

# HEADACHES, MOTOR DISORDERS, AND AMYOTROPHIES

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American College of Osteopathic internists  
Internal Medicine Board Review Course 2018  
Marriott Chicago Downtown  
Chicago, Illinois  
April 25-29, 2018

Scott Spradlin, D.O. FACP, FACOI

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# Board Exam Sample

28-year-old female has a throbbing, one-sided headache three times a month. It occurs suddenly, persists for 2 days.

This is what type of headache?

- A. Migraine with aura
- B. Migraine without aura
- C. Tension-type headache
- D. Cluster headache

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- 
- Answer: Cluster Headache(D)

# Headaches, Motor Disorders, Amyotrophies

## Migraines

- Unilateral, intermittent, throbbing
- Lasts 4 hours-3days
- Light sensitive/sound sensitive
- Associated with prodrome
- Aura- scintillating scotomas
- Triggers
- Acephalic- abnormal transient dysfunction No pain

# Headaches, Motor Disorders, Amyotrophies

- **General Classification of Headaches**
  - Migraine
  - Tension
  - Cluster
  - Coital
  - Post-Traumatic
  - Temporal Arteritis
  - Pseudotumor Cerebri
  - Thalamic

## Headaches, Motor Disorders, Amyotrophies

- **Treatment**

- Acute

- Serotonin agonists (Triptans)
- NSAIDS
- Ergotamine
- Dopamine antagonists
- Narcotics- rarely recommended
- DHE IV- *severe*

- *Chronic- Botulinum Toxin A*

Headaches, Motor Disorders, Amyotrophies

## **Prophylactic**

- Beta blockers

- Tricyclic Antidepressants

- Divalproex

- Topiramate

# Headaches, Motor Disorders, Amyotrophies

## Cluster Headaches

Occur daily for weeks then stop

Ice pick like

Associated with REM or early AM

“Worst Pain” known

Pain peaks in 5-10 min then throbs 2 hours

Ipsilateral Horner’s syndrome

Male

Drinkers and Smokers

Tall and THIN and Hazel eye color



Headaches, Motor Disorders, Amyotrophies

## Treatment

- 5-HT<sub>1</sub> Receptor agonists
- Triptans/Ergot Alkaloids
- Oxygen 8-10 L/min
- Lidocaine intranasal drops
- Corticosteroids
- Prophylactic= Calcium Channel Blockers

## Headaches, Motor Disorders, Amyotrophies

- **Tension**
  - Chronic muscle contraction
  - Can have vascular component
  - Daily
  - Bilateral
  - Tight band feeling
  - **Non throbbing**

Headaches, Motor Disorders, Amyotrophies

## **Treatment**

- NSAIDS

- Muscle Relaxants

- Tricyclics

- Beta Blockers

# Headaches, Motor Disorders, Amyotrophies

## • Other Headaches:

### Coital

- Benign TX: Propanolol / Indomethacin

### Post-Traumatic

- Vascular TX: same as migraine

### Temporal Arteritis

- >55 yr old
- Sudden onset
- Temporal artery tenderness
- Elevated ESR- *usually* >60 Tx: Biopsy/Steroids

### Pseudotumor Cerebri

- Obese premenopausal women
- Diplopia/headache visual field loss papilledema
- CSF=>250 mm H<sub>2</sub>O Tx: Diuretic/Steroids

### Thalamic

- Severe/debilitating after infarct usually has hemianesthesia

# Headaches, Motor Disorders, Amyotrophies

- **Motor Disorders:**

- Parkinsons Disease
- Progressive Supranuclear Palsy
- Huntingtons Chorea
- Essential Tremors
- Tardive Dyskinesia
- Neuroleptic Malignant Syndrome
- Tic Douloureux
- Giles de la Tourette
- Torticollis
- Meige Syndrome
- Creutzfeldt-Jakob disease

