Chapter 8 – Gait and Movement Disorders

William G. Ondo, MD
Rosabel Young, MD

Definition
The term “movement disorder” refers to the group of central nervous system diseases in which the control of movement is altered with relative preservation of strength, muscle bulk, and mechanical range of motion. Instead, there are changes in the patient’s muscle tone, rapidity and smoothness of voluntary movements, or movements may occur involuntarily. Movement disorders are grossly segregated into hyperkinetic movements (tremor, dystonia, chorea etc.) and hypokinetic movements (Parkinsonian conditions).

Epidemology and Scope of the Problem as a Health Care Issue
Movement disorders probably affect about 20 percent of the population. Restless legs syndrome accounts for half of this. Essential tremor is estimated to affect up to 5–10 percent of the population. Parkinson’s disease in persons over the age of fifty is approximately 1 percent. However, some studies indicate that up to 10 percent of the population over age 60 have early symptoms not yet diagnosed as Parkinson’s disease. Although early studies suggested that Parkinson’s disease was more common in Caucasians, some prevalence studies comparing other racial groups and Caucasians in the same geographic areas have demonstrated approximately equal numbers. Other movement disorders may also be more common than previously thought. The various dystonias, especially spasmodic torticollis, may affect up to 0.4 percent population.

Although most movement disorders are not life threatening, they are certainly a threat to the patient’s quality of life. The impact can be enormous, with loss of employment, inability to drive an automobile, and impairment in activities of daily living including personal hygiene. Most do not spontaneously remit so they become lifelong issues. In addition, physicians and patients often face a challenge in obtaining insurance coverage for treatment of these conditions, since many treatment modalities, both pharmacologic and surgical, are relatively new.

Clinical Symptoms And Signs
The motor control system is the part of the nervous system that integrates sensory input and organizes and directs motor output. The structures involved include proprioceptive sensory receptors in the muscles, the spinal cord, brainstem, cerebellum, thalamus, basal ganglia, and cerebral cortex (Figure 8-1).
Figure 8-1: Sagittal view of the brain and brain stem showing the location of the basal ganglia. From these structures there are numerous projections to and from the cerebral cortex as well as descending tracts into the spinal cord. The neurotransmitters involved include: Glutamate, GABA (y-aminobutyric acid), Acetylcholine, Dopamine, Norepinephrine, Serotonin, Somatostatin, Enkephalin, Dynorphin, Cholecystokinin, Neurtensin, neuropeptide Y, and possibly others.

The approach to the patient with a movement disorder is to first determine which aspects of motor control are clinically affected, and then to combine the cluster of symptoms and signs found into a specific diagnosis and etiology whenever such can be identified. Treatment is directed toward the individual symptoms as well as the underlying etiology.

Most movement disorders begin insidiously. A co-worker, spouse, or even child of the patient may notice the problem before the patient does. Patients may complain of “weakness” or “stiffness” in their muscles, or they may have noticed involuntary movements, such as tremors, twitches, or gross movements of their head or extremities. Symptoms may be noticed as they try to perform their routine activities, or may be present only at certain times, such as when walking, turning the head, or handwriting. Thus, in addition to testing strength, examination of the motor system should include testing of tone, kinesis, posture, observation for any spontaneous involuntary movements, and evaluation of coordination and gait with various provocative maneuvers. If the patient tells you that the abnormal movement occurs only with a certain activity or position, such as when writing, always try to reproduce it in your office (Table 8-1).
Table 8-1: Clinical signs of abnormal motor control

<table>
<thead>
<tr>
<th>Movement</th>
<th>Speed/Phenotype</th>
<th>Location</th>
<th>Hallmark Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rest Tremor</td>
<td>4–6 Hz</td>
<td>Arms&gt;legs</td>
<td>Parkinson’s disease</td>
</tr>
<tr>
<td>Postural Tremor</td>
<td>6–12 Hz</td>
<td>Hands</td>
<td>Essential Tremor</td>
</tr>
<tr>
<td>Intention Tremor</td>
<td>2–5 Hz</td>
<td>Arms/legs</td>
<td>Cerebellar lesions</td>
</tr>
<tr>
<td>Dystonia</td>
<td>Tonic Patterned</td>
<td>Any part</td>
<td>Torticollis</td>
</tr>
<tr>
<td>Stereotype</td>
<td>Slow, semi- rhythmic</td>
<td>Any part</td>
<td>Tardive dyskinesia</td>
</tr>
<tr>
<td>Athetosis</td>
<td>Slow, irregular &quot;writhing&quot;</td>
<td>Proximal limbs</td>
<td>Brain lesion</td>
</tr>
<tr>
<td>Chorea</td>
<td>Fast, irregular</td>
<td>Any part</td>
<td>Huntington’s disease</td>
</tr>
<tr>
<td>Myoclonus</td>
<td>Fast, Simple</td>
<td>Any part</td>
<td>Juvenile myoclonic epilepsy</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Cerebral hypoxia</td>
</tr>
<tr>
<td>Tics</td>
<td>Fast, Patterned</td>
<td>Face, neck, shoulders</td>
<td>Tourette’s syndrome</td>
</tr>
</tbody>
</table>

**Muscle tone** refers to the resting activity of agonist and antagonist muscles. Normally, no resistance should be felt when passively flexing and extending the patient’s arm. When the legs are tested by lifting and letting go (in the supine position), the foot should drag on the bed and the whole leg should drop when released. Rigidity and spasticity are two types of increased tone indicating deficits in the basal ganglia or corticospinal tracts, respectively. Cogwheel rigidity refers to increased tone with a ratchety feel when passively moving a limb, as seen in Parkinson’s disease. Tone can vary with certain extrinsic factors. Pain, for example, will temporarily increase tone and even deep tendon reflexes, but this is usually to an equal degree in all four limbs. The inability to relax may artificially increase apparent tone. Muscle relaxants and antispasmodics, such as diazepam, carisoprodol, baclofen, and related agents will decrease tone symmetrically.

