Hyperkinetic Movement Disorders

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James' case

A mother brings her son James, 10, to your office because of repetitive sniffing sounds and repetitive eye blinking. This sniffing has been going on for about a year, but seems to wax and wane; it is worse prior to tests at school and while at home in the evenings, but it was better last summer, when James was away from school. James admits he tends to prevent his sniffing while at school and "lets things out" while he is at home.

Thinking back, James may have had a period of repetitive grunting in the past year, but this is no longer a problem. James admits to having a very neat and tidy room at home, unlike his friends. Nevertheless, he does well in school and has many friends. James' father is very neat and tidy like his son and may also have the same tendency to excessive sniffing.

Your exam of James is essentially normal, except for repetitive eye blinking and mild sniffing.

About Tom

Tom, 60, is right-handed. He presents to your office with tremor. He is concerned that he has Parkinson's disease (PD), as there is a family history of tremor. He says that he has had his tremor for about 5 years and wonders if it is getting worse. He notices the tremor primarily when he holds objects; there is no tremor at rest. The tremor is improved when he consumes wine or beer. His handwriting is described as slightly shaky, but not smaller, and is still legible.

Exam discloses a well-looking man with no head tremor and good facial expression and voice volume. He is also found to have fine tremor in both hands immediately present on holding both arms outstretched and on holding a cup; however, there is no tremor at rest and no evidence of slowness. He has normal tone and an otherwise normal neurologic exam. You have him write a sentence or two in the office and his handwriting is of normal size, but slightly shaky.

For more on Tom, go to page 59.

Hyperkinetic movement disorders are characterized by excess of movement and may be classified as certain tremors, chorea/dyskinesia, dystonia, and tics. The family doctor will likely encounter some of these movement disorders in his/her practice.

More on Tom

Tom returns to your office. His metabolic workup for tremor has been normal. He is not taking any medications that might cause tremor. On the followup visit, he does not find the tremor very disturbing and would prefer to live without medications aimed at treating his tremor. He is very relieved when you inform him, on the first visit, that he does not have Parkinson's disease, but essential tremor.

How is postural tremor different from Parkinson's disease?

Postural tremor is probably the most common movement disorder likely to be encountered by the family doctor. It is a tremor that is present in the hands when the arms and hands are held outstretched or when the patient holds objects, such as a cup. No rest tremor is typically present in a patient with postural tremor and this important abscence, as well as the absence of other signs of Parkinson's disease (PD) helps the doctor distinguish postural tremor from PD (Table 1).

What are the causes of postural tremor?

When the differential diagnosis of a postural tremor is made, and when other disorders are excluded (a metabolic or drug-induced tremor), if there is a family histo-

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Bob's Parkinson's disease

Bob, 65, presents with Parkinson's disease diagnosed by a neurologist 8 years ago. He continues to be managed by his neurologist. He has responded well to levodopa and a dopaminergic agonist over the years; this treatment has improved his now bilateral resting tremor and slowness (bradykinesia) significantly.

Recently, Bob has become concerned that his tremor is becoming worse and he has tried to increase his levodopa consumption. Unfortunately, this does not seem to help. He is now bothered by his movements, which seem to be bad several hours after taking his last levodopa tablets, and then improve in the hours leading up to his next dose of levodopa.

When you examine Bob, 2 to 3 hours after his last dose of levodopa, you find his exam predominated by extra, involuntary movements in his trunk and legs. These are moderately disruptive to him and there is really no evidence of resting tremor or bradykinesia.

ry of tremor that is responsive to alcohol, the diagnosis of essential tremor is made (Table 2).

Table 1
Features distinguishing postural tremor from Parkinson's disease

	Essential tremor	Parkinson's disease
Tremor	Present with holding objects; no tremor at rest	Tremor at rest; no tremor with holding objects
Family history	Common	May be present
Response to alcohol	Often improves tremor	No improvement of tremor
Other signs	Head tremor	Slowness of movement, cogwheel rigidity, soft voice, decreased facial expression
	Shaky handwriting; normal size	Small handwriting (micrographia)

How should essential tremor be treated?

In many patients, medications are not necessary. However, if the tremor is bothersome and interferes with daily activities, first-line medications include propanolol (up to 240 mg/day in divided doses, or other beta blockers) or primidone (up to 250 mg/day in divided doses). Beta blockers are contraindicated in people with asthma and/or diabetes. Common side-effects of beta blockers include fatigue, hypotension, and depression. Common side-effects of primidone include dizziness and fatigue. If these two first-line medications are unsuccessful, other medications, including gabapentin, topiramate, and even mirtazapine, can be tried.

In those few patients who do not respond to medications, but need treatment, surgery may be considered. Creation of a discrete lesion in the thalamus (thalamotomy) or placement of a deep brain stimulator in the thalamus (thalamic stimulation) may prove beneficial in severe cases.

Table 2
Differential diagnosis of postural tremor

- Essential tremor
- Hyperthyroidism
- Renal failure
- Liver failure
- Drug-induced tremor (*e.g.,* lithium, valproate)
- Dystonia

What are tics?

Tics are brief, repetitive, stereotyped movements, such as eyeblinking or grimacing; vocal tics can include throat clearing or grunting. Patients may describe some sort of sensory premonition prior to tic onset; they may have the ability to suppress tics in certain social situations, but also have the need to "let them out" later, in a socially supportive situation.

