Shy-Drager Disease

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Risk

- More common in men than in women.
- Symptoms begin in fifth-seventh decades of life.

Perioperative Risks

- Autonomic dysfunction with CV collapse due to decreased sympathetic outflow and abnormal parasympathetic homeostatic mechanisms
- Aspiration risk

Worry About

- Orthostatic hypotension and intraop fluctuations in BP, particularly during induction.
- Response to sympathomimetic drugs is unpredictable and may be exaggerated owing to denervation hypersensitivity.
- Little or no HR or BP response to indirect sympathomimetic agents (i.e., ephedrine, methamphetamine) or anticholinergic medications (i.e., atropine).
- Hyperresponsiveness of BP to hyperventilation/hypoventilation (hypercapnia/hypocapnia).
- Loss of baroreceptive response leads to hyperresponsiveness to volume status and sudden changes in blood volume.
- Cannot use sweating, tachycardia, or BP as indicators of anesthesia depth.
- Positive-pressure ventilation can decrease venous return and cause dramatic hypotension without associated change in HR.
- Up to 50% of pts will have supine hypertension.
- Liver blood flow can be dependent on posture, so hepatically cleared drugs' plasma levels can be highly dependent on posture.
- Lyte abnormalities: Hypokalemia and hypomagnesemia when treated with fludrocortisone.
- Central sleep apnea: Apneic syndromes due to impaired central regulation of respiration.
- Obstructive sleep apnea: Found in advanced stages.
- Vocal cord paralysis due to laryngeal muscle dysfunction: Found in advanced stages.
- Impaired GI motility increased the risk of aspiration as well as postop ileus.
- Faulty thermoregulatory systems: Hyperthermia-induced hypotension, lack of peripheral vasoconstriction to cold environment, can lead to hypothermia and hypotension.

Overview

- Irreversible, rapidly progressive, and fatal disease causing death usually within 10 y of onset due to postsyncopal cerebral ischemia.
- Primary neurodegenerative disease with primary autonomic failure. Parkinsonism-plus syndrome; however, Shy-Drager involves loss of vascular reflexes. There is secondary autonomic failure in Parkinson disease.
- Clinical manifestations: Orthostatic hypotension, supine hypertension, parkinsonian symptoms, urinary and bowel dysfunction, impaired potency and libido, decreased sweating.
- Autopsies showed diffuse involvement of the CNS and peripheral autonomic nervous system as well as corticobulbar and corticospinal tracts, basal ganglia, and cerebellum.
- Difficult to treat the parkinsonian symptoms, as dopaminergic drugs may exacerbate orthostatic hypotension.

Etiology

- Unknown

Usual Treatment

- Goals of treatment: Reduce venous pooling, increase peripheral vascular resistance, increase plasma volume.
- Nonpharmacologic: Symptomatic relief of orthostatic hypotension with postural change, liberal salt intake, graded elastic stockings, postural training with head of bed elevated during sleep.
- Pharmacologic: Fludrocortisone (mineralocorticoid, started preop); midodrine (peripheral α-adrenergic agonist); pyridostigmine (cholinesterase inhibitor, may increase BP); octreotide (splanchnic vasoconstriction elevates SVR); atomoxetine (inhibits reuptake of norepinephrine); sympathomimetics (ephedrine/methamphetamine effective only in early stages); prostaglandin inhibitors (indomethacin, ibuprofen); MAO inhibitors; tyramine.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Vocal cord paralysis	Obstruction; apnea episodes; stridor, particularly during sleep	Midline position of the cords after induction	Direct laryngoscopy, fiberoptic nasopharyngoscopy
	Obstructive sleep apnea	STOP-BANG OSA risk questionnaire	Neck circumference	Polysomnography
CV	Orthostatic hypotension	Syncope; dizziness, visual changes related to position	BP changes related to position	Tilt table test, Valsalva maneuver testing, cold pressor test, sympathetic and parasympathetic "stress" testing
	Fixed HR			Palpation, ECG
RESP	Irregular breathing			Auscultation, visualization
GI	Gastroparesis Fecal incontinence, diarrhea, constipation, sodium loss	Early satiety, dysphagia, GERD	Loss of rectal sphincter tone	Gastric emptying study Lytes/BMP
GU	Urinary incontinence Atonic bladder Sexual dysfunction	Nocturia; stress/overflow incontinence Sexual impotence		
CNS	Parkinsonian symptoms Anhidrosis Heat intolerance		Cogwheel rigidity Shuffling gait Anisocoria Horner syndrome	Sudomotor testing for anhidrosis
MS	Osteoporosis and aseptic necrosis (may be assoc with autonomic dysfunction)		Muscle atrophy Fasciculations	

