

**MOVEMENT DISORDERS:**  
**Identification and Characterization**  
**of the Abnormal Movements**

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# General concepts

- Movement Disorder
  - Term for a physical sign
  - Term to describe a specific syndrome/condition
- Either excess of movement or paucity of voluntary and automatic movements, unrelated to weakness or spasticity
- Diagnosis of movement disorders requires:
  - **Identify the type and pattern of movement**
  - Isolated or accompanied with other neuro signs
  - Determine probable etiology

# Clinical phenomenology

## Hypokinesias

- **Akinesia/bradykinesia (parkinsonism)**
  - Rigidity
- **Freezing**
- **Apraxia**

## Hyperkinesias

- **Tremor**
- **Dystonia**
- **Myoclonus**
- **Chorea**
  - Ballism
  - Athetosis
- **Tics**
- **Ataxia**
  - Dysmetria
- **Stereotypy**
- **Akathisia**
- **Hyperekplexia**
- **Hemifacial spasms**
- **Restless leg Syndrome/Periodic movements of sleep**
- **Tardive movements**
- **Paroxysmal Dyskinesias**

- **Automatic movements:** learned motor behaviors performed without conscious effort (walk, speak)
- **Voluntary movements:** intentional planned or self initiated, or externally triggered
- **Semi-voluntary (un-voluntary):** induced by an inner sensory stimulus, move to suppress unpleasant sensation, suppressible for short time (tic, akathisia, RLS)
- **Involuntary :** not suppressible (tremor, myoclonus)

Pathology in basal ganglia or connections,  
subthalamic nucleus, substantia nigra EXCEPT:

Cerebellum : asynergy, ataxia, dysmetria  
intention tremor

Myoclonus and many forms of tremors: not  
basal ganglia pathology; arise from cortex,  
brainstem

Tics : basal ganglia and limbic implicated,  
uncertain

# Structures = movement

- Substantia nigra = bradykinesia, rest tremor
- Subthalamic nucleus = ballism
- Caudate nucleus = chorea
- Putamen = dystonia

# Characteristics to classify movements

- Distribution
- Velocity
- Amplitude
- Stereotypy
- Rythmicity
- Suppressibility
- Relationship to position, sleep, activity

# HYPOKINESIAS

# Bradykinesia

- Slowness of movement (parkinsonism)
- Reduced amplitude of voluntary movement
- Slow initiating movement on command
- Loss automatic movements
- Short shuffling steps
- Loss spontaneous movement (gestures)
- Hypomimia (decreased blink)
- Hypophonia
- Aprrosody
- Drool (decreased spontaneous swallow)



# Rigidity

- Increased muscle tone to passive motion
- Present equally in all direction of the passive movement throughout the range of motion
- Distinguish from spasticity (velocity dependent)
- Distinguish from paratonia (inability to relax)

# Freezing

- Motor act halted transiently (several seconds)
- Agonists and antagonist muscles are simultaneously and isometrically contracting
- Start hesitation, turning hesitation, destination hesitation, freeze with obstacle
- Typical and atypical parkinsons

# Apraxia

- Inability to perform complex learned voluntary motor movements (not due to weakness, spasticity, rigidity, sensory loss)
- See in Dementia, atypical parkinsonism, structural lesions (strokes, tumors)



# Frontal gait disorder

- Start hesitation
- Slow small shuffling steps with out other parkinsonian features
- Frontal lobe tumors, CVA, hydrocephalus, severe subcortical white matter changes



# HYPERKINESIAS

# Hyperkinesia

- Establish movement is involuntary
  - exaggerated gestures, mannerisms or compulsive movement
  - Sustained contracted muscle (involuntary) vs. muscle guarding for pain reduction
- Abnormal involuntary movements are exaggerated with anxiety and diminish during sleep

