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YOUNG PARKINSON'S HANDBOOK

A Guide for Patients and Their Families

The American Parkinson Disease Association

THE AMERICAN PARKINSON DISEASE ASSOCIATION, INC.

YOUNG PARKINSON'S HANDBOOK

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This handbook is a guide for Parkinson's disease patients and their families and is not intended as a substitute for medical diagnosis and treatment.

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INTRODUCTION

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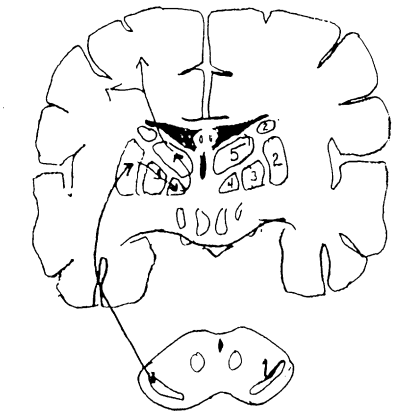
Parkinson's disease (PD) is a disorder of the central nervous system, involving primarily a degeneration of certain nerve cells in parts of the brain called the *basal ganglia*, and in particular a loss of nerve cells (or neurons) in a part of the brainstem called the *substantia nigra*. These cells make the neurochemical messenger dopamine, which is partly responsible for starting a circuit of messages that coordinate normal movement. In the absence, or with significant reduction, of dopamine, the neurons in the receiving area (called *dopamine receptors*) in the next part of the basal ganglia circuit called the striatum are not adequately stimulated, and the result is impairment of movement with *tremor*, slowness (*bradykinesia*), and stiffness (*rigidity*). Under the microscope, the damaged and dying neurons in the substantia nigra show a round, cellular marker called a Lewy body, which is considered the specific pathologic hallmark of PD.

Although the average age of onset of PD is around 60, young-onset PD (before age 40) occurs in about 5% of patients, but this number may be increasing with earlier recognition of symptoms and easy access to good medical care. Some problems in PD are universal regardless of age and disability; but there are frequently issues that are specific to younger patients. It is for these people and their families that this booklet is directed.

This booklet is not meant to be comprehensive and does not cover many topics, such as the basics of the signs and symptoms of PD and the related parkinsonian ("parkinson plus") disorders. For more complete information and details, we suggest you refer to another APDA booklet, the **Parkinson's Disease Handbook.**

For ease of reading, we will use the abbreviation "PD" for Parkinson's disease throughout this booklet.

Finally, the editors of this second edition of **the Young Parkinson's Handbook** would like to acknowledge gratefully Arlette Johnson, editor of the first edition. We hope this new, updated edition will be as useful to the young patients as the first edition was.



1. Substantia Nigra
2. Striatum
3. External Globus Pallidus
4. Internal Globus Pallidus
5. Thalamus

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CHAPTER 1
YOUNG-ONSET PD

Lawrence I. Golbe, MD

The average age at which the symptoms of PD begin ("onset age") is about 60, but about five percent of those with PD experience the first symptoms before age 40, defined as *"young-onset PD."* The medical literature applies the special term *"juvenile parkinsonism"* to those few cases whose symptoms appear before age 21, as patients this young, and many whose symptoms begin during their 20's, do not have typical changes of PD in their brains. (Of note is that the Japanese scientific literature refer to patients under 40 as "juvenile parkinsonism," so the source of information is important in defining age ranges.)

Many of the features of PD vary with the onset age. Most important is that the overall disability proceeds more slowly. Although there is considerable variation among individuals, someone whose PD onset age is 40 can expect to be able to work for another 15 to 20 years on average, while for someone with onset age 60, the average figure would be roughly half that. These figures are based on treatment that was available over the past decade or two. Currently available treatment probably prolongs both figures and future treatment will probably be even more effective at prolonging the productive lifetime of people with PD.

Young-onset PD also is less likely to lead to dementia (memory problems) and balance problems. It is more likely to include *focal dystonia*-that is, episodes of cramping or abnormal posturing of part of the body. A very common initial symptom of young-onset PD, in fact, is an ache or cramp in one shoulder or calf.

The response to levodopa (as in Sinemet) also distinguishes young-onset PD. Younger-onset patients are more sensitive to the benefits of all antiparkinson medication, but they tend to experience the *"dyskinetic"* side effects of levodopa sooner after starting the drug than older-onset patients. These side effects most often take the form of involuntary restless, writhing. or twisting movements of the head, shoulder, hands. or legs and occasionally of the eyelids, mouth, tongue, or trunk. They do not usually cause pain or interference in daily activities, but they can be annoying or embarrassing, often more so to one's family than to the patients themselves. Young-onset PD also tends to exhibit "dose-related fluctuations" at an earlier stage of the disease. These are variations in symptoms that occur over the few hours between one levodopa dose and the next and include *wearing-off* (where the effect of the medication wanes and PD symptoms return) and the *on-off effect* (abrupt changes from good [on] to poor [off] function).

These treatment-related features of young-onset PD have prompted most PD specialists to avoid using levodopa for as long as possible in

younger patients. This postpones the onset of dyskinesias and dose-related fluctuations.

