

# Carotid Sinus Syndrome

## Risk

- Male > female
- 9% of pts with recurrent syncope; history of falls
- Increased incidence with age, rarely below age 50 y
- Peripheral vascular disease
- Head and neck cancer

## Perioperative Risks

- Presence of CSS does not increase rate of mortality, sudden death, or stroke more when compared with pts with similar age and risk factors.
- CSS does increase morbidity, secondary to injuries sustained during syncopal episodes.

## Worry About

- Presence of comorbid conditions: CAD, carotid stenosis, and neck tumor
- Severity of CSS and frequency of syncopal episodes
- Hemodynamic compromise: Bradycardia and/or hypotension

## Overview

- The carotid sinus reflex occurs with changes in transmural pressure of the baroreceptors at the carotid sinus.
- Reflex arc:
  - Afferent signals are sent via glossopharyngeal and vagus nerves to the nucleus tractus solitarius.

- Efferent signaling occurs through sympathetic and vagus nerves to the heart and blood vessels.
- CSH is defined as an exaggerated response to baroreceptor stimulation.
- CSS occurs in pts with CSH when direct CSM or accidental neck stimulation produces symptoms such as dizziness/syncope or bradycardia and/or hypotension.
- Three types of CSS:
  - Cardioinhibitory type, which is due to vagal stimulation of SA and AV nodes, resulting in sinus bradycardia and may be treated with atropine.
  - Vasodepressor type, which results in hypotension due to inhibition of vasomotor sympathetic tone; differentiated with cardioinhibitory type by not responding to atropine treatment.
  - Mixed type, which results in bradycardia and loss of vasomotor tone.
- Diagnosis: Perform CSM in supine position and massage each carotid individually for 5 second. Test is positive if any of the three are true: asystole greater than 3 sec (cardioinhibitory type); decrease in SBP >50 mm Hg (vasodepressor type); and combination or mixed type. There have been some new suggestions that SBP ≤85 mm Hg may be more sensitive in correctly identifying vasodepressor type.

## Etiology

- Afferent overshoot from external pressure due to internal atherosclerotic changes diminishing carotid sinus compliance
- Degenerative process of the nucleus tractus solitarius that occurs with age and is associated with sternocleidomastoid movement (head turning or looking down)
- Possible association with dementia, especially DLB
- Mechanical deformation from neck tumors

## Usual Treatment

- Medication
  - Atropine or vasopressors for acute, symptomatic pt.
  - For vasodepressor type, midodrine has been used with moderate success, and fludrocortisone has been used with limited success.
- Permanent dual chamber cardiac pacing is effective for cardioinhibitory and mixed types of CSS in pts who are symptomatic (pacing is of no benefit in vasodepressor type).
- Surgical denervation of carotid sinus may be attempted to treat vasodepressor type or pts who remain symptomatic despite pacing.
- Blocking the afferent limb (glossopharyngeal nerve) of the reflex with ethanol ablation is controversial due to high complication rate.
- Surgical removal of neck mass causing carotid sinus compression.

## Assessment Points

| System     | Effect   | Assessment by Hx  | PE   | Test                               |
|------------|--|---|--|------------------------------------|
| CNS        | Syncope  | Dementia, DLB   | CSM  | MRI brain                          |
| RESP/HEENT | Potential for difficult intubation/ventilation<br>Bradycardia and/or hypotension | Neck mass, neck surgery, symptoms with neck movement    | Airway exam, tracheal deviation, carotid bruit | Carotid duplex CT/MRI neck         |
| CV         | Bradycardia and/or hypotension   | Syncope with head turning or neck stimulation, CAD, PVD | CSM  | CSM with ECG monitoring and A-line |

**Key References:** Amin V, Pavri BB: Carotid sinus syndrome, *Cardiol Rev* 23(3):130–134, 2015; Solari D, Maggi R, Oddone D, et al.: Assessment of the vasodepressor reflex in carotid sinus syndrome. *Circ Arrhythm Electrophysiol* 7:505–510, 2014.