Headaches, Motor Disorders, Amyotrophies

# Parkinsons Disease

Clinical Diagnosis solely

Decrease dopamine producing cells in the *substantia nigra*

Signs/Symptoms:

- Resting Tremor
- Rigidity
- Retarded movement
- Loss of postural reflexes

**Table 1. Common Presentations of Parkinson's Disease.**

Common Presentations of Parkinson's Disease

Presentation	Parkinsonism	Differential Diagnosis	Distinguishing Signs
Tremor	Asymmetric rest tremor	Essential and other tremors	Symmetric postural and action tremor
Clumsy or weak limb	Bradykinesia	Carpal tunnel syndrome, radiculopathies, and stroke	Altered reflexes, sensation, and strength
Stiff or uncomfortable limb	Rigidity	Musculoskeletal syndromes	Pain and limitation of movement
Gait disorder	Asymmetric slowness, shuffling, reduced arm swing, minimal or no imbalance	Multiple ischemic lesions in the brain, hydrocephalus, and musculoskeletal disorders	Symmetric shuffling, retained arm swing, wide-based gait, prominent imbalance, limited movement at knee and hip

# Headaches, Motor Disorders, Amyotrophies

- **Treatment**

- **Increase the Dopamine**

- Decrease the Acetylcholine

- Dopaminergic is most successful**

- levodopa/carbidopa (Sinemet® or Atamet®)

- Anticholinergics-Artane

- Parlodel/Eldepryl/Mirapex/

- Ropinirole (Requip, Requip XL)

- Rasagiline (Azilect)

- Apomorphine (Apokyn)

- Amantadine

- Toicapone-COMT

- Entacapone-COMT

Deep Brain Stimulation

Surgery-Pallidotomy



**Table 2. Initial Therapy for Symptoms in Parkinson's Disease.\***

Drug Class	Example(s)	Initial Dosage	Usual Dosage	Side Effects
<b>First-line dopaminergic agents</b>				
Carbidopa plus levodopa				
Immediate release (Sinemet)	25 mg carbidopa, 100 mg levodopa	1/2 tablet three times daily	1 to 2 tablets three times daily	At initiation: anorexia, nausea, vomiting, dizziness, hypotension (a 1:4 ratio of carbidopa:levodopa reduces gastrointestinal symptoms), long-term therapy: motor fluctuations, dyskinesias, confusion, hallucinations
Controlled release (Sinemet-CR)	25 mg carbidopa, 100 mg levodopa	1 tablet three times daily	—	Same as for immediate-release preparations
	50 mg carbidopa, 200 mg levodopa	1/2 tablet three times daily	1 tablet three times daily	
Carbidopa plus levodopa plus entacapone (Stalevo)	12.5 mg carbidopa, 50 mg levodopa, 200 mg entacapone	1 tablet three times daily	—	Same as with preparations above, plus diarrhea
	25 mg carbidopa, 100 mg levodopa, 200 mg entacapone	—	—	
	37.5 mg carbidopa, 150 mg levodopa, 200 mg entacapone	—	—	
<b>Dopamine agonists</b>				
Nonergot	Pramipexole (Mirapex)	0.125 mg three times daily	0.5–1.5 mg three times daily	Nausea, vomiting, hypotension, ankle edema, excessive daytime sleepiness, compulsive behavior, confusion, and hallucinations
	Ropinirole (ReQuip)	0.25 mg three times daily	3–8 mg three times daily	Same as for pramipexole
Ergot	Pergolide (Permax)	0.05 mg three times daily	1 mg three times daily	Same as for nonergot drugs plus retroperitoneal, pulmonary, and cardiac fibrosis
<b>Second-line alternatives</b>				
Anticholinergic agents	Trihexyphenidyl (Artane)	1 mg three times daily	2 mg three times daily	Impaired memory, confusion, constipation, blurred vision, urinary retention, xerostomia, and angle-closure glaucoma
	Benzotropine (Cogentin)	0.5 mg twice daily	1 mg twice daily	Same as for trihexyphenidyl
Selective MAO-B inhibitors	Selegiline (Eldepryl)	5 mg daily	5 mg twice daily	Insomnia, nausea, anorexia, hallucinations, potential for interactions with SSRIs and meperidine
NMDA antagonist	Amantadine (Symmetrel)	100 mg twice daily	100 mg twice daily	Dizziness, insomnia, nervousness, livedo reticularis, hallucinations, confusion

