CHAPTER

81

Approach to Hyperkinetic Movement Disorders

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Movement disorders is a group of neurologic conditions that can be divided phenomenologically into slow movements(hypokinetic) and abnormal involuntary movements(hyperkinetic) (Table 1).

The hypokinetic disorders are characterized not only by slowness of movement but also by paucity of movements. The most characteristic feature of hypokinetic movement disorder is bradykinesia, typically present in Parkinsonian disorders.

Hyperkinetic disorders are subdividided into tremor, tics, myoclonus, athetosis, ballismus, chorea, dystonia, sterotype an akathisia. Furthermore ataxia, gait disorders and spasticity are also often included among movement disorders. While the basal ganglia and their connections have been implicated in the pathophysiology of most of the movement disorders, some are caused by altered peripheral input as exemplified by hemifacial spasm and other peripherally induced movement disorders. A subset of movement disorders with varied phenomenology is caused by psychological factors , hence referred to as 'psycogenic movement disorders'. Because the diagnosis of a movement disorder is based on recognition of specific phenomenological features, clinicians who encounter patients with movement disorder must use their powers of observation to carefully categorize the disorder. The phenominological categorization is absolutely critical for formulation of etiological diagnosis and selection of the appropriate treatment. Many hyperkinetic movement disorders manifest with multiple types of movements, which may include a combination of the various hyperkinesias.

The basal ganglia regulate the initiation, scaling, and control of the amplitude and direction of movement.

Table 1 : Classification of Movement Disorders

Hypokinetic	Hyper kinetic
Parkinsonism	Tremor
Apraxia	Tics
Blocking tics	Myoclonus
Catatonia	**A**thetosis
Cataplexy and drop attacks	Ballismus
Hypothyroid slowness	Chorea
	Dystonia
	Dyskinesia

Movement disorders can result from biochemical or structural abnormalities in these structures.

The basal ganglia are a complex of deep nuclei that consist of the corpus striatum, globus pallidus, and substantia nigra. The corpus striatum, which includes the caudate nucleus and the putamen, receives input from the cerebral cortex and the thalamus and, in turn, projects to the globus pallidus.

The substantia nigra is divided into the dopamine-rich pars compacta and the less dense pars reticularis. The pars reticularis is similar histologically and chemically to the medial segment of the globus pallidus, and both project via the thalamus to the premotor and motor cortex. The substantia nigra pars compacta gives rise to the nigral-striatal pathway, which is the main dopaminergic tract.

The output of the basal ganglia projects by way of the thalamus to the cerebral cortex and then to the pyramidal system. Basal ganglia output is sometimes referred to as the extrapyramidal system because it was formerly thought to be in parallel with the pyramidal system. Integration of the basal ganglia with the cortex facilitates motor control.

PHYSIOLOGICALLY MOVEMENTS CAN BE CATEGORIZED INTO ONE OF FOUR CLASSES

1. Automatic
2. Voluntary
3. Semivoluntary or unvoluntary
4. Involuntary

Automatic movements are learned motor behaviors that are performed without conscious effort.

Eg: The act of walking or speaking ;the swing of arms during walking

Voluntary movements are intentional (planned or self initiated) or externally triggered

Eg: Turning the head toward a loud noise or withdrawing a hand from a hot plate. Intentional voluntary movements are preceded by the Bereitschafts potential (or readiness potential), a slow negative potential recorded over the supplemental motor area and contralateral motor cortex appearing 1 to 1.5 seconds prior to the movement. The Bereitschafts potential does not appear with other movements including externally triggered voluntary movements. In some cases learned voluntary motor skills are incorporated within in the repertoire of the movement

disorders such as camouflaging choeric movements or tics by immediately following them with voluntary executed movements so called parakinesias.

Semivoluntary or unvoluntary movements are induced by an inner sensory stimulus (Eg; need to stretch a body part or need to scratch an itch) or by an unwanted feeling or compulsion (Eg: compulsive touching or smelling). Many of the movements occurring as tics are as a response to various senasation (Eg; akathesia and the restless leg syndrome) can be considered unvoluntary because the movemets are usually the result of an action to nullify an unwanted , unpleasant sensation. Unvoluntary movements are suppressible.

Involuntary movements are often non suppressible (Eg most tremors and myoclonus) but some can be partially suppressible (Eg some tremors., chorea, dystonia, stereotypies and some tics).

STEPWISE APPROACH TO MOVEMENT DISORDERS

Step 1: Rhythmic Versus Arrhythmic

Rhythmic

1. Tremor
   a. Resting
   b. Posturing
   c. Action
   d. Intention
2. Dystonic tremor *
3. Dystonic myorhythmia*
4. Myoclonus,segmental*
5. Epilepsia partialis continua
6. Myoclonus,oscillatory
7. Moving toes/ fingers
8. Myorhythmia*
9. Periodic movements in sleep
10. Tardive dyskinesia(tardive stereotypy)*

Arrhythmic

1. Akathitic movements
2. Athetosis
3. Ballism.
4. Chorea
5. Hemifacial spasm
6. Hyperekplexia
7. Arrhthmic myoclonus
8. Stereotypy
9. Tics
10. Dystonia*

*Dystonias often, but not always, has repetitive movements, which were coined as myorhythmia by Herz and now labeled as dystonic tremor and patterned movements. Today , myorhythmia refers to the slow , rhythmic movements, most classically seen in Whipple disease. Segmental myoclonus is typically rhythmical, whereas other forms of myoclonus are arrhythmic. Stereotypies can occur at irregular intervals, and these are in the right hand column above. In contrast, classical tardive dyskinesia movements are continuous, and these stereotypies are placed in the left hand column.

