oral cavity, vascular abnormality, and congenital cardiac malformations.

Worry About

- Seizures, mental retardation, neurologic deficits, headache
- Congenital glaucoma, retinal detachment
- Difficult airway
- Intracerebral angiomas

Overview

- Described by Sturge (1879) and Weber (1929).
- Also known as encephalotrigeminal angiomatosis.
- Involves a triad of (1) vascular malformation (port wine stain); (2) leptomeningeal angioma; and (3) vascular malformation of the eye.
- Facial, extrafacial, and bilateral port wine stain, along with hypertrophy of the facial soft tissue and facial bone:

- Obstructive sleep apnea.
- Difficult mask ventilation and laryngoscopy.
- Seizure:
  - The earlier the onset, the poorer the prognosis.
  - May need multiple antiepileptic drugs.
  - Treatment of dehydration/fever/infection.
- Mental retardation leads to anxiety, agitation.
- Hemiparesis, hemianopsia, hemiplegia.
- Status-like episodes.

Etiology

- Unknown. Suggested etiology includes
  - Failure of primitive cephalic venous plexus to regress during first trimester of pregnancy
  - Failure of superficial cortical veins to develop
  - Thrombosis of veins leading to vascular steal phenomena
  - Deficiency of sympathetic insertion of vessel

Usual Treatment

- Anticonvulsants.
- Control of IOP.
- Antiplatelet therapy.
- Surgery for ocular diseases, epilepsy control, angiomas.
- Require anesthesia for examination, investigations, and surgery.

Assessment Points

System	Effect	History	PE	Test
CNS	Headache (migraine like) Stroke like episodes (hemiparesis) Hemianopsia	Seizures (focal/generalized) Mental retardation		CT scan MRI X-ray: Tram-track calcification
EYE	Choroidal/episclera/conjunctival hemangioma Iris heterochromia Retinal pigment degeneration Retinal degeneration Buphthalmos Optic disc coloboma		Glaucoma Cataract	IOP monitoring Fundoscopy
CV	Septal defects, valvular stenosis and malposition of great vessels			ECG, ECHO
HEENT	Difficult mask ventilation and laryngoscopy	Hypertrophy of the soft tissue and bone	Facial and airway hemangiomas	
ANGIOMATOUS		Pituitary, thymus, lung, thyroid, testis, spleen, and lymph nodes		

Key References: Khanna P, Ray BR, Govindrajan SR, et al: Anesthetic management of pediatric patients with Sturge-Weber syndrome: our experience and a review of the literature, *J Anesth* 29(6):857–861, 2015; Thomas-Sohl KA, Vaslow DF, Maria BL: Sturge-Weber syndrome: a review, *Pediatr Neurol* 30(5):303–310, 2004.

Perioperative Implications

Preoperative Preparation

- Anticonvulsants in pts with convulsions.
- Assess airway and vascular malformation.
- Establish rapport with mentally retarded pts to decrease anxiety.
- Maintain adequate hydration.
- Benzodiazepines premedication.

Monitoring

- Intraop: Intracerebral bleed, convulsion
- ECG, respiration, NIBP, $ETCO_2$, SpO_2, BIS, EEG

Airway

- Hypertrophy of soft tissue and bone
- Facial and airway hemangioma
- Decreased intraoral space/high arched palate
- Difficult mask ventilation and laryngoscopy
- Bleeding during airway manipulation
- Difficult supraglottic placement
- Better option: Videolaryngoscopes

Anesthesia

- Based on history, examination, investigation, and type of surgery.
- Adults: Regional anesthesia:
  - Avoid systemic complications.
  - Modification of antiplatelet therapy before block.
- Children: General anesthesia

Induction

- Inhalational:
  - Use of sevoflurane is controversial for cortical epileptical activities. No persistent neurologic sequelae have been described.
  - Halothane can be used.
- For IV induction, both thiopentone and propofol can be used.

Maintenance

- O_2, N_2O or air, isoflurane or desflurane.
- Vecuronium or atracurium for muscle relaxation.
- Avoid succinylcholine (increases IOP/ICP).
- Analgesia: Fentanyl and NSAIDs.
- Avoid hypercarbia and light plane of anesthesia (increases IOP/ICP).
- Avoid hypoxia, hypoglycemia, hypotension, and hypothermia (to prevent seizure).

Extubation

- Prevent extubation response (increased risk of intracranial bleed, IOP, ICP).

Adjuvants

- Topical anesthesia, local anesthetic infiltration, and nerve blocks.

Postoperative Period

- Continue antiepileptic drugs.
- Maintain hydration.
- O_2 supplementation.

Anticipated Problems/Concerns

- Mental retardation, neurologic deficit, convulsion, facial and airway hemangiomas, and difficult airway (arrange difficult intubation cart)
- CNS hemangiomas: Increased chances of intracranial bleed, postop convulsion, and neurologic deficit (control BP)

Subclavian Steal Syndrome

Dolores B. Njoku | Natalia Hnatiuk

Risk

- Uncommon entity with a variably reported clinical significance
- Male:female ratio: 2:1

Perioperative Risks

- Stroke from a plaque originating from vertebral artery system
- Stroke from a plaque originating from subclavian artery

Worry About

- Worsening neurologic symptoms
- Upper limb ischemia

Overview

- Retrograde blood flow from vertebral artery to distal subclavian secondary to proximal ipsilateral subclavian or innominate artery stenosis or occlusion occurs when the pressure at the subclavian end of the vertebral artery drops below the basilar artery pressure.
- Presence of other extracranial arterial disease is a prerequisite to development of symptoms.
- Criteria for diagnosis (must be symptomatic):
  - Cerebral ischemia causing neurologic symptoms associated with ipsilateral arm exercise.
  - Decreased BP or arm claudication in ipsilateral arm secondary to occlusion or stenosis of subclavian artery proximal to vertebral artery.
- Ratio of left-sided to right-sided SSP is 3:1. The left subclavian artery at increased risk for atherosclerosis secondary to more acute angle of takeoff and turbulent flow.