\* All antiparkinsonian drugs are started at low doses and increased slowly to reduce adverse effects. Likewise, slow withdrawal of these drugs after long-term treatment is prudent to avoid a marked worsening of parkinsonism or even the neuroleptic malignant syndrome (discussed by Keyser and Rodnitzky<sup>20</sup>). MAO-B denotes monoamine oxidase B, SSRI selective serotonin-reuptake inhibitor, and NMDA *N*-methyl-D-aspartate.

Headaches, Motor Disorders, Amyotrophies

## Progressive Supranuclear Palsy

Similar to Parkinsons

Erect Posture

Hyperextension Neck

No tremor

Vertical Ophthalmoplegia- *can't look up or down*

Over 2 yrs unable to walk

No treatment

# Headaches, Motor Disorders, Amyotrophies

## Huntingtons Chorea

Inherited

Autosomal Dominant

Hemiballismus

Facial twitching

Rigidity/Dystonia

Lab:

**H-D Gene**

**Decreased GABA**

**CT/MRI= Bulge of Caudate Nucleus/ enlarged ventricles**

Treatment:

**Tetrabenazine**

**Amantadine or Riluzole**

Headaches, Motor Disorders, Amyotrophies

## **Benign Tremor (Essential)**

Not to be confused with Normal tremor

7 Hz

Autosomal Dominant

Treatment

Beta Blockers

Primidone

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# Tardive Dyskinesia

Effect of Long term antipsychotics

Involves Lips, tongue, face, and neck

Can affect limbs

Treatment

Exchanging the dopamine antagonist  
antipsychotic

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# Neuroleptic Malignant Syndrome

Response to antipsychotics

Dopamine Receptor Blockade

Fever- can be as high as 106

Rigidity

Increased CPK

Altered mental status

Treatment:

- Remove drugs

- Supportive therapy

- Dantrolene/Bromocriptine/Amantadine

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## **Tic Douloureux**

Hemifacial spasm

Pain

Trigeminal neuralgia

80% have basilar artery affecting the facial n.

Treatment: Carbamazepine/Surgery

# Headaches, Motor Disorders, Amyotrophies

## Other:

- Gilles de la Tourette- Neuroleptics-Risperdal/Geodon

- Torticollis-Botulinum toxin

- Meige Syndrome:

  - Bilateral blepharospasm with lip/mouth involvement

- Creutzfeldt-Jakob disease

  - Myoclonus with dementia/brain biopsy/no tx

  - Sudden onset



Headaches, Motor Disorders, Amyotrophies

# Seizures

Excessive abnormal discharges of electrical activity in CNS

Epilepsy is a syndrome of recurrent episodes of seizure activity

Two Types:

Partial-

Generalized-

# Headaches, Motor Disorders, Amyotrophies

## **Partial Seizures**

Also known as “Focal or Local Seizures”

