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Chase Family Movement Disorders Center

TWITCH, JERK or SPASM Movement Disorders Seen in Family Practice

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DEFINITION OF TERMS

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- Movement Disorders – neurological syndromes in which there is either an excess of movement or a paucity of voluntary and automatic movements, unrelated to weakness or spasticity
- Hyperkinesias – excess of movements
- Dyskinesias – unnatural movements
- Abnormal Involuntary Movements – non-suppressible or only partially suppressible
- Hypokinesia – decreased amplitude of movement
- Bradykinesia – slowness of movement
- Akinesia – loss of movement

CLASSES OF MOVEMENTS

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- Automatic movements – learned motor behaviors performed without conscious effort, e.g. walking, speaking, swinging of arms while walking
- Voluntary movements – intentional (planned or self-initiated) or externally triggered (in response to external stimulus, e.g. turn head toward loud noise, withdraw hand from hot stove)
- Semi-voluntary/"unvoluntary" – induced by inner sensory stimulus (e.g. need to stretch body part or scratch an itch) or by an unwanted feeling or compulsion (e.g. compulsive touching, restless legs syndrome)
- Involuntary movements – often non-suppressible (hemifacial spasms, myoclonus) or only partially suppressible (tremors, chorea, tics)

HYPERKINESIAS: major categories

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- CHOREA
- DYSTONIA
- MYOCLONUS
- TICS
- TREMORS

HYPERKINESIAS: subtypes

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Abdominal dyskinesias	Jumpy stumps
Akathisic movements	Moving toes/fingers
Asynergia/ataxia	Myoclonus
Athetosis	Myokymia
Ballism	Myorhythmia
Chorea	Paroxysmal dyskinesias
Dysmetria	Restless legs
Dystonia	Stereotypies
Hemifacial spasms	Tics
Hyperekplexia	Tremors

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HYPERKINESIAS: localization

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- Usually associated with pathologic alterations in the basal ganglia or their connections (e.g. caudate/putamen/pallidum within cerebral hemispheres, subthalamic nucleus in diencephalon, substantia nigra in mesencephalon)
- Some conditions are classically associated with specific localization:
  - Substantia nigra – bradykinesia, rest tremors
  - Subthalamic nucleus – ballism
  - Caudate nucleus – chorea
  - Putamen - dystonia

HYPERKINESIAS: localization

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- Exceptions to basal ganglia localization
  - Cerebellum/cerebellar pathways – dysmetria, ataxia, intention tremors
  - Cerebral cortex – cortical reflex myoclonus
  - Brainstem – reticular reflex myoclonus, hyperekplexia, palatal myoclonus, ocular myoclonus
  - Spinal cord – segmental myoclonus, propiospinal myoclonus

HYPERKINESIAS: general evaluation

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- FIRST QUESTION – is this an involuntary movement?
  - May be a purposeful voluntary movement – exaggerated gestures, mannerisms, compulsive movements
  - May be guarding against pain – sustained contraction of muscles to reduce pain
  - GENERAL RULE – abnormal involuntary movements are exaggerated with anxiety, increase with distraction, diminish during sleep

HYPERKINESIAS: general evaluation

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- SECOND QUESTION: what is the nature of the involuntary movement?
  - General category: chorea, dystonia, myoclonus, tics, tremor
  - Features: rhymicity, speed, duration, pattern, induction (stimulus-induced, exercise, action), complexity of movements (simple vs complex), suppressibility (by volitional attention or sensory tricks), sensory accompaniments (restlessness, pain, urge)
  - Involved body parts

HYPERKINESIAS: general evaluation

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- THIRD QUESTION: what are the possible etiologies of this involuntary movement?
  - Hereditary
  - Sporadic
  - Symptomatic or Secondary
  - Formulate differential diagnosis on the basis of initial categorization and clinical features, then with additional information from diagnostic work up

HYPERKINESIAS: general evaluation

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- LAST QUESTION: how is this movement disorder best treated?
  - Dopaminergic medications
  - GABAergic medications
  - Dopamine receptor blocking agents
  - Anticonvulsants
  - Tremorlytic agents
  - Anticholinergics
  - Opiate narcotics

AKATHISIA

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- From Greek, unable to sit still
- Characterized by feeling of inner general restlessness, reduced or relieved by moving about
- Typical movements: while sitting – cross/uncross legs, rock trunk, squirm in chair, get up from chair and pace, marching, moaning/groaning vocalizations
- May affect whole body or isolated body part
- Can be suppressed transiently
- Etiology: usually iatrogenic/medication induced, either acute (improves with discontinuation of drug) or chronic (may be exacerbated by withdrawal of drug, i.e. tardive