Table 3

Treatment of tics, ADHD, and obsessive compulsive tendencies

A. Treatment of tics
- Supportive: Education of family, teachers, and friends
- Pharmacologic:
  - Clonidine, 0.1 mg bid-tid
  - Tetrabenazine, 25 mg tid
  - Quetiapine, 25 mg bid-tid
  - Pimozide
  - Haloperidol

B. Treatment of ADHD
- Dexedrine
- Methylphenidate

C. Treatment of obsessive compulsive tendencies
- SSRIs

bid: Twice daily
tid: Three times daily
ADHD: Attention deficit hyperactivity disorder
SSRIs: Selective serotonin reuptake inhibitors

What is Tourette's syndrome (TS)?

TS is characterized by multiple motor and one or more vocal tics, onset of tics prior to the age of 18, and presence of tics for more than one year; other causes of tics, such as drugs, infections, or other neurologic disorders, should be excluded. Associated features may include attention deficit hyperactivity disorder (ADHD) and obsessive compulsive traits. Boys are more affected than girls and male members of the family may be affected with some or all of these symptoms and signs.

How should TS be treated?

Treatment of TS is multifaceted.

1. First, it is important to garner the understanding of family and teachers. It must be understood that tics are a part of a medical condition, and not an attempt to disrupt the family or classroom. It should also be recognized that tics wax and wane in severity throughout the year.
2. Second, treatment of tics should be initiated only when tics are severe enough to disrupt school or home life. Treatment should not be initiated because those surrounding the child feel uncomfortable with the tics.

In general, treatment of tics starts with clonidine, then quetiapine or tetrabenazine (the prescription of tetrabenazine may need special approval). Higher potency neuroleptics, such as pimozide or haloperidol, may be used if these initial medications are not helpful and tics are severe.

Table 4
Dyskinesia versus resting tremor and bradykinesia in Parkinson's disease

Dyskinesia	Resting tremor
Writhing, involuntary movements; affecting legs and trunk	Rhythmic, low frequency tremor at rest; can affect arms, legs, or chin
Usually no associated bradykinesia	Associated bradykinesia (slowness)
Peak dose dyskinesias come on several hours after last dose of levodopa	Improvement in resting tremor and bradykinesia soon after medications

However, use of these latter medications carries the risk of tardive disorders later in life. The symptoms of ADHD are typically treated with stimulants, such as dexedrine or methylphenidate, and symptoms of obsessive compulsive tendencies are treated with selective serotonin reuptake inhibitors (Table 3). Complicated cases of TS are best managed with the help of a movement disorders neurologist or pediatric neurologist and pediatric psychiatrist. However, many children outgrow their tics by adulthood, although a period of worsening during puberty may be expected.

PD and drug-induced dyskinesia

PD is traditionally classified as a hypokinetic movement disorder because of the bradykinesia associated with the condition. However, the drugs used to treat PD can produce extra, involuntary writhing movements called dyskinesias. Dyskinesias are quite different from the resting tremor and bradykinesia characteristically associated with the disorder. It is important to encourage patients to distinguish dyskinesia from resting tremor. Furthermore, the treatment of dyskinesia is quite different from the treatment of bradykinetic symptoms and signs (Table 4).

How should dyskinesias be treated?

In general, dyskinesias associated with moderate PD are peak dose dyskinesias and come on several hours after the last dose of levodopa. These involuntary writhing movements in the legs and trunk gradually improve, and are eventually replaced by resting tremor and slowness of movement as time to the next dose of levodopa approaches. This type of dyskinesia may be treated by reducing the individual doses and increasing the frequency of doses of levodopa. Other approaches are to add a dopaminergic agonist, with reduction in levodopa dosages, or to add amantadine. If dyskinesias are mild and not bothersome, no change in medications is needed.

However, as PD patients progress in their disease, other dyskinesia may occur, requiring different management. Patients who fulfill certain specific criteria may be considered for surgery (*i.e.*, subthalamic nucleus stimulator placement). In cases of advanced PD, ongoing care through a neurologist with special expertise in PD will be of benefit to the patient. CME

Suggested Readings
1. Furtado S, Pfeiffer RF, Uitti RJ et al: Neurotherapeutics and Movement Disorders. World Federation of Neurology Web site.www.wfneurology.org/research_advances_neurology.htm
2. Grimes, DA: Tremor-easily seen but difficult to describe and test. Can J Neurol Sci 2003; 30(Suppl 1):S59-63.
3. Martin WR, Wieler M: Treatment of Parkinson's Disease. Can J Neurol Sci 2003; 30(Suppl 1):S27-33.

Take-home message

Tremor

- When the differential diagnosis of postural tremor is made and other disorders are excluded, if there is family history of tremor responsive to alcohol, diagnosis of essential tremor is made.
- First-line treatments for essential temor are propanolol and primidone. Surgery may be considered.

TS

- Treatment for TS starts with clonidine, followed by quetiapine or tetrabenazine.

Dyskinesias

- Dyskinesias are associated with moderate PD.
- Dyskinesias can be treated by reducing the doasge of levadopa, while increasing the frequency of dosing
- Advanced PD requires specialist referral.