Seizure activity occurs in a specific area

Sensory Phenomena

Autonomic manifestations

Psychic manifestations

# Headaches, Motor Disorders, Amyotrophies

## Generalized Seizures

### **Absence:**

Sudden

Brief motor activity

Blank Stare

Unconsciousness

### **Myoclonic:**

Sudden

Uncontrollable

Jerking of single or multiple muscle groups

Unconsciousness

Confusion postictally

# Headaches, Motor Disorders, Amyotrophies

## Tonic Clonic Seizure- *Grand Mal*

May or May not have an Aura

Sudden loss of consciousness

Tonic Phase-

abrupt increase in muscle tone and contraction

Clonic Phase-

Rhythmic muscular contraction and relaxation

# Headaches, Motor Disorders, Amyotrophies

## Status Epilepticus

Continuous seizures

## Complex Partial Seizure

Purposeless repetitive activities

Evolves to secondary generalized

**TABLE 1. PRINCIPAL TYPES OF SEIZURES.**

<b>TYPE OF SEIZURE</b>	<b>CLINICAL FEATURES</b>	<b>ELECTROENCEPHALOGRAPHIC FEATURES*</b>
Partial		
Simple partial seizures (focal)	Signs and symptoms may be motor, sensory, autonomic, or psychic, depending on the location of the electrical discharge; consciousness is not impaired	Focal slowing or sharp-wave activity, or both
Complex partial seizures (temporal lobe or psychomotor)	Seizure may begin with no warning or with motor, sensory, autonomic, or psychic signs or symptoms; consciousness is impaired; automatisms (automatic acts of which the patient has no recollection) may occur; seizure is often followed by a period of confusion	Focal slowing or sharp-wave activity, or both
Secondarily generalized partial seizures (tonic-clonic, or grand mal)	Seizures may begin with motor, sensory, autonomic, or psychic signs or symptoms; consciousness is lost, with tonic increase in muscle tone; subsequent rhythmic (clonic) jerks subside slowly; patient is comatose after seizure and recovers slowly; tongue biting or incontinence, or both, may occur	Focal slowing or sharp-wave activity, or both
Generalized		
Absence seizures (petit mal)	Seizure begins rapidly, with a brief period of unresponsiveness (average, 10 seconds) and rapid recovery; there may be increased or decreased muscle tone, automatisms, or mild clonic movements. Seizure can be precipitated by hyperventilation; age at first seizure, 3–20 yr	Spike-wave pattern (3 Hz)
Primarily generalized tonic-clonic seizures (grand mal)	Loss of consciousness occurs without warning or is preceded by myoclonic jerks; clinical features are similar to those of a secondarily generalized partial seizure	Spike-wave pattern (3–5 Hz)

\*The electroencephalographic features listed are those observed on routine electroencephalography during which a seizure does not occur.

Headaches, Motor Disorders, Amyotrophies

## **MYOPATHIES**

- Hereditary/Congenital

- Metabolic

- Inflammatory

- Toxic

# Headaches, Motor Disorders, Amyotrophies

## Work up for Myopathy

- CK with isoenzymes
- Electrolytes, calcium, magnesium
- Serum myoglobin
- Serum creatinine and BUN
- Urinalysis:
  - Myoglobinuria is indicated by positive urinalysis** with few RBCs on microscopic evaluation.
- Complete blood count
- Erythrocyte sedimentation rate
- Thyroid function tests
- Liver Functions
- EMG-NCV
- Age appropriate cancer screening
- Specific Genetic testing- Cadisil, MELAS, etc



## Differences Between McArdle Disease and CPT Deficiency

	<b>McArdle Disease (glycogenosis V)</b>	<b>CPT Deficiency</b>
<b>Metabolic defect</b>	<b>Glycogen storage</b>	<b>Lipid storage</b>
<b>Exercise</b>	<b>Usually cramps with short strenuous exercise</b>	<b>Usually myalgia and tenderness (without cramps) with prolonged exercise, worse with fasting</b>
<b>Second-wind phenomenon</b>	<b>Present</b>	<b>Absent</b>
<b>Recurrent myoglobinuria</b>	<b>Less frequent (50% of patients)</b>	<b>Common</b>
<b>CK at rest</b>	<b>Increased</b>	<b>Normal</b>
<b>Ischemic forearm exercise test</b>	<b>Absence of normal increase in lactate level</b>	<b>Normal</b>
<b>Muscle biopsy</b>	<b>Usually shows glycogen accumulation</b>	<b>May be normal</b>
<b>Gene location</b>	<b>Band 11q13</b>	<b>Band 1p32 (CPT II)</b>