**Dystonia**, by definition, means a sustained abnormality ("dys-") in muscle tone ("-tonia"). Dystonia can be thought of as a sustained contraction of a group of muscles that produces altered posture of the head, neck, trunk, or limbs. There should be a consistent pattern. Usually this is because of an imbalance of resting tone between agonist and antagonist muscle groups in the same limb, or in the neck. Patients may notice pain before the dystonia becomes visibly apparent. Involuntary movements, especially tremors, are sometimes associated with dystonias. Usually volitional movement initiates or augments the dystonia, although fixed dystonias at rest also occur, usually associated with brain injury. As the dystonia worsens and begins to interfere with daily activities, patients may develop certain compensatory maneuvers. For example, a patient may change the way they hold a pen to overcome hand dystonia known as writer’s cramp.

**Tremor** is the most common presenting symptom in patients with movement disorders. It is a rhythmic oscillation around one or more focal points, usually a joint. Tremor may be seen in the head, face, especially the chin, one or both arms or legs. Tremor occurs when agonist and antagonist muscles supplying the same limb contract involuntarily. They may alternate or actually be synchronous. Tremors vary in frequency, anatomy, quality, and precipitating actions. With loss of neuronal function in the basal ganglia, cerebellum, or certain other structures involved in motor control, this balance is lost, and tremor occurs (Figure 8-2).
Tremor should be observed at rest (muscle not in use), with the arms held forward, and with directional movements, such as the finger-to-nose task. The three main types of tremor and their treatment will be discussed later in this chapter.

There are various types of tremor, which point the neurologist to particular anatomical structures within the central nervous system. Because of the complex neurochemical pathways subserving the basal ganglia, localization is important in determining appropriate treatment (Table 8-3).

**Table 8-3: Classification and differential diagnosis of tremor**

<table>
<thead>
<tr>
<th>Rest Tremors</th>
<th>Action Tremors</th>
</tr>
</thead>
<tbody>
<tr>
<td>Parkinson’s disease</td>
<td>Postural</td>
</tr>
<tr>
<td>Other Parkinsonian syndromes</td>
<td>Essential tremor (familial or sporadic)</td>
</tr>
<tr>
<td>Midbrain (rubral) tremor: rest&lt;postural/action</td>
<td>Task specific tremors (i.e. isolated writing tremor)</td>
</tr>
<tr>
<td>Wilson Disease</td>
<td>Orthostatic tremor</td>
</tr>
<tr>
<td>Essential Tremor-only if severe: rest&lt;postural/action</td>
<td>Physiological tremors</td>
</tr>
<tr>
<td></td>
<td>• Endocrine: Hypoglycemia, thyrotoxicosis, pheochromocytoma adrenocorticosteroid stress, fatigue, anxiety</td>
</tr>
<tr>
<td></td>
<td>• Drugs: beta, agonists, dopamine agonists, amphetamines, lithium, SSRI neuroleptics, theophylline, caffeine, valproic acid</td>
</tr>
<tr>
<td></td>
<td>• Toxins: alcohol withdrawal, mercury (“hatters shakes”), lead, arsenic, others</td>
</tr>
<tr>
<td></td>
<td>Cerebellar tremor (postural&lt;action)</td>
</tr>
<tr>
<td></td>
<td>• Focal cerebellar of brainstem lesions due to multiple sclerosis, trauma, tumor</td>
</tr>
<tr>
<td></td>
<td>• Drugs and Toxins: Chronic Dilantin®, mercury, others</td>
</tr>
<tr>
<td></td>
<td>Asterixis, myoclonus, epilepsy partialis continua, myorrhythmia, others</td>
</tr>
</tbody>
</table>
Myoclonus is a rapid, very brief (< .25 seconds), simple contraction. It may affect any body part, may be rhythmic or non-rhythmic. Although myoclonus can be seen with several epilepsy syndromes, metabolic derangements (hepatic or renal) and brain ischemia are the most common etiologies.

Asterixis, also called negative myoclonus, refers to sudden loss of tone while attempting to maintain a limb in a certain position. Typically, it appears as a “flapping” of the hands when the patient holds the arms out with palms extended as if halting traffic. Asterixis is a classic sign of hepatic encephalopathy, hence the term “liver flap.”

Kinesis simple means movement. Bradykinesia means slowed (“brady-”) movement and hypokinesia means small movements. These are characteristic of Parkinson’s disease. Observe the patient performing various tasks, such as opening and closing the fist or finger-tapping rapidly. PD patients have slowness, and more specifically smallness to these. Often, the amplitude will start normally then decrement. Tapping may stop altogether, then restart at a larger amplitude only to decrement again. This is much more specific for PD compared to general slowness, which could be seen in many disorders. Other PD symptoms and signs such as the “masked facies” (hypomimia), micrographia are consequences of hypokinesia. Retropulsion on pull testing (backward movements caused by the inability to move the feet fast or big enough) is also a sign of hypokinesia.

Dyskinesia technically means any disorder of kinesis. The term is used to describe multiple phenotypes, usually hyperkinetic. It is most commonly used in association with tardive syndromes (below), and levodopa (Sinemet®) when used for PD.

Chorea refers to involuntary movements, which are rapid and unpredictable. Each movement involves one part of the body at a time, but “skips” from one part to another in seemingly random fashion.

Athetosis is a pattern of dyskinesia in which the random involuntary movements occur slowly, seeming to “flow” rather than “skip” to different parts of the body. Athetosis and chorea may actually represent different severities of the same pathophysiology, but have traditionally been individually defined.

