

Idiopathic Parkinson's Disease: Diagnosis and Management



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The typical GP will have two or three cases of idiopathic Parkinson's disease (PD) in their practice. Although it is a relatively common movement disorder, most GPs will only see one new case every several years.

Diagnosis/differential diagnosis

The diagnosis can easily be missed on presentation, especially when there is no tremor at presentation, which occurs about 30% of the time. It is not unusual for the early symptoms to be dismissed as arthritis or "aging" until readily recognized by a family member or health professional, who has not seen the affected person for some time. A wide range of erroneous diagnoses may be considered initially, including stroke and even carpal tunnel syndrome.

Typical PD presents unilaterally with a wide range of symptoms, like patients reporting some awkwardness using one arm and often describing non-specific symptoms such as "weakness" or "numbness." It is always important to avoid taking these symptoms at face value. Some will present with a frozen shoulder syndrome.

Physical exam

When a person presents with functional difficulties with a limb, the physical examination must include assessment of the speed of move-

Meet James

James is a 70-year-old male who presents with awkwardness using the right arm. His symptoms seemed to evolve quickly:

- Minor change in gait and seems slower
- Previously healthy and active
- Initially considered to have had a possible stroke, but CT scan was negative
- Careful re-examination revealed cogwheel rigidity in the right arm and some slowness of rapid movements of the right arm and leg

Turn to page 13 for more on James.

ment, to assess for the presence of bradykinesia. This can be done with a variety of repeated hand/arm movements such as rapidly tapping the thumb and index finger (hand should be wide open, then tap finger/thumb and repeat several times). Other useful methods include rapidly opening and closing the fist or rapidly pronating and supinating the forearm, as if screwing in a light bulb. The best value of these tests is achieved by comparing the sides, as initially, only one side will be affected. Also observe for lack of facial expression, reduced blinking and general slowness of movement. Enquire about change in writing (smaller) and speech (quieter, monotone).

It is also essential to assess muscle tone. The

forearm can be flexed and extended at the elbow and the fist can be rotated at the wrist. Resistance to slow passive movements indicates increased tone and when it has a jerky, ratchet-like quality, it is called cogwheel rigidity, a classic finding in PD. This sign can be enhanced by voluntary activation of the contralateral body part (Froment's sign).

The tremor of PD is classically noted at rest, when attention is not being drawn to the limb. It will commonly be observed when the person is walking with arms relaxed by the side. With action, the tremor diminishes markedly or disappears. With the outstretched arms, the tremor will initially settle, but may re-emerge when a posture is maintained for several seconds. Ultimately, a judgment needs to be made regarding the dominant element of the tremor. Jaw, lip or tongue tremor is most likely due to PD, whereas head tremor is most likely due to essential tremor. Keep in mind many drugs can cause tremor including the neuroleptic drugs, valproic acid and lithium.

Features of PD

A key differential diagnosis when tremor is a dominant feature is essential tremor. In this case, the tremor is maximal with action or maintained posture with little or no tremor at rest. However, it must be kept in mind that essential tremor is common in the whole population and more common in persons with PD. Therefore, a person with PD may have rigidity and bradykinesia with a non-parkinsonian essential tremor or may have both the essential tremor and PD tremor.

It is exceptionally useful to watch the individual walk. In PD, not only can the rest tremor be easily observed, a very distinctive lack of arm swing can be noted. This will be most clearly evident if the person is watched walking along a hallway and will be easily missed if only a few steps are taken in a small examining room. Since PD typically presents unilaterally, this is arguably the

most useful diagnostic tool in the early recognition of PD. Watch and listen to the person's steps while they are walking. A subtle heel scuff indicates reduced length and amplitude of stride of one leg.

Depending on the practice setting, diagnostic errors in PD may exceed 20%. Atypical forms of PD commonly present bilaterally and symmetrically, without tremor. They will be relatively resistant to PD therapy. There may be other associated features early in the presentation, such as:

- impaired downgaze,
- a wide-based ataxic gait,
- autonomic dysfunction of bowel, bladder or sexual function and
- significant postural hypotension.

It is essential to review the drug history as the neuroleptic drugs, especially the typical ones, can lead to typical parkinsonian clinical signs. Other drugs that may result in parkinsonian features include metoclopramide and possibly lithium.

Occasionally, a variety of other conditions may have some features that can, at first look, appear to be PD. This includes multiple lacunar strokes, hydrocephalus and subdural hematoma. While typical PD probably needs no radiological studies, a CT scan will be helpful to assess these other considerations and will be especially important when the clinical features are not typical.

Management

Once the diagnosis is secure, the treatment plan can be established. Good principals of patient education are especially important in this condition. Many people will have erroneous ideas, confusing it with Alzheimer's, amyotrophic lateral sclerosis (ALS), *etc.* A well-educated patient and their family will be well equipped to participate effectively in key therapeutic decisions throughout the course of the illness.