# Headaches, Motor Disorders, Amyotrophies

## Duchenne Muscular Dystrophy

X linked

Progressive weakness

Begins at 2 until young adult

Weakness: proximal>distal

Elevated CPK

Treatment

- Exondys 51 –(eteplirsen)

- Deflazacort

# Headaches, Motor Disorders, Amyotrophies

## **Myotonic Dystrophy**

Inherited neuromuscular disorder

Autosomal dominant

Symptoms-

Weakness

Sleep apnea

Cardiac conduction defects

Mitral valve prolapse

Testicular atrophy

# Headaches, Motor Disorders, Amyotrophies

## Mitochondrial

Mitochondrial myopathy (MELAS)

Inherited maternal

Defect of the mitochondria

Lactic acidosis

Muscle weakness/ptosis/neurological

Cardiomyopathy - arrhythmias

Liver/Kidney problems

Stroke before 40

Red ragged fibers on biopsy

# Headaches, Motor Disorders, Amyotrophies

## **Metabolic**

Addison disease, particularly when fluid and electrolyte problems are present

Cushing disease

Hypothyroidism (CK may be mildly elevated)

Hyperthyroidism (CK may be normal)

Hyperparathyroidism

Conn Syndrome

# Headaches, Motor Disorders, Amyotrophies

## **Periodic Paralysis:**

Normokalemic paralysis causes the **most severe and prolonged** attacks.

Patients usually feel well between attacks, but some have myotonia or residual weakness after repeated episodes.

Acute hypokalemic periodic paralysis may be primary (ie, familial) or secondary to excessive renal or GI losses or endocrinopathy.

Intracellular shift of potassium depolarizes the cell membrane rendering it inexcitable and no muscle contraction can occur.

**Familial periodic paralysis** usually occurs in Caucasian males, is autosomal dominant, and may last as long as 36 hours.

Attacks usually **occur at night or in early morning** upon awakening and can be precipitated by a diet high in carbohydrates, rest following exercise, or glucose and insulin given intravenously.

# Headaches, Motor Disorders, Amyotrophies

## • Inflammatory

- Dermatomyositis / Polymyositis
  - Proximal muscle weakness
  - EMG- myopathic changes consistent with inflammation
  - MRI- shows inflammatory component
  - **Responds to glucocorticoids**
- Inclusion Body Myositis
  - Does NOT respond to steroids
  - BX shows vacuolar inclusions with eosinophils

Headaches, Motor Disorders, Amyotrophies

# Infections

## **Spirocete**

Lyme

## **Bacterial**

Staphylococcal ,Tuberculosis, Clostridium

## **Viral**

HIV, Influenza, EBV, CMV, Coxsackie, Adenovirus



# Headaches, Motor Disorders, Amyotrophies

## Toxic

Ingestion of chemicals or pharmaceuticals:

ETOH

Statins/Fenofibrates

Steroids

AZT

Cocaine

Diuretics

Amiodarone

Colchicine

# Headaches, Motor Disorders, Amyotrophies

## Myasthenia Gravis

Autoimmune- motor end plate disorder

**Associated with thymomas**

Diplopia and ptosis is common

Symptoms worsen as day progresses

Diagnosis:

Anti-Acetylcholine receptor antibodies

**Tensilon test** (while ptosis present)

Treatment:

Anti-cholinesterase agents (mestinon)/ thymectomy

In crisis- Plasma exchange/IVIG

Headaches, Motor Disorders, Amyotrophies

# Lambert-Eaton

Associated with Oat cell carcinoma

Autoimmune

Presynaptic peripheral nerves antibodies that causes acetylcholine release to decrease

Proximal muscle weakness

Dry mouth

Hypo-reflexia- esp lower extremities

Treatment: Anti-cholinesterase agents