Tics are another type of involuntary movements which differ from chorea in that they involve smaller groups of muscles, are more “jerky” in quality, and tend to be stereotyped, recurring with the same or a very similar pattern in the same muscles. Tics are often partially suppressible, and may involve an urge to tic, which grows if the tics are suppressed. They are also suggestible and increase when they are discussed. Tics are most commonly seen in the face but can occur anywhere.

Ataxia means any breakdown of smooth, coordinated movement. It is often segregated into limb ataxia and gait ataxia. When examining the limbs look for “past pointing” on finger to nose testing (the subject misses your finger), breakdown of rapid alternating movements (alternately tapping the plantar and dorsal surface of the hand against the leg is arrhythmic. The inability to quickly stop movements “overshoot” is also characteristic of limb ataxia. When examining for gait ataxia, observe the patient’s posture, the speed of the swing phase, the stride length (normal is 24”–26” for women, 30” for men). Also note the looseness and symmetry of the arm swing and watch for the foot pivoting when the patient makes turns. A true cerebellar ataxic gait is wide-based and the patient has difficulty walking in tandem (“tightrope walk”). The arms are
often abducted to improve balance. The hallmark of sensory ataxia (a pseudo--
ataxia caused by impaired sensory input, usually from a neuropathy) is a positive
Romberg; gait improves if the patient looks at his feet. Gait impairment due to
basal ganglia dysfunction is manifested by slowing, shuffling and poor postural
reflexes, most easily observed when the patient makes turns.

Diagnostic Evaluation

The focus of the diagnostic work-up will be guided by the information acquired
from the history and physical examination (Table 8-2A and 8-2B). All differential
diagnosis are mostly based on the neurologic examination.

The Neurologic Examination

Table 8-2A—Additional clues from the history

<table>
<thead>
<tr>
<th>Question</th>
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<tbody>
<tr>
<td>Was the onset acute, subacute, or chronic?</td>
</tr>
<tr>
<td>Was the onset related to another illness (cardiac arrest, an auto accident or fall)?</td>
</tr>
<tr>
<td>Are there other “unrelated” symptoms or illnesses (e.g., liver disease, headaches, dysphagia, visual, speech, or memory disturbance)?</td>
</tr>
<tr>
<td>If there is syncope or other alteration of consciousness, or if the movement disorder is episodic, the patient should be evaluated for a seizure disorder.</td>
</tr>
<tr>
<td>Was the onset associated with ingestion of a drug (accidentally or prescribed)?</td>
</tr>
<tr>
<td>Has the patient identified aggravating or relieving factors, such as caffeine to worsen or a sip of alcohol to relieve tremor?</td>
</tr>
<tr>
<td>Is motor function better or worse with the time of day?</td>
</tr>
<tr>
<td>Does it disappear or worsen during sleep (ask the spouse)?</td>
</tr>
<tr>
<td>How does the movement problem interfere with daily life (walking, driving, eating, talking, sports, and public appearances)?</td>
</tr>
</tbody>
</table>

Table 8-2B—Additional clues from the examination

<table>
<thead>
<tr>
<th>Question</th>
</tr>
</thead>
<tbody>
<tr>
<td>Seborrhea and abnormalities in sweating are associated with Parkinson’s disease and other conditions of the basal ganglia.</td>
</tr>
<tr>
<td>Orthostatic hypotension and other cardiovascular symptoms may point to a problem with autonomic nervous system regulation, which also lies in the domain of the basal ganglia.</td>
</tr>
<tr>
<td>Psychiatric disease may be a clue to drugs as the etiology, even prescribed drugs, such as the neuroleptics or antiemetics.</td>
</tr>
<tr>
<td>Dementia is part of some movement disorders, such as the late stages of Parkinson’s disease.</td>
</tr>
<tr>
<td>Hepatic insufficiency may be a clue to certain entities, some common (alcoholism) others less so (Wilson disease).</td>
</tr>
</tbody>
</table>

Serological and other laboratory examinations depend entirely on the
phenotype. Recommended serologies range from none (classic PD phenotype) to many (ataxia and chorea have large differentials). A large number of metabolic abnormalities may result in movement disorders. Screening for hepatic failure, uremia, electrolyte abnormalities, including calcium and parathyroid, thyroid testing, inflammatory/rheumatological screening, and metals (serum and urine copper, heavy metals, may be considered.

Neurology consultation should be sought when no etiology is found to explain
the patient’s symptoms after routine office and laboratory evaluations have been performed. Imaging studies such as CT and MRI may reveal structural lesions such as strokes, tumors, or severe atrophy, but since the basis of most
movement disorders is biochemical rather than structural, these are helpful only
in a minority of cases. For example, brainstem and cerebellar atrophy is seen in
olivopontocerebellar atrophy.
Neurophysiologic studies should be performed whenever there is pain, numbness, weakness, or paresthesias. Electromyography (EMG) is valuable in identifying patterns of muscle hyper- and hypo- activity in various types of movement disorders, especially the dystonias, however this would have to be specifically ordered as standard EMG usually concentrates on neuropathy and myopathy. Nerve conduction studies (NCV) are indicated in the evaluation of movement disorders associated with sensory disturbances.

Electroencephalography (EEG) should be performed if the abnormal movements are paroxysmal (sudden onset with brief duration of seconds to minutes) or if there is associated alteration of consciousness or behavior. As mentioned above, myoclonus is often part of an epileptic syndrome that may include absence and generalized tonic-clonic seizures in later life. Myoclonus is also associated with metabolic, toxic, and hypoxic encephalopathies. Therefore it is imperative to perform electroencephalography in any patient with myoclonus.

**Disease Overviews**

**Essential tremor** is one of the most common movement disorders. Prevalence studies indicate that it is about 5–20 times more common than Parkinson’s disease. Onset is usually in early adulthood, although it can occur at any time. Progression is variable but as a rule, the amplitude increases and the frequency decreases over years. High amplitude correlates with disability so patients often have tremor for many years before presenting to a physician. A positive family history can frequently be elicited. Amelioration by alcoholic beverages is another common feature. The hands are almost always involved but any part may tremor.