ATAXIA/DYSMETRIA/ ASYNERGIA

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- Asynergia - decomposition of movement due to breakdown of normal coordinated execution of voluntary movement, seen only with voluntary movement and not appreciated when limb is at rest
- Usually associated with hypotonia, loss of check (unable to stop precisely with voluntary ballistic movement) and rebound (sudden displacement of limb results in overcorrection)
- Dysmetria - tendency to miss target and worsens as target is approached, over/undershooting (hyper/hypometria)
- Dysdiadochokinesia – break up and irregularity of movement when rapid alternating movements of limb
- Gait ataxia – unsteady, wide based, body swaying, unable to tandem
- Differential Diagnosis: cerebellar or cerebellar tract dysfunction; multiple sporadic, iatrogenic, and familial etiologies

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ATHETOSIS

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- Slow writhing continuous involuntary movements, with direction of movement changing randomly and in a flowing pattern
- Usually affects the limbs, distal > proximal, but can also involve axial muscles, face, neck, tongue
- Can be augmented by voluntary motor activity if not present at rest, i.e. overflow
- Often associated with dystonia (sustained contractions) or chorea (faster movements)
- Differential diagnosis: iatrogenic, sporadic, secondary, familial
- Pseudoathetosis – distal athetoid movements due to loss of propioception (sensory deafferentation/sensory athetosis)

BALLISM

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- Very large amplitude choreic movements of proximal parts of the limbs causing flailing/flinging movements
- Usually unilateral, i.e. hemiballism
- Differential diagnosis: lesion of contralateral subthalamic nucleus or its connections, multiple small infarcts in contralateral striatum, iatrogenic dopaminergic excess

CHOREA

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- Involuntary irregular purposeless non-rhythmic, abrupt rapid unsustained movements which seem to flow from one body part to another
- Movements are unpredictable in timing direction and distribution (random)
- Can be partially suppressed and can camouflage some movements by incorporating into semipurposeful movements (parakinesias)
- Accompanied by motor impersistence, inability to maintain sustained contraction (tongue protrusion, milk-maid grip)
- Treatment with dopamine depleters (tetrabenazine), dopamine receptor blocking agents, benzodiazepines
- Differential diagnosis: Huntington's disease, Syndenham's chorea, iatrogenic dopaminergic excess, secondary

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DYSTONIA

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- Twisting sustained movements progressing to abnormal postures, frequently repetitive and involve same group of muscles (patterned)
- Speed of movement can be slow (athetotic dystonia), shock like (myoclonic dystonia), or very brief contractions (dystonic spasms)
- Initially noted with voluntary action but progression leads to occurrence at rest
- Can be focal, segmental, multifocal or generalized, can lead to fixed contractures when involves chronic posturing
- Treatment with BoNT, anticholinergics, GABAergics, levodopa, dopamine depleters (tetrabenazine)
- Differential diagnosis: DYT mutations, iatrogenic, secondary

HEMIFACIAL SPASMS

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- Unilateral facial muscle contractions, generally repetitive and continual, with occasional sustained spasms, but with periods of quiescence
- Induced by voluntary forceful contraction of facial muscles, with spasms occurring once the face relaxes
- Affects upper and lower facial muscles but eyelid closure is usually more bothersome than lower facial spasms; differentiate from blepharospasms which is bilateral eyelid closure/spasms
- Differential diagnosis: may be due to compression of CN VII by aberrant blood vessel

HYPEREKPLEXIA

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- "Startle disease" – excessive startle reaction to sudden unexpected stimulus
- Can be short jump or more prolonged tonic spasm causing falls; related to "jumping disorders"
- After initial jump to startle, may see automatic speech or automatic behavior (e.g. striking out)
- Differential diagnosis: familial or sporadic

PAINFUL LEG MOVING TOES
(or painful hand moving fingers)

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- Painful leg moving toes syndrome - toes of one or both feet in continual flexion-extension with some lateral motion, associated with deep pain in leg, movement has sinusoidal quality
- Movements and pain are continuous and occur even in sleep (albeit may be reduced)
- Leg pain usually more bothersome than movements
- Analogous disorder – painful arm moving fingers
- Differential diagnosis – may have peripheral nerve or lumbar root lesions/injury, secondary or familial; different from Restless Leg Syndrome

MYOCLONUS

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- Sudden brief shock like involuntary movements caused by muscular contraction (positive myoclonus) or inhibitions (negative myoclonus, e.g. asterixis)
- Usually irregular (arrhythmic) but can be rhythmic (palatal or ocular myoclonus)
- Synchronized when involving different body parts
- Occurs at rest, with voluntary action, or triggered by sudden stimuli like sound, light, visual threat or movement
- Persists in sleep
- Propiospinal myoclonus – segmental, rhythmic, and may involve flexion axial jerks triggered by distant stimulus traveling spinal pathway
- Differential diagnosis: structural lesion of brainstem or spinal cord (rhythmic or segmental myoclonus), hypoxia, familial degenerative disorders