Step 2: Sustained Versus Non Sustained

Sustained

1. Rigidity
2. Dystonia
3. Oculogyric crisis
4. Paroxysmal dystonia
5. Dystonia tics
6. Sandifer syndrome
7. Stiff person syndrome
8. Congenital torticollis
9. Orthopedic torticollis

Non Sustained

1. All others

Step 3 : Paroxysmal Versus Continual Versus Continuous

Paroxysmal

1. Tics
2. PKD
3. PNKD
4. PED
5. Paroxysmal ataxia
6. Paroxysmal tremor
7. Hypnogenic dystonia
8. Stereotypies
9. Akathitic movements
10. Jumpy stumps
11. Moving toes
12. Myorrhythmia

Continual

1. Ballism
2. Chorea
3. Dystonic movements
4. Myoclonus,arrhythmic
5. Some stereotypies
6. Akathitic moaning

Continuous

1. Abdominal dyskinesias
2. Athetosis

3. Tremors
4. Dystonic postures
5. Minipolymyoclonus
6. Myoclonus, rhythmic
7. Tardive stereotypy
8. Tic status
9. Myokymia

- Continual means over and over again; Continuous means without stopping or unbroken.

Step 4: Present While Asleep Or Awake

Appears during sleep and disappears when awakened

1. Hypogenic dyskinesias
2. Periodic movements in sleep

Persists during sleep

1. Secondary palatal myoclonus
2. Ocular myoclonus
3. Ocu;ofacialmasticatory myorhythmia
4. Moving toes
5. Myokymia
6. Neuromyotonia(Isaacs syndrome)

Diminishes during sleep

1. All others

Step 5: Present At Rest Or With Action

At rest only (disappears with action)

1. Akathitic movements
2. Paradoxic dystonia*
3. Resting tremor
4. Restless legs
5. Orthostatic tremor(only on standing)

With action only

1. Ataxia
2. Action dystonia
3. Action myoclonus
4. Orthostatic tremor*
5. Tremor: postural,action,intention
6. Task- specific tremor
7. Task- specific dystonia

At rest and continues with action

1. Abdominal dyskinesias
2. Athetosis
3. Ballism
4. Chorea
5. Dystonia*
6. Jumpy stumps
7. Minipolymyoclonus
8. Moving toes/ fingers
9. Myoclonus*
10. Myokymia
11. Pseudodystonias*
12. Tics

*Paradoxical dystonias refers to dystonias refers to dystonias that is present only at rest disappears with action; orthostatic tremor is tremor of the thighs and legs (spreading to the trunk) that occurs only on prolonged standing and disappears with walking or sitting ; most dystonias and myoclonus that are present at rest are also present and often worse with action as well; pseudodystonias refer to neuromyotonia and other causes of stiff musclaes or postures that are not due to dystonia (common are orthopredic deformities and pain).

Step 6: Patterned and Non Patterned Movements

Patterned (i.e., same muscle groups)

1. Abdominal dyskinesias
2. Dystonias
3. Hemifacial spasm
4. Moving toes/fingers
5. Segmental myoclonus
6. Myorhythmia,Myokymia
7. Tardive stereotypy
8. Tremor

Non patterned

1. All others

Step 7: Combination of Varieties of Movements

1. Psychogenic movement disorders
2. Tardive syndromes
3. Neuroacanthocytosis
4. Wilson disease
5. Huntington disease
6. DRPLA
7. Dystonia*

*Patients with dystonia have additional dyskinesias that are part of the spectrum of classical torsion dystonia. These include tremor,myoclonus and choreic – like movements. Dystonia – plus syndromes can have features of parkinsonism or myoclonus in addition to dystonia.

Step 8: Speed (Fast Versus Slow)

Fastest →

1. Minipolymyoclonus
2. Myoclonus
3. Hyperekplexia

4. Hemifacial spasm

Intermediate →

1. Chorea
2. Ballism
3. Jumpy stumps
4. Tremors
5. Tardive steretypy

Slowest

1. Athetosis
2. Moving toes/fingers
3. Myorhythmia
4. Akathitic movements

Step 9: Amplitude

Ballistic

1. Ballism

Not Ballistic

1. Chorea and all others

Very Small

1. Minipolymyoclonus

Step 10: Force

Powerful

1. Stiff person
2. Jumpy stumps

Intermediate

1. Dystonia

Easy to overcome

1. All others

Step 11: Suppresibility

Suppressible

1. Stereotypies>tics,akathitic movements>chorea>ballism>dystonia>tremor>moving toes

Not suppressible

1. Hemifacial spasm
2. Minipolymyoclonus
3. Myoclonus
4. Hyperekplexia
5. Myorhythmia
6. Moving toes

Step 12: Vocalizations

1. Vocal tics: simple or complex
2. Akathisia: moaning
3. Huntington disease
4. Neuroacanthocytosis
5. Cranial dystonia

Step 13: Presence of Self Mutilation

1. Lesch- Nyhan Syndrome
2. Neuroacanthocytosis
3. Tourette Syndrome
4. Psychogenic movement disorders

Step 14: With Complex Movements

1. Tics
2. Akathitic movements
3. Compulsions
4. Stereotypies
5. Psychogenic movements

Note: Each of the above can have simple movements as well.

Step 15: Presence Of Sensory Component

1. Akathisia
2. Moving toes,painful legs
3. Restless legs
4. Tics

Step 16: Presence Of Ocular Movements

1. Tics
2. Oculogyric crises
3. Opsoclonus
4. Ocular myoclonus
5. Ocular myorhythmia
6. Ocular dysmetria
7. Nystagmus

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