The most commonly used medications for ET include propranolol and primidone. Propranolol and nadolol are probably the most effective beta blockers for ET, and can be used on an “as needed” basis. Their effect is dose-dependent and they help most cases to some extent. Primidone is less dose-dependent, cannot be used as needed, and usually either helps significantly, or does not help at all. The usual dose should begin at 25 mg to minimize side effects. Many other medications may help ET: topiramate, pregabalin, gabapentin, zonisamide, benzodiazepines. Botulinum toxin and brain surgeries can also be used. (See below)

**Orthostatic tremor** is a very interesting tremor disorder that does not present with tremor. Rather, patients complain of balance difficulties specifically while standing. The subjective balance immediately improves with walking. Although patients report a dramatic feeling of decreased balance and exhibit distress while standing, they do not actually fall. Upon close examination while standing, a very fine, rapid (14 Hz) tremor can be palpated, and sometimes seen, in the calves. This stops when walking. Numerous medications may improve orthostatic tremor (clonazepam, gabapentin, topiramate, phenobarbital, and dopaminergics) but the efficacy often lessens over time.

**Physiologic tremor** is the term used to describe a tremor that is enhanced by certain physiologic states, such as hyperthyroidism. All people have some tremor, the amplitude of which can be enhanced in a variety of situations. Physiologic tremor is usually low amplitude and high frequency and seen with posture or during activity of the upper limbs, rather than at rest. Treatment should be aimed first at the underlying etiology (Table 8-4) although propranolol
is very effective.

Table 8-4. Drugs or toxins that may potentiate physiologic tremor

<table>
<thead>
<tr>
<th>Beta-adrenergic agonists</th>
<th>Psychiatric drugs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Terbutaline</td>
<td>Lithium</td>
</tr>
<tr>
<td>Metaproterenol</td>
<td>Neuroleptics</td>
</tr>
<tr>
<td>Isoetharine</td>
<td>SSRI</td>
</tr>
<tr>
<td>Epinephrine</td>
<td></td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th>Stimulants</th>
<th>Heavy metals</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amphetamine</td>
<td>Mercury</td>
</tr>
<tr>
<td>methylyphenidate</td>
<td>Lead</td>
</tr>
<tr>
<td>midodrine</td>
<td>Arsenic</td>
</tr>
<tr>
<td>cocaine</td>
<td>Bismuth</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Anticonvulsants</th>
<th>Methylxanthines</th>
</tr>
</thead>
<tbody>
<tr>
<td>Valproate sodium</td>
<td>Caffeine</td>
</tr>
<tr>
<td></td>
<td>Theophylline</td>
</tr>
</tbody>
</table>

Cerebellar outflow tremor, aka intention tremor, is a tremor that worsens with directed movement. Closed head trauma and alcoholic cerebellar degeneration are common causes in adults. Numerous hereditary ataxias, multiple sclerosis, Wilson disease, and posterior fossa tumors may also present this way. Certain drugs and toxins affecting these pathways, such as phenytoin and carbamazepine, may cause intention tremor.

Idiopathic Parkinson’s disease (PD) is the most common degenerative movement disorder. Men are affected somewhat more commonly than women, and rural areas have a somewhat high prevalence of disease. PD is manifested, and clinically diagnosed, by rigidity, bradykinesia, postural instability, and tremor. Motor symptom onset is usually unilateral, and that side will always be worse. The most common symptom that leads the patient to seek medical attention is tremor. However, tremor is not necessarily the first symptom to appear, nor is it the most disabling symptom. Patients may not suspect that bradykinesia and rigidity are early symptoms. In fact non-motor symptoms such as REM behavioral disorder (acting out dreams), impaired olfaction, and constipation may precede the motor symptoms of PD by more than a decade. This suggests that the neurodegeneration occurs for many years prior to the motor symptom onset. Multiple genetic causes of PD have been identified, although most subjects do not have any family history. PD is in fact multiple different diseases that present with similar symptoms caused by dopaminergic cell loss.

Treatment

The treatment of PD is complex. Many non-motor aspects (depression, psychosis, urinary urgency, constipation, sleep disorders, dementia) often need to be addressed pharmacologically. Non-pharmacologic treatments, especially specific physical therapies (visual cuing, rhythmic training etc.) also aid the movement problems. The following summary included only pharmacological treatments for the cardinal motor symptoms.

1. Levodopa (combined with carbidopa as Sinemet®) is the most potent treatment for Parkinson disease. (Figure 8-3) Unfortunately, the short dose response of the drug requires frequent dosing as the disease progresses, at intervals as short as every 1–1/2 to 2 hours. Sustained-release Sinemet CR® can reduce this inconvenience, but absorption is reduced, requiring higher total daily doses. Side effects include
orthostatic hypotension, dyskinesias, hallucinations, and nausea. Dyskinesia (involuntary excessive chorea type movements) develops over time and is the greatest limitation with levodopa, and greatest argument for alternative dopaminergic therapies.

2. **Dopamine agonists**, such as bromocriptine (Parlodel®) and pergolide (Permax®), pramipexole (Mirapex), ropinirole (Requip®), rotigotine (Neupro®), and apomorphine (Apokyn®) were initially introduced to help control fluctuations in movement control that patients with Parkinson’s disease often develop on levodopa. It is often recommended that therapy begin with these drugs as they do clearly delay the onset of dyskinesia. They are somewhat less potent than levodopa and tend to have more hallucinations, peripheral edema, and sedation, and impulsivity (increased spending, gambling, sex drive) than levodopa. They also require more titration.

Monoamine oxidase (MAO) is one of the enzymes that breaks down dopamine. MAO inhibitors (selegiline, rasagiline) increases brain concentrations of dopamine and should lead to improvement of Parkinson’s disease symptoms. In practice, however, the efficacy is modest, although side effects are also mild.