## Perioperative Implications

### Preoperative

- ECG, increased workup if advanced CAD
- CXR and/or CT scan to r/o tracheal compression if neck mass present
- Interrogate pacemaker, convert to DOO mode if unipolar cautery is to be used

### Monitoring/Lines

- Consider arterial line for symptomatic pts (strongly encouraged if position other than supine or head turning is needed during surgery).
- If no pacemaker is present, have external pacer readily available.

### Airway

- Minimize neck extension during laryngoscopy; inline immobilization may be used.
- Consider asleep fiberoptic if pt has frequent symptoms with neck movement.

### Positioning

- Avoid turning pt's neck.
- Ensure that instruments or personnel are not causing pressure to pt's neck.

### General Anesthesia

- Emergency drugs may be required based on type of CSS.
- General anesthesia may be preferred because inhalational agents have been shown to attenuate baroreceptor reflexes.
- Avoid hypotension on induction if coronary or carotid disease is present.
- Avoid long-acting beta-blockers or antihypertensive drugs.

### Regional Anesthesia

- Glossopharyngeal nerve block for CSS treatment may be performed.
- As an adjuvant to general anesthesia, local anesthetic may also be injected around the carotid sinus before

ipsilateral neck dissection to attenuate the baroreceptor response.

### Postoperative Period

- Strict postop orders in PACU outlining no head turning or neck compression.
- If intraop asystole has occurred, anesthesiologist/surgeon may need to place a temporary transvenous pacer.

## Anticipated Problems/Concerns

- Potentially difficult intubation.
- Assess pacemaker function if present.
- Pt may undergo profound hypotension or asystole at any time in the periop setting; emergency drugs should be readily available.
- Avoid neck stimulation/movement and maintain hemodynamic stability.

# Carpenter Syndrome (Acrocephalopolysyndactyly Type II)

Ray Munroe | Lee A. Fleisher

## Risk

- Extremely rare; global estimate: 1:1,000,000 births
- Over 100 reported cases

## Perioperative Risks

- Airway obstruction
- Difficult mask ventilation and/or difficult intubation

- Increased ICP from craniosynostosis
- Coexisting congenital anomalies, especially cardiac

## Overview

- Manifestation:
  - Craniosynostosis
  - Polysyndactyly
  - Cardiac defects

- Obesity
- Maxillary or mandibular hypoplasia
- Craniosynostosis usually with metopic and sagittal sutures (midline suture fusion)
- Cardiac defects in up to 50% of cases (tetralogy of Fallot, transposition of great vessels, pulm artery stenosis, VSD, and ASD)
- Other features: Brachydactyly, cognitive impairment, umbilical hernia, macrosomia, cryptorchidism in

males, molar agensis, high-arched narrow palate, broad cheeks, shallow supraorbital ridges, hypertelorism, and genu valgum

### Etiology

- Currently not elucidated

- Autosomal recessive
- Mutation in RAB 23 gene on chromosome 6P12.1-q12 shown in most cases; at least five different mutations in RAB 23 gene
- Some cases of mutation in MEGF8 gene

### Usual Treatment

- Surgery is the only option to correct cranial or cardiac abnormalities

### Assessment Points

| System | Effect   | Assessment by Hx                                 | PE   | Test             |
|--------|--|--|--|------------------|
| HEENT  | Craniosynostosis<br>Mandibular or maxillary hypoplasia |  | Acrocephaly<br>Abnormal facies<br>Overbite/underbite<br>Restricted mouth opening | CT head          |
| RESP   | Obstructive sleep apnea                                | Daytime somnolence Hx snoring                    | Macroglossia, tonsillar hypertrophy,<br>redundant soft tissue                    | Polysomnography  |
| CV     | Congenital cardiac defects                             |  | Varies with type of defect and<br>Hx or repair                                   | ECG, TTE, CBC    |
| GI     | None   |  |  |                  |
| CNS    | Increased ICP Cognitive delay                          |  | Acrocephaly  | CT head          |
| HEME   | None   |  |  |                  |
| METAB  | Obesity  | Highest percentile on pediatric<br>growth charts | Central adipose deposits   | BMP, lipid panel |

