Parkinson's Disease Foundation

Understanding Parkinson's Plus Syndromes and Atypical Parkinsonism

The term "parkinsonism" refers to a group of diseases that are all linked to an insufficiency of dopamine in the basal ganglia — the part of the brain that controls movement. Symptoms include tremor, bradykinesia (extreme slowness of movement), flexed posture, postural instability, and rigidity. A diagnosis of parkinsonism requires the presence of at least two of these symptoms, one of which must be tremor or bradykinesia.

By far the most common form of parkinsonism is idiopathic, or classic, Parkinson's disease (PD), but for a significant minority of diagnoses — about 15 percent of the total — one of the Parkinson's plus syndromes (PPS) may be present.

These syndromes, also known as atypical parkinsonism, include corticobasal degeneration, Lewy body dementia, multiple system atrophy, and progressive supranuclear palsy. Each of these syndromes has its own distinctive set of symptoms that can help doctors diagnose them. Although there is currently no cure for Parkinson's plus syndromes, researchers are making advances to better understand and manage them.

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Corticobasal Degeneration: The main symptoms of corticobasal degeneration (CBD) are apraxia (inability to perform coordinated movements or use familiar objects), pronounced asymmetry, stiffness that is more severe than classic Parkinson's, and myoclonus (twitching or jerking) usually in the hand.

Lewy Body Dementia: Lewy body dementia (LBD) is one of the most common types of progressive dementia. Its central feature is progressive cognitive decline, combined with pronounced fluctuations in alertness and attention, complex visual hallucinations and motor symptoms, such as rigidity and the loss of spontaneous movement.

Multiple System Atrophy: Multiple system atrophy (MSA) is characterized by symptoms of autonomic nervous system failure (such as lightheadedness or fainting spells, constipation, erectile failure in men, and urinary retention), combined with tremor and rigidity, slurred speech, or loss of muscle coordination.

Progressive Supranuclear Palsy: The cardinal symptoms of progressive supranuclear palsy (PSP) include frequent falls, an inability to aim the eyes properly, especially in a downward gaze, and emotional and personality changes.

Diagnosis: Is It PD or PPS?

Initial signs indicating that the diagnosis may be PPS and not PD include early and severe dementia, falling, and difficulty with voluntary eye movements. Tremor is often not a presenting symptom. Stepwise deterioration (a series of episodes that result in a sudden decline in functioning followed by periods of time where the individual's condition remains relatively stable, but resulting in an overall worsening of that condition) might also point toward a PPS diagnosis.

Another measure used to differentiate PPS and PD is the individual's response to medications typically used to treat Parkinson's. When given these medications (primarily levodopa), individuals with PPS generally have only a slight response or no response at all. In some cases, there may be an initially strong response to the medication, but this response will not be long lasting. For this reason, a doctor may conduct a trial of levodopa at doses higher than those normally used to treat Parkinson's. If this approach is used, it is important that the individual with PPS and his/her care partner have easy access to the doctor during this time. If these high doses do not signifi-

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cantly help the symptoms, the doctor may gradually lower the dose and possibly discontinue the medication altogether.

Treatment Approaches for PPS

For some people with PPS, a low dose of levodopa helps with mobility and reduces rigidity and stiffness. At this time, people with PPS are not considered to be candidates for any of the surgical treatment techniques available for PD including deep brain stimulation (DBS). Because there are currently no medications that effectively treat PPS, the prognosis for these syndromes is not as positive as for medication-responsive PD.

A common problem for people who live with PPS is swallowing or choking. In such cases, the doctor will usually order a swallow study, which involves a series of x-rays that allow the doctor to see how a liquid such as barium passes from the mouth to the throat and down the esophagus. Depending on the results, the doctor may direct adjustments in diet, such as the pureeing of food and/or the thickening of liquids. At some point, gastrostomy (a surgical procedure that involves the placement of a tube through the skin of the abdomen into the stomach for feeding purposes) may be considered.

Cognitive problems may also arise for some individuals with PPS. If a person is experiencing hallucinations, these may be alleviated somewhat with the use of quetiapine or clozapine. For dementia, a comprehensive neuropsychological evaluation once a year can accurately measure the decline of cognitive abilities and suggest coping strategies. There are techniques that can reduce frustration and encourage the maximum amount of independence for the person with PPS. These include limiting the number of choices offered and cueing. For additional information, please see the PDF Fact Sheet, "Coping with Dementia: Advice for Caregivers," at www.pdf.org.

A growing number of clinical trials are studying PPS. For more information on these trials and to find one in your community, please visit www.PDtrials.org.

Complementary Treatment Approaches for PPS

Although there are currently no effective treatments for Parkinson's plus syndromes, some relief may be found in complementary approaches, including exercise and physical, occupational and speech therapy. For all PPS conditions (as for classic Parkinson's disease), a regular daily exercise program is vital for maintaining muscle tone, strength, and flexibility. A physical therapist trained in neurological conditions and PD can design an appropriate program as well as suggest a walker (one with wheels and brakes) that meets the special needs of a person who lives with one of the PPS conditions. An occupational therapist can be brought in to assess the individual's abilities and home environment and make recommendations that will allow for greater independence while at the same time assuring safety. A speech therapist can be asked to develop a program to improve voice articulation and volume.

In summary, Parkinson's plus syndromes can be very difficult to diagnose and are difficult to treat. Because the cardinal symptoms of the individual disorders may take a long time to become visible or may never appear at all, a doctor may not be able to say exactly what is causing the parkinsonism. In addition, because these syndromes are complex and rare, complete diagnostic clarity may only come after the patient has been followed for several years by a movement disorder specialist (MDS) — that is, a neurologist who has completed additional, specialized training in movement disorders. For more information or to locate a MDS in your area, please contact PDF's Parkinson's Information Service helpline at (800) 457-6676.

For support and additional information:

Lewy Body Dementia Association
(404) 935-6444
Helpline: (800) 539-9767
www.lewybodydementia.org

Shy-Drager/MSA Support Group
(866) 737-4999/5999
www.shy-drager.org

Society for Progressive Supranuclear Palsy
(410) 785-7004
www.psp.org

If you have or believe you have Parkinson's disease, then promptly consult a physician and follow your physician's advice.
This publication is not a substitute for a physician's diagnosis of Parkinson's disease or for a physician's prescription of drugs, treatment or operations for Parkinson's disease.

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