Key References: Bevan DR: Shy-Drager syndrome. A review and a description of the anaesthetic management, *Anaesthesia* 34(9):866–873, 1979; Mustafa HI, Fessel JP, Barwise J, et al.: Dysautonomia: perioperative implications, *Anesthesiology* 116(1):205–215, 2012.

Perioperative Implications

Preoperative Preparation

- Reduce venous pooling, increase PVR, and increase plasma volume.
- Oral fludrocortisone can be started up to 2 wk preop.

Monitoring

- Arterial and central venous cath to guide fluid replacement and use of vasopressors
- Temperature: Avoid hypothermia/hyperthermia.

Airway

- Vocal cord paralysis and dysautonomia with gastroparesis may make awake intubation or rapid sequence intubation with cricoid pressure more desirable.

Preinduction/Induction

- Avoid drugs that would cause decrease in cardiac output, HR, or SVR as profound hypotension may occur due to decreased sympathetic outflow.
- Regional anesthesia can be used if indicated for procedure.
- Consider steroid supplementation if pt is on fludrocortisone preop.

Maintenance

- Positive-pressure ventilation and pneumoperitoneum during laparoscopic surgery may decrease venous return and exaggerate hypotension.
- Use direct-acting sympathomimetic drugs in small doses titrated to effect. Pt may have an exaggerated response due to denervation hypersensitivity. Vasopressin has been used successfully in cases in which hypotension was not responsive to other sympathomimetics.
- Atropine may not increase the HR owing to parasympathetic deficiency.
- Rapid blood loss or rapid fluid administration can precipitate exaggerated changes in BP.
- Maintain normothermia.

Extubation

- Ensure pt can protect airway.

Postoperative Period

- Continue invasive BP monitoring if necessary.
- Monitor volume status, respiratory mechanics, and temp closely.
- Abnormal lytes can affect BP and response to medications.
- Restart home dysautonomia medications as soon as possible postop.

Anticipated Problems/Concerns

- Autonomic dysfunction with CV collapse
- Aspiration risk

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Sick Sinus Syndrome

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Risk

- Highest incidence occurs in pts >60 y.
- Common in pts who have had congenital heart defect repair surgery.
- Sinus node dysfunction is a common indication for pacemaker implantation.

Perioperative Risks

- Syncope, symptomatic bradycardia, and asystole.

Worry About

- Sinus bradycardia can be poorly responsive to atropine and require a pacer intraop.
- Termination of a tachy-brady event where an atrial tachycardia occurs can lead to a prolonged bradycardia or asystole.
- A tachy-brady event can lead to demand myocardial ischemia and can precipitate heart failure in pts with related comorbidities.

Overview

- A significant portion of elderly pts with SSS frequently have other cardiac comorbidities, such as CAD.
- SSS includes several ECG abnormalities: Noniatrogenic persistent spontaneous sinus bradycardia that is inappropriate for physiologic circumstance, sinus arrest or exit block, or alterations of paroxysmal atrial tachyarrhythmia (often fibrillation or flutter) and period of bradycardia—tachy-brady syndrome.
- Class I indications for pacemaker insertion in SSS include SND with documented symptomatic bradycardia, or sinus pauses; SND as a result of essential long-term therapy; or SND with symptoms of chronotropic incompetence.
- A large portion of pts with chronotropic incompetence (defined as the inability to achieve 80% of age predicted HR during physiologic stimulus) also have SND.