**Key References:** Bissonnette B, Luginbuehl I, Marciniak B, et al.: Acrocephalopolysyndactyly syndromes. In Bissonnette B, Luginbuehl I (eds): *Syndromes: rapid recognition and perioperative implications*. New York, NY, 2006, McGraw-Hill; Kadakia S, Helman SN, Healy NJ, et al.: Carpenter syndrome: a review for the craniofacial surgeon. *J Craniofac Surg* 25(5):1653–1657, 2014.

### Perioperative Implications

#### Preoperative Preparation

- Equipment available for difficult airway
- Congenital heart disease: may warrant prophylactic antimicrobial therapy
- Increased ICP: may warrant avoiding preop sedation

#### Monitoring

- Arterial line if indicated
- CVP/PA catheter: Consider as indicated

#### Airway

- Equipment available for difficult airway
- Surgeon available if surgical airway required

#### Preinduction/Induction

- Risk for obstruction and difficult mask ventilation is increased.
- Potential for agitation in pt with cognitive impairment.
- Maintain spontaneous ventilation while securing airway if possible.

#### Maintenance

- Adjustments required if pt Hx of repaired cardiac defects.
- Adjustments required if ICP is increased.
- Judicious opioid use in setting of increased risk for postop airway obstruction.

#### Extubation

- Difficult airway precautions

#### Postoperative Period

- Increased risk for upper-airway obstruction

### Anticipated Problems/Concerns

- Difficult airway in periop period
- Complications from increased ICP
- Complications from congenital heart defects

## Central Neurogenic Hyperventilation

Sarah C. Fausel | Kirk Lalwani

### Risk

- True CNH is exceedingly rare; the exact incidence is unknown.
- In pts with neurologic injury, it is most often associated with pulm dysfunction or shunting (aspiration, pneumonia, pulm edema, and baseline disease).
- Primarily seen in comatose pts.
- No association with age or gender.

### Overview

- A diagnosis of exclusion in neurologic disorders and in cases of hyperventilation; life-threatening causes of hyperventilation (hypoxemia, ischemic bowel, and acidosis) must be ruled out.
- Primary diagnostic criteria are hyperventilation that persists during sleep; low PaCO<sub>2</sub>, high PaO<sub>2</sub>, and absence of drug or metastatic causes.
- Associated primarily with brainstem inflammation and brainstem tumors with inconsistent involvement of midbrain, pons, and/or medulla.

- CNS lymphomas and astrocytomas are the most common tumor types with gliomas, lymphomatoid granulomatosis, and medulloblastoma; also reported in metastatic tumors.
- May result from seizure activity that stimulates the ventilatory response.
- May be associated with acute intermittent porphyria.
- Effects of GA unknown.

### Etiology

- Exact etiology and level of brainstem dysfunction not known.
- Probable etiology:
  - Uninhibited stimulation of inspiration and expiration centers in the medulla and/or loss of descending inhibitory control of ventilation by cerebral cortex with brainstem lesion.
  - Ultimate control of respiration, which may lie in the medulla (dorsal and ventral respiratory groups) with fine control from the pneumotaxic

center of the pons with input from cerebral cortex, hypothalamus, chemoreceptors and mechanoreceptors, and vagal nerve.

- Stimulation of most areas of cerebral cortex except motor/premotor areas, which inhibit respiration.
- Has been associated with brainstem infarction and malignancy.
- Tumor, which may reduce local pH in the brainstem and activating respiratory chemoreceptors located in the ventral brainstem at the junction of the pons and the medulla.
- Destructive lesions of midbrain or pons in animal studies, which do not produce CNH. (It is unclear if animal models serve as an adequate model of the human brain in this instance.)