Chorea, Ataxia, and other Movement Disorders

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Control of motor function

Descending Systems
- Upper Motor Neurons
  - Motor Cortex: Planning, initiating, and directing voluntary movements
  - Brainstem Centers: Basic movements and postural control

- BASAL GANGLIA: Gating proper initiation of movement
- CEREBELLUM: Sensory motor coordination

Local circuit neurons: Reflex coordination
- Motor neuron pools: Lower Motor Neurons
- Spinal cord and brainstem circuits
- Skeletal muscles
Control of motor function

• The activities of the basal ganglia and cerebellum modulate the corticospinal and cortical-brainstem-spinal systems.
• Often referred to as extra-pyramidal system
Movement disorders

Hyperkinetic Movements

Hypokinetic Movements
Categorization of Movement Disorders

Movement Disorders

- Pyramidal Syndromes
  - Spasticity

- Basal Ganglia Disorders

- Cerebellar Disorders
  - Ataxia

Hypokinesias
- Akinesia
- Rigidity
- Tremor

Hyperkinesias
- Dystonia
- Myoclonus
- Chorea
- Athetosis
- Tics
- Stereotypies

Miscellaneous
- Motor
- Sensory
- Behavioral
- Compulsions
- Mannerisms
- Akathisia
- Restless Legs

Jankovic
Epidemiology of hyperkinetic movement disorders in Africa per 100,000

<table>
<thead>
<tr>
<th>Study</th>
<th>Chorea</th>
<th>Athetosis</th>
<th>Dystonia</th>
<th>Cerebellar ataxia</th>
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<tbody>
<tr>
<td>El Tallawy et al. 2013 (Al Quseir, Egypt)</td>
<td>21.0</td>
<td>15</td>
<td>39.1</td>
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<td>El Tallawy et al. 2010 (Al Kharga, Egypt)</td>
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<td>Kandil et al. 1994 (Assiut, Egypt)</td>
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### Hypokinetic Movement Disorders

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Frequency</th>
<th>Valid percent</th>
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<tbody>
<tr>
<td>Idiopathic PD</td>
<td>85</td>
<td>69.7</td>
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<tr>
<td>Drug induced PD</td>
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<td>Vascular PD</td>
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<td>4.1</td>
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</table>

### Hyperkinetic Movement disorders

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Frequency</th>
<th>Valid percent</th>
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</thead>
<tbody>
<tr>
<td>Essential tremor</td>
<td>14</td>
<td>11.5</td>
</tr>
<tr>
<td>Cerebellar/ thalamic tremor</td>
<td>11</td>
<td>9.0</td>
</tr>
<tr>
<td>Myokymia</td>
<td>2</td>
<td>1.6</td>
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<tr>
<td>Choreo-athetosis</td>
<td>2</td>
<td>1.6</td>
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<tr>
<td>Dystonia</td>
<td>1</td>
<td>0.8</td>
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<tr>
<td>PD plus syndrome</td>
<td>1</td>
<td>0.8</td>
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<tr>
<td>Tics</td>
<td>1</td>
<td>0.8</td>
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<tr>
<td>Writers cramps</td>
<td>0</td>
<td>0.0</td>
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<tr>
<td>Tardive dyskinesia</td>
<td>0</td>
<td>0.0</td>
</tr>
<tr>
<td>Restless leg</td>
<td>0</td>
<td>0.0</td>
</tr>
</tbody>
</table>

**Total** 122 100
Hyperkinesias

• Due to imbalance of activity in the complex basal ganglia circuits
• Related to excessive dopaminergic activity in the basal ganglia
Cortico-Basal-thalamic circuits
Basal ganglia circuits
Chorea, athetosis, ballism, dystonia

• Not a single entity, but a spectrum
• Myoclonus  Ballismus  Chorea  Athetosis  Dystonia

Movements become – less violent / explosive / jerky
- smoother and more flowing
- more sustained
Chorea

• Chorea means ‘dance”
• Irregular, unpredictable brief jerky/fidgety movements that are usually of low amplitude
• Semi-purposeful
• If subtle, it can be missed
• May be generalised/segmental/localised
• Videos
Causes of chorea

• Hereditary
  – Dominant- Huntington’s disease
  – Recessive- Wilson’s disease
• Autoimmune
  – Rheumatic chorea (Sydenham’s)
  – Chorea gravidarum
  – Systemic lupus erythematosus
• Metabolic
  – Hypo-hypernatraemia, hypo-hyperglycaemia, hypocalcaemia, renal failure
• Toxins- mercury, carbon monoxide
• Drugs- neuroleptics (ie TARDIVE DYSKINESIA), metoclopramide, L-dopa, anticonvulsants, steroids, oral contraceptives
• Inflammatory
  – HIV/AIDS
  – encephalitis
• Vascular
  – Basal ganglia infarcts
Causes of chorea in Africa

- 62/100,000 – Rheumatic chorea
- 21/100,000 – Huntington’s chorea
- 17/100,000 – Vascular disease
- 12/100,000 – Post-encephalitic

Sample of 42,000 participants from 7,000 families in Assiut, Egypt. (Kandil et al., 1994)
Athetosis

