- 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.

- Symptoms may be obscured by concomitant carotid insufficiency.
- Spontaneous resolution of vertebrobasilar symptoms may be related to the establishment of extracranial collaterals to the subclavian circulation.

Etiology

- Most common atherosclerosis.
- Other causes include Takayasu's arteritis, tumor, history of aortic stenting/grafting for aneurysm, and previous surgery, as well as trauma.
- Rare causes include congenital atresia of first portion of left subclavian, hypoplastic arch with severe coarctation, or stenosis of left subclavian at old suture site of a coarctation repair, as well as Blalock-Taussig shunts.

Usual Treatment

- Surgical:
  - Common carotid to subclavian artery bypass graft
  - Subclavian-to-subclavian artery bypass graft
  - Axillary-to-axillary artery bypass graft
- Nonsurgical: Percutaneous transluminal angioplasty and stent placement

Assessment Points

System	Assessment by Hx	PE	Test
CV	Claudication	Bruit	Difference in brachial systolic BP of at least 20 mm Hg Diminished pulse in ipsilateral arm Bruit at base of neck or supraclavicular area on affected side (proximal subclavian artery) Reactive hyperemia: Temporary cuff inflation causes peripheral vasodilation distal to cuff, when released results in increased demand leading to neurologic symptoms Color Doppler ultrasound: Ipsilateral vertebral artery flow reversal with a parvus tardus waveform in the ipsilateral subclavian artery confirms the diagnosis of SSP Vascular structures well demonstrated by contrast-enhanced MRA Flow reversal well demonstrated by flow-encoded MRI
CNS	Vertigo Rarely cortical visual disturbances, ataxia, syncope, dysarthria		Retrograde cath Angiogram Transcranial Doppler
MS	Paresis/paresthesias		See CV

Key References: Wood RJ, Walmsley AJ: Subclavian artery stenosis and blood pressure control, *Anaesthesia* 61(4):409–410, 2006; Potter BJ, Pinto DS: Subclavian steal syndrome, *Circulation* 129(22):2320–2323, 2014.

Perioperative Implications

Preoperative Preparation

- Bilateral upper extremity BP in pts undergoing surgery is characterized by large variations in hemodynamic status or in pts with previous internal mammary-coronary bypass grafts.
- Neurologic evaluation prior to surgery.

Monitoring

- Consider arterial cath, since BP maintenance may be essential for cerebral perfusion.
- Consider CVP monitoring and/or PA cath if contributing factors in pt.

Maintenance

- Consider maintaining arterial BP and heart rate near preop levels to facilitate cerebral perfusion.

Extubation

- None

Postoperative Period

- Neurologic evaluation at end of surgery.

Anticipated Problems/Concerns

- Pts with internal mammary grafts may experience a similar syndrome of coronary-subclavian steal. There is a gradient in systolic brachial blood pressure of 60 mm Hg. In such situations, myocardial ischemia that is refractory to medical management may occur.

Subphrenic Abscess

Betsy Ellen Soifer | Lee A. Fleisher

Risk

- Prior abdominal surgery, either open or laparoscopic
- Blunt or penetrating trauma
- GI perforation (malignancy, appendicitis, diverticulitis)
- Inflammatory bowel disease
- Immunocompromised pt

Perioperative Risks

- Developing sepsis

Worry About

- Respiratory compromise (pleural effusion, atelectasis, V/Q mismatching, ARDS)
  - Preop ileus/bowel obstruction; aspiration risk
  - Sepsis, including septic shock and associated renal failure and/or coagulopathy
  - Increased capillary permeability (hypovolemia)
  - High-output cardiac failure/LV dysfunction
- Lyte and acid-base disturbances

Overview

- Classic findings include fever, leukocytosis, and abdominal pain.
- Associated findings include atelectasis, pleural effusions, elevated diaphragm, ipsilateral shoulder pain, and/or hiccups secondary to diaphragmatic irritation.
- May be right- or left-sided, or both; above or below the liver or spleen.
- Fistulas may form to any abdominal or thoracic organ, including pericardium or bronchi.
- Disease severity ranges from mild to moribund.

Etiology

- Primary: Associated with perforated viscus such as duodenal ulcer, diverticulitis, appendicitis, primary liver abscess, immunocompromised state. (Pathogens include *Escherichia coli*, *Enterococcus* spp, *Bacteroides fragilis*, *Clostridium* spp and are often polymicrobial.)
- Secondary: Following surgical intervention, critical illness, or blunt abd trauma. (Pathogens include *Candida* spp, *Enterococcus* spp, *Enterobacter* spp, *Staphylococcus epidermidis*, *E. coli* and are often polymicrobial with anaerobic bacteria outnumbering or equal to aerobic bacteria in all but postbiliary surgeries.)

Usual Treatment

- Broad-spectrum antibiotics ± antifungals. Narrow coverage after cultures obtained based on culture and sensitivity.
- Percutaneous or surgical abscess drainage (80–90% successful resolution).
- Supportive therapy: Appropriate monitoring, nutrition, oxygenation, hydration, vasopressors, as indicated using the surviving sepsis recommendations.