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MYOKYMIA/SYNKINESIAS

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- Fine persistent quivering or rippling of muscles ("live flesh"), most common in facial muscles, can persist in sleep
- EMG shows regular groups of motor unit discharges, in doublets or triplets, occurring with a regular rhythmic discharge
- Also benign fasciculations (e.g. of orbicularis oculi), but not associated with characteristic EMG findings
- Synkinesias – involuntary movements in one part of face accompanying voluntary contraction of another part (post-Bell's palsy)
- Fasciculations – small amplitude intermittent contraction of muscles innervated by a motor unit (anterior horn cell disease)
- Differential diagnosis: usually due to pontine lesions, particularly Multiple Sclerosis (abates after wks or mos) or pontine glioma (persists indefinitely)

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PAROXYSMAL DYSKINESIAS

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- Mixed movements that occur "out of the blue" then disappear after seconds, minutes or hours; may be normal for months in between attacks or have multiple attacks per day
- Kinesigenic (PKD) – triggered by sudden movement, lasts secs to a few mins, dystonic/ballistic/choreic, familial or sporadic, responds to anticonvulsants
- Nonkinesigenic (PNKD) – triggered by stress, fatigue, caffeine/EtOH, lasts mins to hrs, familial or sporadic, may respond to benzodiazepines or acetazolamide

RESTLESS LEGS SYNDROME

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- Unpleasant crawling sensation in legs, particularly when relaxing in evening, before falling asleep or with prolonged sitting, disappears when walking or with movement
- May include periodic limb movements of sleep, myoclonic jerks, sustained dystonic postures, or stereotypic movements in the late evening
- Responds to dopaminergic medications, benzodiazepines, opiates
- Differential diagnosis: idiopathic or secondary

STEREOTYPIES

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- Coordinated movements that repeat continually and identically, may have intervals of several minutes or occur very frequent, occurs at irregular intervals
- Uniform repetitive movements for prolonged periods of time, e.g. classic tardive dyskinesias or orobuccolingual dyskinesias
- May respond to tetrabenazine, reserpine, propranolol
- Differential diagnosis: usually neuroleptic induced, or with mental retardation and autism

TICS

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- Motor (involuntary movements) and phonic tics (involuntary sounds), can be simple or complex, occurring abruptly for brief moments from a background of normal motor activity
- Usually vary in severity over time, have remissions and exacerbations
- Paroxysmal and usually repetitive repertoire, preceded by uncomfortable feeling or sensory urge that is relieved by completing the movement, can be transiently suppressed but followed by rebound increase burst of tics
- Responds primarily to dopamine receptor blockers, dopamine depleters
- Differential diagnosis: Tourette's syndrome, benign tics, neurodegenerative disorders

TICS

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- Simple tics: shoulder shrug, head jerk, blink, darting of eyes, nose twitch or an abrupt isolated movement indistinguishable from a myoclonic jerk
- Complex tics: coordinated pattern of sequential movements that appear in different body parts and not necessarily identical: touching tics, nose pick, head shake with shoulder shrug, leg kick, jumping, obscene gestures (copropraxia)
- Phonic tics: simple throat clearing, sniffing, grunts or complex verbalizations or cursing (coprolalia)

TREMORS

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- Oscillatory usually rhythmical and regular movement produced by alternating or simultaneous contraction of agonist and antagonist muscles, affecting one or more body part (e.g. limbs, neck, tongue, chin, vocal cords)
- Rate, location, amplitude and constancy varies with different types of tremors
- Occurs at rest (limb supported against gravity), with postural sustension (limb extended), action (writing or pouring water), intention (FTNT), or task/position specific (orthostatic tremors)
- Differential diagnosis: Parkinson's disease, Wilson's disease (wing-beating tremor), Essential tremors, cerebellar tremors, iatrogenic

DIFFERENTIAL DIAGNOSIS: rhythmicity

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RHYTHMICAL	ARRHYTHMICAL
TREMORS	AKATHISIC MOVEMENTS
DYSTONIC TREMORS	ATHETOSIS
MYORHYTHMIA	BALLISM
SEGMENTAL MYOCLONUS	CHOREA
OSCILLATORY MYOCLONUS	DYSTONIA
MOVING TOES/FINGERS	HEMIFACIAL SPASMS
PERIODIC MOVEMENTS OF SLEEP	HYPEREKPLEXIA
TARDIVE STEREOTYPIES	ARRHYTHMIC MYOCLONUS
	STEREOTYPIES
	TICS