Etiology

- Causes are numerous and not clearly defined.
- In the elderly, likely due to fibrosis of SA node and hypersensitivity to autonomic changes.
- In adults with congenital heart disease (especially with ASD repair, or extensive atrial reconstruction), likely due to surgical (direct or inflammatory) trauma to the SA node.

Usual Treatment

- When SSS remains undiagnosed until an episode of bradycardia in the OR, administer atropine (0.5 mg–3 mg IVP) and epinephrine (10 µg IVP with rapid up-titration based on HR), along with pacing (external or transvenous). Pts with SSS often have a poor response to atropine.
- When SSS is diagnosed preop (i.e., ECG changes with symptoms), suggest a pacemaker (often DDD or AAI). A pacemaker does not control tachyarrhythmia, but instead it allows antiarrhythmic therapy for tachycardia by pacing during bradycardia caused by the therapy.
- Anticoagulation with warfarin is used if continuous ECG monitor detects paroxysms of atrial tachyarrhythmia, since this subset of pts has an increased thromboembolic risk.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Low cardiac output Tachy-brady event	Syncope, presyncope, lightheadedness, decreased exercise capacity, dyspnea on exertion, fatigue, confusion, memory loss, CVA (especially in elderly) Palpitation, angina, CHF symptoms	Bradycardia, tachycardia	ECG Continuous ECG monitor with symptom diary
RENAL	Accentuate SSS			Potassium (hypokalemia)

Key References: Semelka M, Gera J, Usman S: Sick sinus syndrome: a review, *Am Fam Physician* 87(10):691–696, 2013; Staikou C, Chondrogiannis K, Mani A: Perioperative management of hereditary arrhythmogenic syndromes, *Br J Anaesth* 108(5):730–744, 2012.

Perioperative Implications

Preinduction/Induction

- Factors that alter autonomic balance to favor a parasympathetic state can produce sinus bradycardia: Eye surgery, increased ICP, severe hypoxia, and cervical/mediastinal tumors.
- Optimize extrinsic factors that can decrease heart rate: hypoxia, hypothermia, ICH, and hypothyroidism.
- Volatile anesthetics, propofol, and vecuronium all decrease sinus node activity in a dose-dependent fashion. Consider using NMBA with vagolytic properties (e.g., rocuronium).
- Standard monitors (pulse oximetry, ECG).
- In pt population that is dependent on atrial systole for sufficient cardiac output (such as those with ischemic cardiomyopathy, aortic stenosis, or diastolic dysfunction), an atrial tachyarrhythmia (as part of the tachy-brady phenomenon) can lead to hypotension.
- Consider placing transcutaneous pacing pads on pt prior to induction, or placing a central venous introducer in preparation for a transvenous pacing wire if needed.
- In cases of asystole or bradycardia, attempt therapeutic medications: Atropine 0.5–1 mg IV q 3–5 min up to 0.04 mg/kg (max 3 mg), or ephedrine 5–5 mg IV bolus, or infusions of dopamine 5–20 µg/kg per min IV, or epinephrine 2–10 µg/min IV, or isoproterenol 2–10 µg/min. Electrical pacing is most likely to succeed.
- If an episode of tachycardia occurs in pt with pacemaker, use normal agents for control of arrhythmia, as the pacemaker should rescue bradycardia. If no pacemaker, use short acting agents for rate control such as esmolol.

Regional Anesthesia

- Case reports demonstrate that SSS may manifest from several types of blocks, including a regional block that produces sympathectomy, stellate ganglion blockade, thoracic epidural, and spinal anesthesia.
- Standard monitoring is recommended.
- If history is strongly suggestive of SSS, consider having pacing pads in the room.

Postoperative Period

- If SSS manifests in a pt without pacemaker intraop, consult cardiology to further evaluate for permanent pacemaker placement.

Anticipated Problems/Concerns

- An episode of atrial tachyarrhythmia that terminates in asystole or significant bradycardia.
- Atrial tachyarrhythmia in a susceptible population (aortic stenosis, diastolic dysfunction) may itself compromise cardiac output.
- Symptomatic sinus bradycardia may not respond robustly (or at all) to atropine. It may need a pacing if a second agent (isoproterenol, epinephrine, ephedrine, or dopamine) does not work.