

EVALUATION OF AUTONOMIC FUNCTION

HISTORY AND TESTING

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Case# 1

Loss of consciousness and vehicular homicide. Guilty or innocent?

A 52-year-old man collides head on with another car while driving a van on the wrong side of the highway. Five people die in the collision. He suffers only broken ribs and other minor injuries. Despite freezing cold weather on the afternoon of the accident, the car window on the driver's side was open. Right after the accident, neurological examination was normal, he had no ETOH in blood and toxic screen was negative.

The diagnosis is:

- a) Seizure
- b) Acute psychosis /Suicidal attempt
- c) Narcolepsy
- d) Reckless driving while using a cell phone
- e) Cough syncope

Case# 2

A lightheaded university professor

A 54-year-old university Professor complains of lightheadedness and shortness of breath after climbing stairs. He first noted lightheadedness after prolonged standing in his mid forties. Over the past 5 years he has had 2 brief syncopal episodes, both in the morning after breakfast on warm days. He describes erectile dysfunction and has retrograde ejaculation. He complains of constipation and nocturia.

His general physical and neurological examinations were normal. Blood pressure while supine was 140/80 mmHg with a heart rate of 63 beats per minute and after standing for 3 minutes his blood pressure fell to 86/58 mmHg while heart rate increased to 67 beats per minute. Serum Na concentration was 134mEq/l, K was 3.8 mEq/l and morning serum cortisol was 20 micrograms per deciliter (ug/dl).

Plasma norepinephrine concentration while supine was 32 pg/ml; dopamine was 23 pg/ml and epinephrine was 10 pg/ml. Serum cortisol concentration was 20 micrograms per deciliter (ug/dl) in the morning.

The diagnosis is:

- a) Autoimmune autonomic neuropathy
- b) Chronic fatigue syndrome
- c) Pure autonomic failure
- d) Dopamine-beta hydroxylase deficiency
- e) Adrenal insufficiency.

Case# 3

Daily loss of consciousness in a 14 year-old girl.

An otherwise normal 14-year old girl is referred for evaluation of recurrent episodes of sudden loss of consciousness with falls occurring up to six times a day for the last three months. Episodes occur while she is standing or sitting, frequently are preceded by raising her arms or turning her head. She is unaware of the

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episodes, which last only a few seconds and leave no neurological sequelae. She describes trouble concentrating and reading and is not doing well in school. There was a questionable diagnosis of sleep paralysis.

The diagnosis is:

- a) Benign autonomic seizures (Panayiotopoulos syndrome)
- b) Long QT syndrome
- c) Carotid body tumor
- d) Ehlers-Danlos Syndrome
- e) Psychiatric disorder

Case # 4

A bus driver in his late 40s with orthostatic and heat intolerance.

A 49 year-old bus driver complains of lightheadedness on standing, heat intolerance, urinary frequency and urgency. Symptoms occur each time he stands up and are worse in the morning. He describes a pressure on his shoulders only when standing. He has nocturia and erectile dysfunction.

Video of neurological exam.

The diagnosis is:

- a) Recurrent vasovagal syncope
- b) Diabetes
- c) Parkinson disease
- d) Multiple system atrophy
- e) Amyloidosis

Case #5

A retired neurosurgeon with orthostatic hypotension and a very dry mouth

81 year old retired neurosurgeon consults because of 9-month history of lightheadedness on standing followed on several occasions by brief episodes of loss of consciousness. He also complains of a very dry mouth, weight loss, diminished appetite and early satiety. He describes constipation, nocturia and urinary frequency as well as erectile dysfunction. His past medical history includes Hashimoto's thyroiditis and vitiligo. He had a cardiac pacemaker inserted because of sinus bradycardia.

The diagnosis is:

- a) Sjogren's disorder
- b) Paraneoplastic syndrome with autonomic failure
- c) Auto-immune autonomic ganglionopathy
- d) Pure autonomic failure
- e) Pacemaker syndrome

Case #6

A young man with convulsions in a parking lot.

A lawyer in his early 30's was diagnosed with epilepsy 4 months prior after 3 apparent grand mal seizures and is now is taking 2 anticonvulsants. One of the 3 apparent seizures was witnessed. He was sitting in his car waiting for his girlfriend in a parking lot. He lost consciousness and abnormal bilateral jerky movements were witnessed.

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He had a normal neurological examination. Blood pressure was 120/80 mmHg while supine with a heart rate of 62 beats per minute; standing 120/82 mmHg with a heart rate of 69 beats min.

Video of tilt table testing

The diagnosis is:

- a) Epilepsy
- b) Vasovagal syncope
- c) Munchausen syndrome
- d) Subclavian steal syndrome
- e) Adrenoleukodystrophy.