# Tremor

- Oscillatory, rhythmical and regular movement
- Produced by rhythmic alternating or simultaneous contractions of agonists and antagonists
  - **Physiologic tremor**
  - **Rest tremor**
  - **Action tremor** – during voluntary contraction muscles
    - **Postural tremor** – voluntarily maintained against gravity
    - **Kinetic tremor** – during any voluntary movement
      - Simple kinetic tremor – during non target directed voluntary movement
      - Intention tremor – with increasing amplitude at end of movement
    - **Task-specific tremor** – during specific activity



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# Essential Tremor

## Diagnostic criteria

- Bilateral, largely symmetric
- Postural or kinetic tremor
- Hands and forearms
- Additional or isolated tremor of head, without abnormal posturing
- Long duration >3 years
- Positive family history
- Beneficial response to alcohol

## Red Flags indicating not ET

- Unilateral tremor, leg tremor, rigidity, bradykinesia, rest tremor - PD
- Gait disturbance – PD, cerebellar tremor
- Focal tremor – dystonic tremor
- Isolated head tremor – dystonic tremor
- Sudden or rapid onset – psychogenic tremor, toxic tremor
- Current drug treatment that may cause or exacerbate tremor – drug induced or toxic tremor



# Other Tremors

- **Dystonic tremor**
  - Irregular amplitude
  - Involve any body part affected by dystonia
  - 4-7 Hz
- **Cerebellar tremor**
  - Slower <5Hz
  - Intention tremor (postural may be present)
  - Usually unilateral
  - Proximal > distal muscles
- **Holmes' tremor**
  - Irregular combination of rest, posture and kinetic (often intention) tremor
  - Slow 2-4 Hz
  - Proximal > distal muscles
  - Midbrain lesion
- **Drug induced/toxic tremor**
  - Full range tremor phenomenology, 2-12 Hz, variable severity
  - Exacerbate normal physiologic tremor or cause other types of tremor
  - Asthma meds, cardiac antiarrhythmics, lithium , amphetamines, valproic acid, neuroleptics

# Dystonia

- Involuntary, sustained muscle contraction, frequently causing twisting and repetitive movements and abnormal postures.
- Progress to prolonged abnormal postures
- Repeatedly involve the same group of muscles (unlike chorea)
- Relatively long duration (compared to myoclonus and chorea)
- Agonists and antagonists contract simultaneously
- Dynamic disorder that changes severity depending on activity or posture.
  - Speed varied from slow (athetoid dystonia) to shock like (myoclonic dystonia)

# Features unique to Dystonia

- **TASK SPECIFIC:** selective activation of involuntary movements by specific tasks (write, computer mouse, musical instrument)
- **GESTE ANTAGONISTE:** sensory trick improves the dystonia while it is applied
- **FUNCTIONAL VARIABILITY:** variation in severity of dystonia with specific actions (present walk forward not backward, present while talk not eat)

# Dystonia

- **Primary Dystonia**
- Dystonia in isolation without other neurologic abnormality
- Focal/segmental/generalized
  - Young onset: start LE then generalize
  - Adult onset: start cervical/cranial remain focal/segmental
    - Blepharospasms
    - Cervical dystonia
    - Spasmodic dysphonia
    - Focal limb dystonia
- Genetic/sporadic
- **Secondary Dystonia**
- More likely to begin at rest
- Structural cerebral lesion
  - CP, infection, trauma, vascular insult
- Drug induced
  - Acute dystonic reaction
  - Tardive dystonia
  - Anticonvulsants
- **Hereditary dystonia**
  - Dystonia occurs as feature of a generalized inherited neurodegenerative disorder
  - Wilson's ds, lysosomal storage ds, inborn errors metabolism



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# Primary Dystonia

- Primary torsion dystonia (DYT1)
  - Autosomal Dominant, Torsin A gene mutation
  - Onset childhood, usually one LE, initially action induced, progressive generalization
- Dopa responsive dystonia
  - AD, mutation GTP cyclohydrolase 1 gene
  - Onset childhood, usually one LE, diurnal variation
  - Marked response to low dose carbidopa-levodopa



