



NOTES

AUTONOMIC DISEASES

GENERALLY, WHAT ARE THEY?

PATHOLOGY & CAUSES

- Autonomic nervous system (ANS) disorders (dysautonomia)
- Normative autonomic function
 - Balanced impulses of sympathetic, parasympathetic ANS
 - One/both components fail → symptoms
- Etiology
 - Genetic, environmental factors

CAUSES

- Primary
 - Pure autonomic failure, familial dysautonomia, multiple system atrophy, postural orthostatic tachycardia syndrome (POTS)
- Secondary (neuropathy)
 - Alcoholism, diabetes mellitus, trauma, HIV infection, multiple sclerosis, Lyme disease, Parkinson's disease, porphyria, nerve compression (tumor), drug toxicity (vincristine)

SIGNS & SYMPTOMS

- Breadth of autonomic function → wide symptomatic variation
- Common autonomic disease symptoms
 - ↑↓ heart/respiration rate
 - ↑↓ blood pressure
 - Bowel/bladder/erectile dysfunction
 - Hypohidrosis/hyperhidrosis
 - Syncope

DIAGNOSIS

DIAGNOSTIC IMAGING

- See individual diseases

LAB RESULTS

- Nerve biopsy
 - Neuropathy detection

OTHER DIAGNOSTICS

- Autonomic function test battery
 - Monitor heart rate, autonomic functions for pathological changes
- Valsalva maneuver
 - ↑ intraspinal pressure → neuropathic symptom exacerbation
- Quantitative sudomotor axon reflex test (QSART) test
 - Electrical current → sweat gland stimulation
- Tilt table test
 - Individual lies on table → table tilted upright → detects sudden blood pressure change

TREATMENT

- Treat underlying cause if possible
- Mostly symptomatic treatment

HORNER'S SYNDROME

osms.it/horners-syndrome

PATHOLOGY & CAUSES

- AKA oculosympathetic paresis
- Clinical syndrome
 - Damaged sympathetic neural pathways to eye, associated structures
- Sympathetic innervation to eye
 - Three neurons comprise pathway
 - 1st order neurons: in posterolateral hypothalamus, preganglionic fibers
 - 2nd order neurons: in ciliospinal center (Budge's center) in intermediolateral segment of spinal column (C8–T2) → preganglionic fibers travel to superior cervical ganglion (SCG) → synapse with 3rd order neurons
 - 3rd order neurons: in SCG, postganglionic fibers follow different paths upon leaving SCG → flushing, absent sweating not mandatory signs
- Manifests ipsilaterally

CAUSES

- Condition manifests following pathway interruption
- Congenital/acquired
 - **Congenital:** may present with heterochromia iridis as eye pigmentation under sympathetic innervation during development
- Classification based on lesion's level
 - 1st order neuron lesion: Arnold–Chiari malformation, cerebrovascular insult, basal skull tumor
 - 2nd order neuron lesion: trauma, cervical rib, Pancoast tumor, neuroblastoma, aorta dissection
 - 3rd order neuron lesion: herpes zoster, internal carotid artery dissection, cluster headache

SIGNS & SYMPTOMS

- Classic triad: ptosis, anhidrosis, miosis
- May present with anhidrosis (if 2nd order neurons affected), flushing (impaired vasoconstriction), apparent enophthalmos (ptosis)



MNEMONIC: PAM

Signs & symptoms of Horner's syndrome

Ptosis

Anhidrosis

Miosis



Figure 62.1 An individual with Horner's syndrome demonstrating ptosis and miosis of the left eye.

DIAGNOSIS

DIAGNOSTIC IMAGING

X-ray

- Detects Pancoast tumor, shoulder trauma

MRI

- Detects aneurysm, dissection

LAB RESULTS

- Vanillylmandelic acid (VMA) level
 - Detects neuroblastoma

OTHER DIAGNOSTICS

- Neurological exam
- Pharmacological diagnostics
 - Disorder detection, lesion level determination
 - **Cocaine drops**: norepinephrine missing from synaptic cleft → absent mydriasis

- **Apraclonidine**: upregulation of α_1 receptors (\uparrow apraclonidine sensitivity) → mydriasis occurs
- **Hydroxyamphetamine**: 1st or 2nd order neuron lesion → mydriasis occurs (postganglionic fibers undamaged); 3rd order neuron lesion → weaker/absent mydriasis in affected eye

TREATMENT

- Treat the underlying cause if possible

ORTHOSTATIC HYPOTENSION (OH)

osms.it/orthostatic-hypotension

PATHOLOGY & CAUSES

- Sudden, sustained systolic blood pressure ($> 20\text{mmHg}$)/diastolic blood pressure ($> 10\text{mmHg}$) drop within three minutes upon standing/tilting head upright $\geq 60^\circ$
- Delayed/lowered lower-body vasoconstriction
- Lower-body blood accumulation while seated/supine → lower-body vasoconstriction delayed upon standing → \downarrow cardiac output → \downarrow cerebral perfusion → dizziness, blurred vision, syncope

CAUSES

- Neuropathy impairs vasoconstriction
- Baroreceptor reflex impairment (α_1 blockers inhibit vasoconstriction)
- Hypovolemia (absolute/relative); atherosclerosis; diabetes mellitus; Addison's disease; Parkinson's disease; anorexia nervosa; alcohol, THC intoxication; medication (MAOI)
- Occurs in elderly/postpartum individuals

COMPLICATIONS

- Postural orthostatic tachycardia syndrome (compensatory mechanism for chronic \downarrow cardiac output), syncope, injury (falling)

SIGNS & SYMPTOMS

- Pale skin, vertigo, blurred vision, nausea, heart palpitations

DIAGNOSIS**LAB RESULTS**

- Measure blood pressure
 - Confirm sudden drop

OTHER DIAGNOSTICS

- Tilt table test
 - Provokes OH episode

TREATMENT**MEDICATION**

- Corticosteroids
- Antihypertensives
- Supplemental measures (caffeine)

OTHER INTERVENTIONS

- Increase blood pressure via increased fluid/salt intake
- Treating underlying cause