A physiotherapist must be involved from the beginning, to instil the principals of a regular exercise program. Physiotherapy assessments will also be needed periodically during the course of the illness to focus on various problems that emerge, particularly as abnormal posture and instability develop. Principals of drug management are summarized as follows:

- Early on, drug therapy may not be needed, if functional difficulties are minimal
- There is emerging evidence that the Monoamine oxidase (MOA-B) inhibitor rasagiline has a disease modifying/neuroprotective benefit
- Amantadine is a useful and easy drug to use (100 mg one to three times per day). It is often effective for a year or two before other therapies are needed
- 3,4-dihydroxy-L-phenylalanine (L-DOPA) remains the single most effective drug therapy for PD. It is given with a DOPA decarboxylase inhibitor (such as carbidopa) to prevent the peripheral metabolism of L-DOPA. In Canada, the most widely used form is levodopa and carbidopa. A convenient dose format is levodopa and carbidopa 100/25, initially about three times a day, gradually increasing as necessary. The controlled release format of levodopa and carbidopa, has about 20% less bioavailability, but is a reasonable option with a more gradual onset and longer duration of effect. L-DOPA therapy can be the first-line of therapy in persons > 70-years-of-age
- Dopamine agonists (ropinirole, pramipexole) are very useful in moderate stages of the disease, in conjunction with L-DOPA therapy, particularly to help manage fluctuations. They are also indicated for early onset PD before the introduction of L-DOPA therapy to delay the emergence of dyskinesia, which are more commonly a problem in early onset PD

FAQ

When should I refer a patient with probable Parkinson's Disease (PD)?

Many patients will benefit from involvement in a specialized movement disorder clinic.

Indications for referral include:

- Early onset PD, perhaps before the age of 60
 - Diagnostic uncertainty/atypical features
 - Emergence of fluctuations such as wearing off phenomenon, peak dose dyskinesia
 - Failure to respond to therapy
- A catechol-O-methyltransferase inhibitor (entacapone), which inhibits the peripheral metabolism of L-DOPA is a useful add-on therapy for wearing off phenomenon
 - Surgical management is a useful adjunct to the management of moderate stages of PD, in person's who clearly respond to therapy, but have fluctuations that are difficult to control

Follow-up

The primary care physician in the course of follow-up, should monitor for the emergence of motor complications of the disease and its treatment. After several years of treatment, the effect of L-DOPA will not last from one dose to the next, marking the emergence of wearing off phenomenon. Another common problem is peak dose dyskinesias, involuntary movements occurring as the effect of dopaminergic therapy becomes established. Some persons will fluctuate widely and unpredictably. In more advanced disease, excessive periods of slowness may follow a large protein meal.

Postural instability invariably occurs eventually and while it is important to optimize medical therapy, this symptom is usually refractory to

medications and guidance from a physiotherapist will be critical to educate the patient and family to limit the risk of falls.

The primary care physician should also monitor for the non-motor side-effects of treatment and non-motor manifestations of the disease. Common side-effects of dopaminergic therapy include:

- nausea,
- postural dizziness,
- nightmares and
- visual hallucinations.

The dopamine agonists may cause drowsiness and infrequently sudden sleep episodes. They also cause a distinctive impulse control disorder which may result in pathological gambling, sexual behaviour and impulsive shopping. Peripheral edema occurs with dopamine agonists and amantadine; levido reticularis occurs with amantadine.

Non-motor manifestations of the disease include cognitive impairment which can eventually occur in at least 30% of persons with PD. When this is a prominent feature at presentation, consider Lewy body dementia. Autonomic features can emerge later in the course of typical PD. It is of particular value to inquire about orthostatic dizziness and to periodically check for postural hypotension, as this may contribute to falls and faints. Depression is common. Swallowing can be affected and if so, a speech language pathologist must be involved and drooling can also be problematic.

In summary, the typical GP will encounter a new person with PD every few years. When a person presents with functional difficulties of the limbs, the clinical examination must include

James' case cont'd

James has 2 of the diagnostic criteria for PD:

- There was reduced arm swing on the right
- There were no other abnormal findings

In the absence of other clinical findings and no antecedent medical or drug history, this is most likely typical PD. In the absence of tremor, the key to accurate and early diagnosis is to consider the diagnosis, which should prompt a look for the key findings of rigidity and bradykinesia.

observations of tone and speed of movement. When tremor is present, it must be distinguished from essential tremor and drug-induced tremor. In typical cases, the FP can initiate therapy. Referral to a neurologist will be useful in several situations. The primary care physician will also need to monitor for the many drug side-effects and the motor and non-motor complications of the disease.

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