- Athetosis is a slow continuous stream of sinuous, writhing movements, typically of the hands and feet
- Causes
  - dyskinetic motor fluctuation in PD
  - athetoid cerebral palsy
- If athetosis becomes faster, it sometimes blends with chorea i.e. choreoathetosis or choreo-athetoid movements
Myoclonus

• Brief, isolated, random, non-purposeful jerks of muscle groups in the limbs.
• Generalised/ segmental/ focal
• Rx. Valproate drug of choice.
• May respond to benzodiazepines eg clonazepam
• Video
Ballismus

- Essentially a focal form of chorea (dramatic)
- Hemiballism – proximal high-amplitude flailing chorea limited to one side of the body
- Biballism if bilateral
- Monoballism if in a single limb

Causes
- Usually vascular (e.g., basal ganglia infarct; usually of the sub-thalamic nucleus)
- Metabolic
- Good response to antipsychotics, usually self-limiting
Tics

- Consist of abnormal movements and abnormal sounds
- Vary in severity over time
- Usually preceded by uncomfortable feeling or sensory urge that is relieved by carrying out the movement
- May be simple or complex
- Often suppressible
- Video
Stereotypy

- Refers to coordinated movements that repeat continually and identically
- Resemble motor tics but there is no driving urge
- Often repeat themselves in a uniform, repetitive fashion for long periods of time
Akathisia

• “Unable to sit still”
• Feeling of inner, general restlessness that is reduced or relieved by moving about
• Complex and usually stereotyped movements
• Can be both generalized and focal
• Can usually be briefly suppressed
• Most common cause is iatrogenic
Approach to a patient with hyperkinesia

• **History taking**
  - Patient description of problems
  - Course of present illness
  - Age at onset
  - Past medical history
  - Birth and family history
  - Infections
  - Drug/toxin exposure (eg neuroleptics, alcohol)
Approach to a patient with hyperkinesia

- Neurological examination
  - spend time *observing* the *patient overall*
- Establish **type** of movement disorder
  - hyperkinetic
- Establish **pattern** of movement disorder
  - Focal vs segmental vs generalised
  - Particular characteristics of the movement disorder
    - Rhythmic/intermittent
    - Rest/posture/action
    - Precipitating features (startle/posture/task specific/drugs
    - Associated features – eg. Dementia, epilepsy
- Establish **aetiology** of movement disorder
Aetiology of dyskinesias

• Hereditary
  - Huntington’s dx
  - Wilson’s dx
  - Neuroacanthosis
  - Porphyria
• Childhood
  - Cerebral birth injury
  - Kernicterus
  - Cerebral palsy
• Infective/inflammatory
  - Post-streptococcal
  - Henoch-Scholein purpura
  - Creutzfeldt-Jakob disease
  - SLE
• Drugs
  - Levodopa/dopamine agonists
  - Phenothiazines
  - tricyclics
• Vascular
  - CVA (infarct / hemorrhagic)
  - Arteriovenous malformation
  - Antiphospholipid antibody syndr
• Endocrine
  - Pregnancy
  - Thyrotoxicosis
  - Hypoparathyroidism
  - hypoglycaemia
ATAXIA

• Ataxia is the inability to perform smooth, accurate and coordinated movements
• Ataxia can arise from disorders of
  • Cerebellum (most common)
  • Sensory pathways (sensory ataxia)
  • Frontal lobe lesions (via fronto-cerebellar associative fibres)
Cerebello-cortical loops
Motor ataxia

• Motor Ataxia
  – Caused by cerebellar disorders
    • Intact sensory receptors and afferent pathways
    • Integration of proprioception is faulty
    • Midline cerebellar lesions cause truncal ataxia
    • Lateral cerebellar lesions cause limb ataxia
    • Thalamic infarcts may cause contralateral ataxia with sensory loss
Sensory ataxia

- Sensory Ataxia
  - Failure of proprioceptive information to the CNS
  - May be due to disorders of spinal cord or peripheral nerves
  - Can be compensated for by visual inputs
Approach to patient

• History
  – Onset
  – Rapidity
  – Previous symptoms
  – PMH
  – Medications
  – Social
    • Alcohol intake
    • Illicit drug use

• Associated Symptoms
  • Headache
  • Drowsiness
  • Dizziness
  • Vertigo
  • Tinnitus
  • Fever
  • Nausea/vomiting
  • Weakness
  • Paresthesia
Approach to patient

- Physical Exam
  - Gait testing
  - Tandem gait
  - Orthostatic VS

- Full neurologic exam
  - Cerebellar function
    - Dysmmetria
    - Dysdiadochokinesia
    - Dyssynergia
    - Stewart-Holmes rebound sign
    - Rhomberg
ATAXIA CAUSES

- Acquired vs inherited
- Acute vs subacute vs chronic
Ataxia causes

- Acquired
- Infection (abscess, cerebellitis)
- Vascular (bleed, infarct, TIA)
- Demyelinating (MS, PN)
- Malignancy (tumor)
- Toxin (INH, Li, cyclosporin, cytosine, phenytoin)
- Paraneoplastic
- Metabolic (Vit E deficiency, hypothyroidism)
Ataxia causes

- Inherited
- Intermittent hyperammonia
- Progressive
  - Mitochondrial encephalomyopathy
  - Wilson’s disease
Investigations

- Rule out acquired causes
- CT scan or MRI
- LP
- EMG, NCS
- TFT
- Lipids
THANK YOU FOR LISTENING