3. **Anticholinergics** (Artane®, Cogentin®) may be prescribed as adjunctive therapy to levodopa-carbidopa, and may be helpful early in the course of Parkinson disease if tremor is significant. Unfortunately, elderly patients are less tolerant to these agents because of side effects, such as cognitive impairment, dry mouth, and urinary retention. In recent years, the use of anticholinergic medications in the treatment of Parkinson disease has waned, but these drugs are very addictive and difficult to wean off.

Although the availability of a growing armamentarium of pharmacologic agents for Parkinson disease has changed the outlook for many patients, the degree of functional impairment may still be prohibitive. Initially, early onset Parkinson’s disease patients, who often became disabled by the disease in their prime employable years, were the prime candidates for surgery, but advances in techniques have occurred so rapidly that surgery is considered an option for all ages as long as the patient meets the medical screening criteria.

4. Before L-dopa became available, **thalamotomy** was one of the few treatment options available for persons with Parkinson’s disease and certain tremors (Markham et al, 1966). Unfortunately, cumulative experience with thalamotomy showed that this procedure is beneficial only for a few months after it is performed. Moreover, thalamotomy appears to effectively decrease contralateral tremor, but bradykinesia usually remains, and rigidity improves variably.

Collaboration among neurologists, neurosurgeons, neurophysiologists and neuroradiologists led to the development of a thalamic stimulation technique that could reproduce the benefits seen early in response to thalamotomy. The ventral intermediate (VIM) nucleus of the thalamus was found to be the locus of neurons that appear to be responsible for tremor; thus, it was predicted that the VIM would be a useful lesioning target for the reduction of tremor. High-frequency stimulation at rates over 100 Hz reproduces the same physiologic effect as lesioning. Known
as “DBS” (for Deep Brain Stimulation), the procedure involves the insertion of an electrode wire that is inserted into the VIM under electrophysiologic guidance. The other end of the wire is connected to a pulse generator, which resembles a cardiac pacemaker in size and shape. This pulse generator unit is implanted subcutaneously at the pectoral area. The patient activates this unit by passing a small (about 2 inches) hand-held magnet over the chest. Within 30 seconds to 5 minutes, the tremor resolves on the contralateral side, and the patient can go about his usual activities until he chooses to pass the magnet over the unit again to turn off the stimulation. Although bradykinesia and rigidity usually remain, for patients whose tremor is the primary disabling feature the results are dramatic.

5. **Pallidotomy**, a procedure in which the medial portion of the globus pallidus is lesioned permanently, gained popularity among the general public after several anecdotal reports of success appeared in the news media and the Internet in the mid-1990s. There is greater morbidity with bilateral procedures, with complication rates greater than 15 percent. If the procedure is performed under careful electrophysiologic monitoring and restricted to one side, preferably the nondominant, results are more favorable, with reduction in dyskinesia, but lesser effect on rigidity, bradykinesia and tremor. Pallidotomy is rarely performed now that the technique of Deep Brain Stimulation has been standardized and more widely accepted.

6. More recently, the **subthalamic nucleus** has emerged as the DBS surgical treatment of choice for Parkinson’s disease. This DBS approach was first reported in Grenoble, France, and Pamplona, Spain, and has undergone successful clinical trials in Canada and the United States as well. Subthalamic DBS ameliorates Parkinsonian features other than tremor, including bradykinesia, rigidity and even dyskinesias and motor fluctuations (Limousin et al, 1995; Obeso, 6/2000). It reduces dyskinesia and allows reduced medication doses. The ideal candidate for a STN DBS is relatively young, cognitively intact, and has a dramatic change with levodopa. In fact most surgical centers will not implant a STN DBS unless the patient has marked dyskinesia. Symptoms that do not respond to levodopa i.e. voice, balance, psychiatric, autonomic, do not respond to STN DBS.

**Multiple Systems Atrophy** is pathologically related to PD and may present with similar symptoms. Later, however patients may develop marked autonomic abnormalities (orthostatic hypotension, incontinence, inspiratory strider, etc.) and/or true ataxic features. The prognosis is much worse as patients do not respond as well to medications.

**Progressive Supranuclear Palsy** (PSP) and Cortical Basal Degeneration (CBD) are pathologically related conditions that present with parkinsonism. PSP is manifest by loss of upgaze, marked balance problems, and early bulbar symptoms. CBD presents very unilaterally (often isolated for three years prior to involvement of the other side). Patients present with apraxia (an inability to use their hand or leg, despite normal strength. They may also have PSP symptoms. The prognosis for both is poor and the medications do not respond to treatment.
Dystonias

The dystonias are usually classified by anatomical involvement: “generalized,” “hemi-,” “segmental,” or “focal.” Most generalized dystonias are genetically inherited, or at least idiopathic, and present in childhood. Hemidystonia usually results from a brain lesion (stroke etc.). Adults usually have focal dystonias of the face, neck, and limbs. When multiple contiguous areas are involved, the term segmental is applied.

- **Idiopathic Torsion Dystonia**, formerly termed, “Dystonia Musculorum Deformans,” is a generalized dystonia that usually begins in childhood with twisting of one foot while walking. Dystonic posturing may be precipitated by a specific action, which the patient can often reproduce on request. Other actions that use the same muscles, such as walking backward, often do not exhibit dystonia. The hereditary forms may show an autosomal dominant, recessive, or X-linked pattern. Only one (DYT-1) is commercially available for testing.