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# Myoclonus

- Sudden brief shock like involuntary movements from muscle contraction (positive myoclonus) or inhibition (negative myoclonus)
  - Rhythmic or arrhythmic
  - Generalized, focal or multifocal
  - Stimulus sensitive or action sensitive
  - Symmetric or asymmetrical
- Involuntary movement – no preceding urge as seen in Tic
- Arise from any point in neuroaxis
  - Cortex – can be associated with seizures
  - Subcortical
  - Brainstem
  - Spinal cord
  - Peripheral nerve

# Differential diagnosis of Myoclonus

- Physiologic – hypnic jerk, hiccup, benign infantile myoclonus
- Epileptic – epilepsy partialis continua, infantile spasms, juvenile myoclonic epilepsy
- Progressive myoclonic epilepsy – inborn errors metabolism, lysosomal storage diseases, mitochondrial disorders, etc.
  - Heterogeneous group of disorders characterized by epilepsy, myoclonus, progressive neurological deterioration
- Symptomatic
  - Post hypoxic
  - Post traumatic
  - Myoclonic dementias – CJD, AD, LBD
  - Toxic
  - Metabolic
  - Drug induced
  - Post infectious
  - Inflammatory
  - Neurodegenerative basal ganglia disorders – CBGD, PD, HD, MSA, etc.

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# **Pilot Tolerability and Efficacy Trial of Sodium Oxybate in Ethanol-Responsive Movement Disorders**

**SJ Frucht, Y Bordelon,  
WH Houghton, D Reardan**

*Movement Disorders*  
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# Symptomatic Myoclonus

- **Metabolic**
  - Hepatic failure
  - Renal failure
  - Dialysis dysequilibrium
  - Hyponatremia
  - Hypoglycemia
  - Nonketotic hyperglycemia
- **Toxic**
  - Bismuth
  - Aluminum
  - Mercury
  - Toluene
  - Dichloroethane (dry cleaning fluid)
  - Oven cleaner
  - Drugs of abuse (THC, cocaine, methamphetamine)
- **Medications**
  - Psychotropic:
    - tricyclic antidepressants, SSRI's, MAO-I, lithium, buspirone, antipsychotic agents, benzodiazepine withdrawal
  - Anticonvulsants
    - Phenytoin, valproic acid (asterixis), carbamazepine, gabapentin, lamotrigine
  - Narcotics
    - Morphine, meperidine, hydromorphone, fentanyl
  - Cardiovascular drugs
    - Diltiazem, nifedipine, verapamil, flecainide, propafenone, carvedilol
  - Other meds
    - Levodopa, metoclopramide, pseudoephedrine, albuterol, physostigmine, spinal anesthesia, propofol, amantadine, cimetidine, diclofenac
  - Antibiotic/antiviral
    - Penicillin, carbenicillin, ticarcillin, cefmetazole, monolactam, isoniazid, piperazine, acyclovir, vidarabine, gatifloxacin

# Chorea

- Involuntary, irregular, purposeless, non-rhythmic, rapid, unsustained movements
- Flow from one part body to another
- Unpredictable in timing, direction, distribution (random)
  - Other rapid non-choreic movements: tics, myoclonus, dystonia (repeat themselves in set distribution of the body)
- Partially suppressed, often camouflage into other semi-purposeful movements
- Chorea often accompanied by motor impersistence

# Differential diagnosis of Chorea

- Neurodegenerative
  - Huntington's Ds, other genetic neurodegenerative Disease (Wilson's, spinocerebellar ataxia)
- Neurometabolic Disorders
  - Lysosomal storage disease, mitochondrial Ds., amino acid disorders
- Drug induced
  - Oral contraceptives
  - Anticonvulsants
  - Antiparkinsons medications
  - Thyroid replacement
  - Tricyclics
  - Stimulants – cocaine, amphetamines
  - Neuroleptics
    - Tardive
    - Withdrawal emergent syndrome



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# Differential diagnosis Chorea

