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RUSH UNIVERSITY
MEDICAL CENTER

PARKINSON NEWSLETTER

Atypical Parkinsonism: Multiple System Atrophy, Progressive Supranuclear Palsy & Corticobasal Degeneration

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Many patients in our practice do not have typical Parkinson's disease (PD). Rather, they have "atypical parkinsonism," which includes several different diagnoses: "multiple system atrophy," "progressive supranuclear palsy," or "corticobasal degeneration."

Patients with these atypical parkinsonian conditions often have symptoms that are quite different from PD. However, they may be indistinguishable from PD early in the disease. As care providers, we routinely "rethink" diagnoses and it is common practice to tell a patient we thought had PD that the disease has evolved in such a way that another diagnosis is more appropriate. This article will discuss some of the more common forms of atypical parkinsonism.

How are PD and atypical parkinsonism alike and how are they different?

The atypical parkinsonisms share some core features with PD: most prominently, slowness of movement, muscular rigidity and disorders of walking and balance. Yet there are important fundamental differences. Patients with atypical parkinsonism are much less likely to have tremor of body parts at rest and may have symptoms that are pretty symmetric on both sides of the body. Their symptoms are much less likely to improve when taking medications used for PD such as carbidopa/levodopa and others. Patients with atypical parkinsonism are more likely to have symptoms such as marked changes in blood pressure when going from sitting to standing, difficulty moving the eyes around, severe speech and swallowing disturbances, prominent disorders of thinking and memory, urinary incontinence and impotence.

Within the general family of atypical parkinsonism, there are many specific subtypes that will be discussed below.

How is the diagnosis of atypical parkinsonism made?

There are no specific laboratory or other tests that can reliably distinguish between PD and atypical parkinsonism. The diagnosis is made based on a history and physical examination. Following a patient over time is often necessary to be sure, because patients with atypical parkinsonism may be difficult to distinguish very early in the disease. A general rule of thumb is that most atypical parkinsonism patients can be identified by the time they have been sick 5 years.

What are the most common atypical parkinsonisms?

Multiple system atrophy (MSA) is the most common atypical parkinsonism, affecting about 4 out of every 100,000 people. Patients become sick at an average age of about 57 years. MSA patients have slowness, stiffness and trouble walking similar to that seen in PD, but patients are less likely to have tremor and more likely to have symptoms that are symmetric from side to side.

There are many other clinical features that help point the doctor to the correct diagnosis including a characteristic downward pulling of the head to the chest (anterocollis), very soft and high-pitched voice or a voice that is quite irregular in loudness and rhythm.

When there is tremor, it tends not to be a tremor at rest. Rather, patients may have tremor when holding the arms outstretched or when using the hands or arms actively. The gait of MSA patients is slow and there may be a drunken quality with staggering and falling or prominent "freezing" (feet getting stuck to the floor) with falling. The most characteristic

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feature of MSA is derangement of autonomic functions. Autonomic functions are those that occur automatically, such as maintenance of blood pressure and urination, defecation and sexual function. Patients with MSA often have very high blood pressures when lying down and substantial drop in blood pressure when standing. When standing blood pressure is quite low, a feeling of faintness or even fainting can result. Other signs of autonomic dysfunction include incontinence of urine or stool, loss of normal sweating and impotence (See Table 1).

Table 1: Common features of MSA

Feature	Percentage
Urinary incontinence	55
Faintness when standing	51
Impotence	47
Fecal incontinence	12
Wide, unsteady walking	49
Clumsiness using arms	47
Intellectual deterioration	25

The progression rate of MSA is faster than in PD. About 50% of MSA patients will require a walker by 3 years or wheelchair by 5 years.

The treatment of MSA includes medications normally used to treat PD, (especially carbidopa/levodopa and amantadine). These may help MSA patients move better or have better balance, but they may worsen the blood pressure regulation. Low blood pressure can be managed by adding salt and caffeine to the diet, sleeping with the head of the bed elevated 30 degrees, elastic stockings or medications (fludrocortisone, midodrine). Bladder medications or intermittent catheterization may help urinary symptoms. Antidepressants may be useful for depression. Speech and physical therapy can improve functional outcome. Patients with MSA have a more rapidly progressive course than do those with PD. The average survival is about 9 years from onset.

Progressive supranuclear palsy (PSP) is also rare, affecting about 6 out of every 100,000 people. The age at onset averages 66 years. PSP usually begins as a gait disorder with staggering and early falls. Other common features of PSP include a growling speech pattern, worried or startled facial expression, spontaneous eyelid closure or difficulty opening the eyes, rigidity of the neck and trunk, personality changes with increased emotionality, progressive lessening of speech output, and prominent trouble with eating (stuffing the mouth) and swallowing. Like PD, PSP patients have slowing of movement; but unlike PD, the slowing tends to be symmetrical. Treatment with carbidopa/levodopa or other PD medications, is generally not successful. Functional state may be improved by speech and physical therapy. Patients with PSP deteriorate faster than those with PD. The average survival is about 8 years.

Corticobasal degeneration (CBD) is the rarest atypical parkinsonism and it may produce several different clinical pictures. In typical cases, it is a very asymmetric motor illness. Some of the symptoms can be like those in PD, such as slowness and rigidity, but CBD patients usually have other symptoms such as abnormal arm postures (dystonia), and muscle jerking (myoclonus). Sometimes, profound difficulty controlling the arm is seen and many patients develop a limb that seems to have "a mind of its own," reaching out, interfering with activity or grabbing onto things on its own (alien limb). Some patients begin with prominent difficulty with language or with loss of thinking and memory functions. Some patients look very much like those with PSP.