- **Blepharospasm** is involuntary bilateral contraction of the peri-ocular muscles, sometimes with sustained bilateral eye closure. The eyes are not actually involved. This may be so severe as to impede the patient’s functioning and pose great risk, particularly when driving. It may spread to other parts of the face and be called “cranial dystonia.” Interestingly blepharospasm improves while patients are talking. It often worsens with lights, especially headlights, or other objects coming at the patient i.e. walking in a crowd. There is usually a gritty sensory component, and many patients are misdiagnosed with “dry eyes.” It is usually idiopathic but may be seen with Parkinsonian disorders.

- **Spasmodic torticollis** (cervical dystonia) is a common focal dystonia of the neck muscles that results in abnormal neck and head posturing. This could result in rotation (torticollis); tilt (laterocollis); flexion; or extension (antero- or retrocollis). As with all dystonias, the pattern, although not necessarily intensity, should be stable. A head tremor very much resembling an essential tremor is often seen in patients with cervical dystonia.

- **Writer’s cramp and other task specific dystonias** occur in the setting of repetitive motor tasks. Writing is the most common task specific dystonia. It may begin with an involuntary movement (flexion more often than extension) of the hand precipitated by a single letter. Eventually it will occur with any word writing and later with any movement involving a writing instrument (drawing a straight line). Patients may notice the actual movement or just report “tightness” or “muscle fatigue.” Writing is the classic precipitant of this form of dystonia, but many other activities may produce it as well. Musicians commonly develop task specific dystonias, often ending their careers. Typing and sports i.e., “golf yipps” are other examples.

**Dystonia treatment** depends on the severity and anatomy of the muscle involvement. Botulinum toxin injections are widely considered to be the most effective treatment for all focal dystonias. These compounds (BOTOX, Dysport, Myobloc) eloquently inhibit the release of acetylcholine from the nerve into the neuromuscular junction. Without this trigger, the muscle will not contract, and begins to atrophy. The toxin is extremely potent and must be injected into the muscle that is targeted. Therefore a good knowledge of muscle activity and
function is essential for proper injection technique. There are many debated technical issues to identify the appropriate muscles and optimize placement of the toxin into those muscles. In general, activity lessens for 3–5 months, then gradually returns. Repeat injections are then required. The greatest problem with botulinum toxins are the high cost, especially when large muscles require injections. Numerous muscle relaxant medications can be used for dystonia but their effect is very inconsistent and they have numerous side effects, especially sedation. In general muscle relaxants have relatively greater utility in generalized or hemi-dystonia, when botulinum toxin becomes impractical. Finally baclofen pumps and deep brain stimulation of the globus pallidus internus are also commonly used for generalized and segmental dystonias.

Restless legs syndrome (RLS) is a very common condition affecting women more than men. It manifests as an urge to move the legs, which improves with movement, worsens with inactivity, and worsens at night. Subjects with RLS usually have periodic leg movements of sleep (PLMS) seen on sleep studies, but this is not part of the diagnosis, which is made exclusively on history. RLS pathology shows reduced iron in the brain, even body stores of iron, and iron testing, are normal. Several medical conditions including systemic iron deficiency, uremia, pregnancy, and possibly neuropathy are associated with RLS. Most other patients have a family history, and multiple genetic loci have been found.

RLS usually responds dramatically to low doses of dopamine agonists. The dose should be administered 1–2 hours before the onset of symptoms. Other treatments include levodopa, gabapentin, and opioids.

Hemifacial spasm (HFS) refers to rapid synchronous, involuntary unilateral contractions of the facial muscles, which may range in severity from simple twitching of the corner of one eyelid, to sustained and even painful contraction of one entire side of the face. All involved muscles are innervated by the facial nerve (CN VII). HFS is usually caused by a compression of the facial nerve near its exit from the brainstem, usually a blood vessel. This causes a “short circuit” in the nerve and spontaneous firing. Hemifacial spasm may occur months or years following Bell’s palsy in association with synkinesis due to aberrant reinnervation.

HFS responds dramatically to low doses of botulinum toxin. Seizure medications are sometimes moderately effective. A surgical procedure where the offending blood vessel is stented away from the nerve is also usually effective.

Drug induced movement disorders

There are five main drug induced movements disorders. All are associated with the use of medicines that block dopamine receptors. These include neuroleptics, nausea and GI drugs. Although data is lacking, metoclopramide may be the most common offender.

- Tardive dyskinesia may occur while on an offending medication or only after stopping it. The phenotype is variable but most commonly involved repetitive, loose perioral and lingual muscles. The movements appear with the patient at rest and can be volitionally suppressed for a few seconds at a time. They also tend to decrease with voluntary activity requiring use of the involved muscle groups. Risk factors include a longer duration of use, older age, and female sex.

- Acute dystonic reactions occur within days of starting a dopamine
blocker. Children are most commonly effected by this dramatic dystonic extension posturing. Although it looks serious, it almost always resolves with a single dose of anti-histamine and will not usually recur. Therefore, it does not preclude the continued use of the drug.

- **Akathisia** is an intense urge to move the body. Patients will stand up, pace, and rock back and forth. In most cases it resolves upon discontinuation of the offending agent.

- **Neuroleptic malignant syndrome** is a severe life threatening condition manifest by acute rigidity, fever, and altered mental status. Muscle breakdown may result in renal failure. Treatment is supportive along with dopamine agonists, and possibly other muscle relaxants. Recovery can take months.

- **Drug induced parkinsonism** cannot be consistently differentiated from idiopathic PD on clinical grounds. It may be more symmetric and have a higher frequency tremor. Removing the offending agent results in improvement but this may take months. Up to 40 percent of patients with drug induced parkinsonism may actually have latent PD that was unmasked by the drug.

*Wilson disease* is a systemic illness caused by accumulation of copper primarily in the liver and brain. The characteristic flapping tremor of the arms is seen proximally, and best brought out with the patient’s arms in “chicken wing” position. The phenotype is actually quite broad. The well-known finding of Kayser-Fleischer rings at the circumference of the irises might not be detected without a slit-lamp examination. Lever functions may be abnormal but are not needed to suspect the diagnosis. A low serum ceruloplasmin and “reversed” serum/urine copper ratio confirm the diagnosis.