- Toxin
  - Ethanol (intoxication/withdrawal)
  - Carbon monoxide
  - Mg, mercury, thallium, toluene
- Immune mediated
  - Sydenham's
  - Post infection
  - SLE
  - Antiphospholipid Antibody Syndrome
  - MS
  - Vasculitis
- Metabolic/Infectious
  - Pregnancy (chorea gravidarum)
  - Hyperthyroidism
  - Hyperglycemia
  - Hyponatremia
  - HIV
  - Borreliosis



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# Athetosis

- Slow, writhing, continuous, involuntary movements
- Affect limb especially distally, can involve face neck, tongue, axial structures
- Often blends with chorea (choreoathetosis)

# Ballism

- Large amplitude choreic movements of proximal parts of limbs
- Flinging and flailing limb movements
- Lesion in contralateral subthalamic nucleus or connections, or multiple lacunes in contralateral striatum or over dosage levodopa



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# Tics

- Abnormal movement (motor tics) or abnormal sounds (phonic tics) or both tourette syndrome
- Vary in severity over time, remissions and exacerbations
- Simple or complex, occur abruptly for brief moments from a background of normal activity
- Precede by urge (un-voluntary) , can be suppressed for various periods of time, inner tension, relieved by increased burst of tics

# Differential Diagnosis TICS

## Primary tic disorder

- Transient
  - Motor or vocal < 1 year
  - 15% children (male>female)
  - Mild usually single movement
- Chronic single tic disorder
  - Motor or vocal > 1 year
- Adult onset (recurrent) tics
- Tourette syndrome
  - Motor and vocal > 1 year
  - Onset < 21 year old

## Secondary tic disorders

- Drugs
  - CNS stimulants: amphetamines, methylphenidate, pemoline, cocaine
  - Neuroleptics: tardive tics
  - Levodopa
  - Anticonvulsants: carbamazepine, lamotrigine, phenytoin, phenobarbital
- Hereditary
  - HD, Wilsons, others
- Neurodevelopmental disorders
  - MR, perinatal injury, chromosomal disorders
- Brain injury
  - Stroke, encephalitis, trauma, CO poison
- Infections
  - Sydenham's chorea, PANDAS
  - Postviral encephalitis, lyme, HIV



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# Stereotypy

- Coordinated movements that repeat themselves continually and identically
- Irregular intervals between movements
- Distinguish from tic, compulsions, gestures and mannerisms
  - Tic and compulsion preceded by urge
- See in patients with tardive dyskinesia, schizophrenia, mental retardation, autism



Seit es keine Zeit für die Teilnahme an Aktivitäten auf der Arbeit gibt, ist die Stimmung im Büro oft schlechter. © iStockphoto.com

# Conclusions

## Recognize the clinical phenomena

### **Hypokinesias**

- Akinesia/bradykinesia (parkinsonism)
- Freezing
- Apraxia

### **Hyperkinesias**

- Tremor
- Dystonia
- Myoclonus
- Chorea
- Tics

# Akathesia

- Feeling of inner general restlessness
- Sensation reduced by moving
- Complex movements, stereotyped
  - Other complex movements: tics, compulsions, stereotypies
- Can be vocalizations (humming, moaning)
- Transiently suppressed
- Etiology: antidopaminergic meds

# Restless Leg Syndrome

- Criteria for RLS diagnosis
  - Urge to move (un-voluntary movement) the limb with uncomfortable sensations
  - Symptoms are worse or exclusively at rest or during periods of inactivity
  - Symptoms are partially or totally relieved by movement
  - Circadian rhythm: symptoms must be worse or exclusively in the evening or night

# Etiology RLS

- Role of Iron – ferritin decreased/transferrin increased suggests low brain iron stores
- Role of Dopamine – RLS and akathisia occur with dopamine antagonists

# Etiology of RLS

- **Primary (idiopathic)**
  - Most cases
  - Hereditary (AD)
  - Multiple chromosomal abnormalities identified as possible loci
- **Secondary (symptomatic)**
  - Iron deficiency
  - Pregnancy
  - End stage renal disease
  - Medications
    - TCA, SSRI's, MAO-I, lithium, antihistamines, dopamine antagonists
    - Other: caffeine, smoking, alcohol
  - Peripheral neuropathy