HEADACHES, MOTOR DISORDERS, AND AMYOTROPHIES

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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*

Prophylactic

-Beta blockers

-Tricyclic Antidepressants

-Divalproex

-Topiramate

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-HT1 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 H2O — 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

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 sucessful

    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-Palliodotomy

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

Drug Class	Example	Initial Dosage	Usual Dosage	Side Effects
First-line dopaminergic agents				
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		—	
Dopamine agonists				
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
Second-line alternatives				
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
	Benztropine (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[20]). MAO-B denotes monoamine oxidase B, SSRI selective serotonin-reuptake inhibitor, and NMDA *N*-methyl-D-aspartate.

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

Tardive Dyskinesia

- Effect of Long term antipsychotics
- Involves Lips, tongue, face, and neck
- Can affect limbs
- Treatment
  - Exchanging the dopamine antagonist antipsychotic

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

Headaches, Motor Disorders, Amyotrophies

Tic Douloureux

- Hemifacial spasm
- Pain
- Trigeminal neuralgia
- 80% have basilar artery affecting the facial n.
- Treatment: Carbamazine/Surgery

Headaches, Motor Disorders, Amyotrophies

Other:

-Giles 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.

TYPE OF SEIZURE	CLINICAL FEATURES	ELECTROENCEPHALOGRAPHIC FEATURES*
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

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

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 Paralyisis:

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

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

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

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