If found early, therapies that chelate copper can completely ameliorate the disease, and result in a normal life. Without treatment, the disease is usually fatal.

**Choreas**

- **Huntington’s disease** is an autosomal dominant hereditary form of chorea that is relentlessly progressive. It can be associated with other central nervous system manifestations, especially dementia, personality changes, gait disorders, and bulbar symptoms. Interesting the same disease in childhood has nearly opposite symptoms including parkinsonism and tremor. Genetic and neuroimaging studies of large families have led to the discovery of the autosomal dominant Huntington’s disease gene on chromosome 4. The question of whether to conduct testing presents an ethical dilemma. While there is no “right” decision, a team counseling approach involving the patient’s family physician, neurologist, and significant relatives, can result in peace of mind.

- The chorea can be treated with amantadine, dopamine antagonists, or tetrabenazine. Anti-depressants appear to help but there is not effective treatment for the entire disease.

- **Sydenham’s chorea**, now rare, tends to occur in children and teenagers after streptococcal infection. The exact relationship to the Strep infection is unknown, some have negative ASO titers. Although steroids have been used with some success in shortening the duration of the initial attack,
the condition usually remits spontaneously over 3–5 months and may nevertheless relapse years later.

- Numerous other rare conditions can cause chorea. Chorea is associated with pregnancy, lupus and other autoimmune disorders, several other genetic conditions, and brain ischemia.

**Tourette’s syndrome** is defined by having multiple motor tics and at least one sound tic. It is not thought to be intrinsically different from other arbitrarily defined tic disorders (multifocal motor tic disorder etc.). Often patients also have features of obsessive–compulsive disorder. The mean age of onset is seven but peak intensity is the early teens. Boys are affected more than girls. There is a strong familial tendency for both the tic and behavioral components of this condition that appears to be transmitted in autosomal dominant fashion. Although the motor and vocal tics can be willfully suppressed by the patient, this requires significant effort and produces great anxiety. The anxiety that builds up is released upon allowing the movements or vocalizations to occur.

**Psychogenic movement disorders** are relatively common and can present diagnostic difficulties. The most common is probably tremor, followed by dystonia. In general these have an acute onset, which is fairly uncommon in organic movement disorders. Psychogenic tremor tends to have variable frequencies, is distractible, and will entrain (take the same frequency when the subject is asked to volitionally mimic tremor in the other limb). Psychogenic dystonia tends to be fixed (most commonly foot plantar flexion that cannot be pushed back). The psychopathology ranges from a true conversion disorder to frank malingering. This diagnosis should be made by someone with expertise in movement disorders.

**Community resources**

Because the psychosocial impact of movement disorders is so great, a number of organizations have been created to provide support and medical information in lay terms, largely through the efforts of patients and families. Online question-and-answer “newsgroups” have also proliferated through the Internet in recent years. These sources can answer questions about legal issues, such as driving, and can offer help to the caregivers.

**Patient support groups and foundations**

The following is a list of active movement disorder foundations and support groups in the United States. A comprehensive “Resource Handbook for Movement Disorders” and a directory of international organizations can be obtained from:

**WE MOVE** (Worldwide Education and Awareness for Movement Disorders)
204 W. 84th Street
New York, NY 10024
[www.wemove.org](http://www.wemove.org)
Benign Essential Blepharospasm Research Foundation, Inc.
2929 Calder Avenue, Suite 304
Beaumont, TX 77702
(409) 832-0788
www.blepharospasm.org

Dystonia Medical Research Foundation
8383 Wilshire Blvd.
Beverly Hills, CA 90211
(310) 852-1630
www.dystonia-foundation.org

Huntington’s Disease Society of America
140 West 22nd Street
New York, NY 10011-2420
(800) 345-HDSA
www.hdsa.org

National Parkinson Foundation, Inc.
1501 Ninth Avenue NW
Miami, FL 33136
(800) 327-4545
www.parkinson.org

The Parkinson’s disease Foundation
640 West 168th Street New York, NY 10032
(212) 305-3480 or (800) 457-6676
www.pdf.org

Society for Progressive Supranuclear Palsy
2904-B Marnat Road
Baltimore, MD 21209
www.psp.org

Shy-Drager Syndrome Support Group
Dorothy Trainor-Kingsbury
1607 SE Silver Avenue
Albuquerque, NM 87106
(505) 243-5118
www.shy-drager.org

National Spasmodic Dysphonia Association
PO Box 266
Birmingham, MI 48012
www.dysphonia.org

National Spasmodic Torticollis Association, Inc.
PO Box 873
Royal Oak, MI 48068-0873
Fax: (313) 362-4552
www.torticollis.org
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International Essential Tremor Foundation
P.O. Box 14005
Lenexa, Kansas 66285-4005
913-341-3880
FAX: 913-341-1296
Toll-free: 888-387-3667
www.essentialtremor.org

Wilson Disease Association
PO Box 75324
Washington, DC 20013
(202) 208-0934
www.wilsonsdisease.org

Restless Legs Syndrome Foundation
819 Second Street SW
Rochester, MN 55902-2985
Phone: 507-287-6465
Fax: 507-287-6312
www.rls.org

REFERENCES


Self-Assessment Questions On Gait And Movement Disorders

Please select one BEST answer to each of the questions below.