Treatment with PD drugs is common, though many patients do not have a positive response. Drugs to help with other symptoms (baclofen, clonazepam, etc.) may also be used. Like the other atypical parkinsonisms, CBD patients decline more rapidly than PD patients, dying an average of 6-8 years after the disease onset.

Summary

A number of different diseases overlap with PD, and when PD isn't the correct diagnosis, atypical parkinsonism often is. The atypical parkinsonisms are rarer than PD and frequently misdiagnosed in the early stages. By the time the patient has been sick 5 years, the diagnosis of atypical parkinsonism may be clear. The three most important parkinsonisms are MSA, PSP and CBD. All show core features resembling PD, but with important differences, and all respond more poorly to medications and progress to death over a shorter time period. Treatment involves PD medications with medications to treat other symptoms as well as speech and physical therapy.☺

The Rush Parkinson's Disease Symposium 2009

Friday, April 17, 2009
12:00 pm to 4:30 pm
(Registration/sign-in begins at 11:00 am)

Hyatt Regency Hotel - O'Hare

Announcing the 9th Patient/Caregiver Symposium entitled: "The Rush Parkinson's Disease Symposium 2009" which will be held on Friday, April 17, 2009 from noon to 4:30 p.m. at the Hyatt Regency Hotel - O'Hare, 9300 W. Bryn Mawr Road, Rosemont, IL. This Patient/Caregiver Conference is sponsored by the Movement Disorder Section of the Department of Neurological Sciences at Rush University Medical Center in Chicago and the Parkinson's Disease Foundation (PDF). The moderators of the conference are Dr. Katie Kompoliti and Dr. Christopher Goetz. The conference is directed to the education of Parkinson's disease patients and their caregivers. The overall goal is to provide an understanding of the roles of the various drugs and surgical techniques in the approach to specific problems experienced by the Parkinson's disease patient. Refer to the newsletter insert for a preliminary agenda and topcis to be presented.

The last conference of this type was held in May of 2008 and had full attendance. Admission to the conference is free, but seating is limited. Advanced registration is required. In order to reserve your seat, please complete the registration form on page 3 of this newsletter and send it to us.

On the day of the conference, **sign-in will begin at 11:00 a.m.** The conference will be held in the Rosemont Ballroom of the Hyatt Regency Hotel. The hotel has an adjoining parking garage with a discounted self parking charge of $10.00. Valet parking is also available at the prevailing rate.

CANCELLATIONS:

After reserving your seat, if for any reason you find you cannot attend the conference, please contact our office at 312-942-8002 so that we may open your seat(s) up to other patients/caregivers.

The conference is partly supported by the following: The Parkinson's Disease Foundation.

(Complete form, detach and return)

REGISTRATION FORM

(Please print or type)

I would like to attend the Parkinson's Disease Patient/Caregiver Conference

Name (1):____________________

Name (2):____________________

Address:____________________

____

City:__________ State______ Zip______

Phone Number (____)________________

Number of persons attending:________________

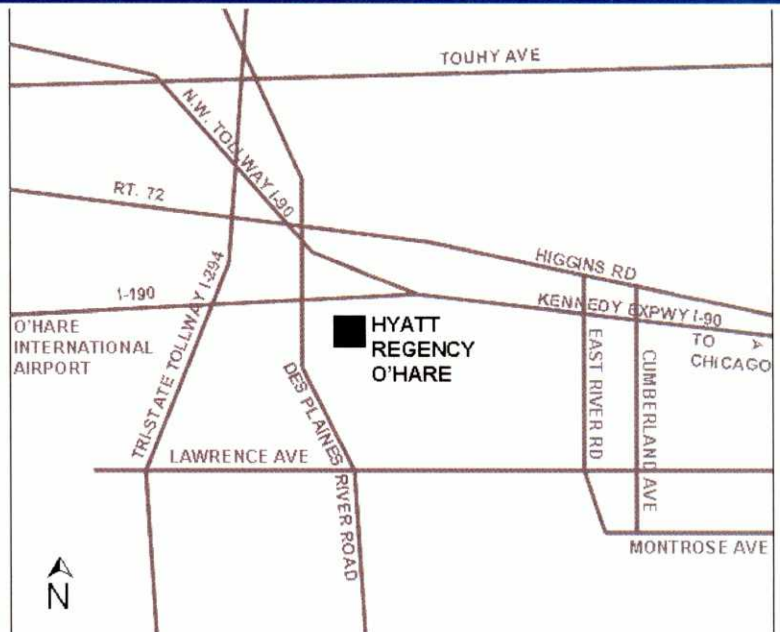
Please mail your completed registration form to:

Ms. Teresa Chmura
Rush University Medical Center
Department of Neurological Sciences
1725 W. Harrison Street, Suite 755
Chicago, IL 60612
Fax: (312) 563-2684 / Phone: (312) 942-8002

From O'Hare International Airport:
Take I-190 East to River Road South Exit. Hotel is on the left side.

From Midway Airport:
Take Cicero Avenue North to I-55 South to I-294 North. Follow directions to O'Hare. Exit River Road South. Hotel is on the left.

Venue: Hyatt Regency Hotel - O'Hare
Address: 9300 W. Bryn Mawr Rd., Rosemont, IL 60018
Phone: 847-696-1234



Monthly Educational and Support Program

WHEN: Second Saturday of each month, 10:00 am to 12:00 noon
LOCATION: Oak Park Hospital (Back of Cafeteria)

March 14: Dr. James Young
April 11: Dr. Emily Wang
May 9: Dr. Jeffrey Kordower
June 13: Dr. Melany Danehy

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Supported by the Parkinson's Disease Foundation

Your invitation to the PD Patient/Caregiver Symposium is included in this issue (see page 3)