DIFFERENTIAL DIAGNOSIS: sustained

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SUSTAINED CONTRACTION	NON-SUSTAINED CONTRACTION
RIGIDITY	ALL OTHERS
DYSTONIA	
OCULOGYRIC CRISIS	
PAROXYSMAL DYSTONIA	
DYSTONIC TICS	
STIFF PERSON	
NEUROMYOTONIA	
TORTICOLLIS	

DIFFERENTIAL DIAGNOSIS: periodicity

PAROXYSMAL	CONTINUAL	CONTINUOUS
TICS	BALLISM	ABDOMINAL DYSKINESIAS
PKD	CHOREA	ATHETOSIS
PNKD	DYSTONIC MOVEMENTS	TREMORS
PAROXYSMAL ATAXIA	ARRHYTHMIC MYOCLONUS	DYSTONIC POSTURING
PAROXYSMAL TREMOR	STEREOTYPY	CLASSIC TD
HYPNOGENIC DYSTONIA		MYOKYMIA
		TIC STATUS
		MOVING TOES
		RHYTHMIC MYOCLONUS

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PAROXYSMAL - episodic
CONTINUAL – over and over again
CONTINUOUS – without stopping or unbroken

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DIFFERENTIAL DIAGNOSIS: sleep

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APPEARS IN SLEEP	PERSISTS IN SLEEP	DIMINISHES IN SLEEP
HYPNOGENIC DYSKINESIAS	PALATAL MYOCLONUS	ALL OTHERS
PERIODIC LIMB MOVMTS	OCULAR MYOCLONUS	
	MYORHYTHMIA	
	MOVING TOES	
	MYOKYMIA	

DIFFERENTIAL DIAGNOSIS: action

AT REST ONLY	WITH ACTION ONLY	REST & CONTINUES W/ACTION
AKATHISIA	ACTION DYSTONIA	ABDOMINAL DYSKINESIAS
PARADOXICAL DYSTONIA	ACTION MYOCLONUS	ATHETOSIS
REST TREMOR	ORTHOSTATIC TREMOR	BALLISM
RESTLESS LEGS	POST/ACTION/INT TREMOR	CHOREA
	TASK-SPECIFIC TREMOR	DYSTONIA
	TASK-SPECIFIC DYSTONIA	MOVING TOES/FINGERS
		MYOCLONUS
		MYOKYMIA
		PSEUDODYSTONIAS
		TICS

DIFFERENTIAL DIAGNOSIS: patterned

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PATTERNED	NON-PATTERNED
ABDOMINAL DYSKINESIAS	ALL OTHERS
DYSTONIA	
HEMIFACIAL SPASMS	
MOVING TOES/FINGERS	
SEGMENTAL MYOCLONUS	
MYORHYTHMIA	
MYOKYMIA	
TARDIVE STEREOTYPY	
TREMOR	

** PATTERNED – same muscle groups

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DIFFERENTIAL DIAGNOSIS: combination

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COMBINATIONS OF VARIETIES OF MOVEMENTS
PSYCHOGENIC MOVEMENT DISORDERS
TARDIVE SYNDROMES
NEUROACANTHOCYTOSIS
WILSON'S DISEASE
HUNTINGTON'S DISEASE
DYSTONIA

DIFFERENTIAL DIAGNOSIS: speed

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FASTEST	INTERMEDIATE	SLOWEST
MYOCLONUS	CHOREA	ATHETOSIS
HYPEREKPLEXIA	BALLISM	MOVING TOES/FINGERS
HEMIFACIAL SPASMS	TREMORS	MYORHYTHMIA
	TARDIVE STEREOTYPY	AKATHISIA

NB: tics and dystonic movements can be of all speeds

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DIFFERENTIAL DIAGNOSIS: amplitude

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BALLISTIC	NON-BALLISTIC
BALLISM	CHOREA AND ALL OTHERS

DIFFERENTIAL DIAGNOSIS: force

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POWERFUL	INTERMEDIATE	EASY TO OVERCOME
STIFF PERSON	DYSTONIA	ALL OTHERS

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DIFFERENTIAL DIAGNOSIS: suppressibility

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SUPPRESSIBLE

stereotypy>tics/akathisia>chorea>ballism>dystonia>tremor

NON-SUPPRESSIBLE

hemifacial spasms, myoclonus, hyperekplexia, myorhythmia, moving toes

DIFFERENTIAL DIAGNOSIS: vocalization

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VOCAL TICS: SIMPLE or COMPLEX
AKATHISIA: MOANING
HUNTINGTON'S DISEASE
NEUROACANTHOCYTOSIS
CRANIOCERVICAL DYSTONIA

DIFFERENTIAL DIAGNOSIS: sensory

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AKATHISIA
MOVING TOES/PAINFUL LEG
RESTLESS LEGS SYNDROME
TICS

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Connect to healthier.