1. Spasmodic torticollis is an example of:
   A. a variant of Parkinson’s disease
   B. a movement disorder caused by botulinus toxin
   C. a genetic disorder caused by triplicate nucleotide repeat sequences
   D. a form of dystonia affecting the neck
   E. a form of epilepsy affecting the neck

2. Among the following, the most prevalent movement disorder is:
   A. Parkinson’s disease
   B. essential tremor
   C. Huntington’s disease
   D. Amyotrophic Lateral Sclerosis
   E. idiopathic torsion dystonia

3. Dystonic reactions may be associated with any of the following except:
   A. Compazine
   B. Benadryl
   C. haloperidol
   D. metoclopramide
   E. Navane
4. Ataxia is the term used to describe:
   A. a jerking or “flapping” of the hands seen in hepatic disease
   B. repetitive jerking movements of the body seen in certain drug withdrawal states
   C. the shuffled gait seen in Parkinson’s disease
   D. the wide-based gait seen in cerebellar disease
   E. a tremor exhibiting irregular frequency

5. A patient complains of neck pain and on examination you find that his head posture is asymmetric, with enhanced drifting to the right when the patient closes his eyes. The remainder of the physical examination is normal, without sensory or motor deficit of the upper or lower limbs and symmetric reflexes. The next step in the management of this patient should be:
   A. brain MRI
   B. cervical spine MRI
   C. cervical spine CT scan
   D. treat with clonazepam
   E. refer for consideration of botulinus toxin injections

6. An 11-year-old child is described as having facial twitching during sleep. Birth and development history are normal. The next step in the management of this patient should be:
   A. electroencephalography (EEG)
   B. brain MRI
   C. botulinus toxin injections for hemifacial spasm
   D. prednisone for idiopathic Bell’s palsy
   E. serum and urine copper and serum ceruloplasmin levels

7. On a routine follow-up examination of a 61-year-old man with a ten-year history of Parkinson’s disease you find new weakness and hyperactive reflexes on the right arm and leg. The significance of these changes is:
   A. motor fluctuations commonly seen in advanced Parkinson’s disease
   B. the dose of Levodopa is too low
   C. a brain MRI scan should be obtained
   D. the patient is exhibiting dominant-side dystonia
   E. these changes may be a side effect of bromocriptine
8. A 56-year-old woman comes to see you because of a tremor she has developed in her arms. She is also reporting recent symptoms of anxiety and excessive sweating. Your next step in the management of this patient is:
   A. prescribe carbidopa-levodopa for probable Parkinson’s disease
   B. prescribe primidone for her tremor
   C. order thyroid function studies
   D. order MRI scan of the neck
   E. all of the above

9. Blepharospasm is:
   A. seen in association with cranial dystonia
   B. a type of partial epilepsy
   C. seen in association with idiopathic Parkinson’s disease
   D. a movement disorder manifested by twitching of one side of the face and eyelid
   E. A and C

10. A 26-year-old school teacher comes to your office because of painful cramping of the hand brought on by writing. You are able to reproduce the symptom in your office by having her write a few words down on paper, and, in addition, you note that her wrist assumes a hyperflexed posture with the digits hyperextended, so that she is unable to continue writing. Your next step in the management of this patient is to:
    A. administer calcium IV STAT, for hypocalcemic tetany
    B. prescribe trihexyphenidyl (Artane®) for writer’s cramp
    C. refer the patient to an orthopedist for carpal tunnel release surgery
    D. administer Benadryl iv STAT for acute dystonic reaction
    E. refer the patient to a neurologist

11. A 45-year-old man with a long history of schizophrenia returns to see you after being lost to follow up for about one year. On examination, you note that his posture is stooped, his gait and overall mobility are slow, and his right hand exhibits a tremor when resting on his lap. You decide to:
    A. start carbidopa-levodopa (Sinemet®)
    B. refer the patient back to the psychiatrist for evaluation of impending catatonia
    C. order MRI of the brain
    D. review his list of medications to look for iatrogenic causes
    E. administer a test dose of Benadryl®
12. A 60-year-old woman with recently diagnosed Parkinson’s disease complains of nausea. You believe this may be related to the Sinemet® prescribed last week by the neurologist. Your next step in management should be to:

A. discontinue the Sinemet®
B. add bromocriptine to the patient’s regimen
C. treat the nausea with prn metoclopramide (Reglan®)
D. tell the patient to take Sinemet® on an empty stomach
E. contact the neurologist

13. A 70-year-old man with Parkinson’s disease complains of visual hallucinations. He is currently treated with Sinemet CR® and bromocriptine (Parlodel®). The best approach to manage the hallucinations is to:

A. discontinue Mirapex® and begin Requip®
B. decrease the dose of Mirapex® and increase the dose of Sinemet CR®
C. decrease the dose of Sinemet CR® and add regular Sinemet® to the regimen
D. begin clozapine 25 mg po bid
E. begin Haldol® 0.5–1.0 mg po tid and increase the dose gradually as needed

14. A 45-year-old bank executive whose essential tremor had initially responded to propranolol (Inderal®) complains of problems with concentration and memory. He is certain that the drug is responsible because he discontinued it and his performance at work improved, according to his colleagues. You decide to:

A. discontinue Inderal® and begin Artane® for treatment of the tremor
B. refer the patient to a psychiatrist for possible dementia
C. add donepezil (Aricept®) to his treatment regimen
D. refer the patient to a neurologist for medication change
E. order MRI of the brain

15. A patient reports problematic unpleasant sensations in the legs at night with an urge to move. She often gets up and walks around to temporarily improve the symptoms. They also occur some evenings when she is forced to sit still. It would be most reasonable to:

A. obtain a sleep study
B. treat with a dopamine agonist
C. obtain serologies including thyroid tests, B12, and folate
D. treat with a sleeping pill
E. treat with low dose haloperidol
16. Resources that are available to patients with movement disorders include:
   A. patient support groups at the national level and local chapters for the patient and family
   B. foundation newsletters
   C. the primary care physician, neurologist, and non-physician team members
   D. Internet websites
   E. all of the above

**Answers**
1. D
2. B
3. B
4. D
5. E
6. A
7. C
8. C
9. E
10. E
11. D
12. E
13. B
14. D
15. B
16. E
17. A