The jury is still out on whether young-onset PD is more likely to be familial, or run in families, than older-onset PD. Those with young-onset PD are more likely to have relatives from the previous generation who are alive and can report symptoms, so there is probably a "reporting bias." Aside from a few families with large numbers of especially young-onset members, young-age appearance of PD in one person does not correlate well with the age at onset of any other family members who may develop PD. In other words, if you have PD that began at age 40, your sibling or your child is probably no more likely to develop PD than would be the case if your PD developed at age 60 or 70. Furthermore, the age at which that relative would develop PD, if they do, is unlikely to be as young as your own onset age.

Probably the most important aspect of dealing with young-onset PD is adapting one's family life, career, finances and emotional outlook to the various stresses of a potentially disabling condition. Talking with other people with the same condition is usually helpful-far more relevant and less distressing than talking with older people with PD. The APDA publishes a Young Parkinson's Newsletter and many referral centers maintain special support groups for young people with PD.

CHAPTER 2
MEDICAL TREATMENT OF PD

Jacob I. Sage, MD and Margery H. Mark, MD

Before reading this chapter, the reader may find it helpful to review some of the points discussed in the APDA booklet, *Parkinson's Disease Handbook*. The chapter on medical management in that pamphlet is a more complete discussion of the various options of treatment, from choices in early, mild disease through therapeutic decisions in the later stages of PD. For the most part, management of problems associated with advanced stage PD is not much different in younger and older patients. The major differences between younger and older patients is considering how to proceed at the beginning of treatment, and the issues of motor fluctuations and dyskinesias (abnormal involuntary movements).

Motor Fluctuations and Dyskinesias

As was noted in a previous chapter, younger patients may be more likely to develop earlier and more severe motor fluctuations than older PD patients. Significant disability from involuntary motor movements *(dyskinesias)* seems to be more frequent in young-onset patients. The exact percentage of patients whose PD will be complicated by dyskinesias is difficult to estimate. Traditional thought has been that 50% of the general PD population will have some motor fluctuations with dyskinesias within 5 to 10 years after diagnosis and beginning treatment. A recent comprehensive study, however, detected about a 20% frequency of fluctuations five years after the initiation of treatment in a large group of PD patients. Despite this more modest estimation, dyskinesias and motor fluctuations may eventually affect a larger percentage of young-onset patients; therefore, strategies aimed at reducing these problems are critical in the approach to treating PD.

We must admit at the outset that there is no definitive, proven strategy to reduce the incidence or delay the onset of fluctuations and dyskinesias in PD patients. Many factors must be taken into account (for example, specific symptims; job, social, family demands), and therapeutic decisions must be individualized , because every patient is different. Nevertheless, physicians and patients have many options for treatment. In the following paragraphs, we will discuss the rationale for different approaches to the treatment of early PD in younger patients and some of the theoretical considerations that underlie each of the various treatment regimens. In addition, since no one knows the reasons that younger patients may be more likely to develop fluctuations and dyskinesias than older patients, we can only speculate as to the causes, and we will discuss two hypotheses that may account for these phenomena. It is worth recalling at this point that when dyskinesias begin in an individual, they occur at doses of levodopa that *previously* had not produced

any abnormal involuntary movements. The reasons for this turn of events may be that receptors (nerve cells which receive the message from the chemical messenger) for dopamine are now hypersensitive to dopamine, that they have undergone some fundamental change over the years, and that a patient's PD has progressed. There are several ways to explain this increased receptor sensitivity:

Hypothesis 1. As PD progresses, there is a gradual decline in dopamine-producing cells. These cells normally send messages (via dopamine) to the cells on which the receptors are located. With dopamine loss, receptor cells do not receive an adequate number of messages and begin to behave abnormally, with increased sensitivity to the chemical messenger, dopamine. Levodopa, the chemical precursor of dopamine, eventually crosses into the brain (the brain will not allow dopamine to cross in) and is converted to dopamine in the brain. Early in the disease, when there is a fair amount of the brain's own reserve of dopamine, the patient experiences a smooth response with the addition of levodopa. As time progresses, however, concentrations of dopamine that used to provide a normal response now produce dyskinesias.

Hypothesis 2. Generally, levodopa is given several times per day. In the earlier stages of disease, brain cells can store the excess dopamine provided by multiple doses of levodopa for use throughout the day. As these cells and their capacity to store dopamine declines, the dopamine receptor cells receive "pulses" of dopamine: that is, they begin to see larger, intermittent amounts of dopamine in the first couple of hours of each dose of levodopa, followed by a significant drop in dopamine levels until the next dose is taken by the patient, and the cycle begins again. As the dopamine receptors see intermittent dopamine stimulation over the course of many years, they begin to react abnormally, and patients begin to experience motor fluctuations and dyskinesias.

It is important to note that the two hypotheses outlined above are just that: they are hypothetical and not proven. There are certainly other reasons to explain the onset of fluctuations that may come to light over the next few years. One up-and-coming area of research is genetics, which may explain why some patients get PD at a younger age while others get it when old or not at all. Such genetic programming may explain why only certain patients get complications of PD such as motor fluctuations while others do not. In the meantime, however, physicians try to use strategies, based on these hypotheses, aimed at minimizing fluctuations, and perhaps even delaying their onset.

Initiating Treatment

If we could slow down the progression of PD, one treatment option would be to begin with a neuroprotective agent. There are, however, no medications that are proven to slow the progression of PD. Our therapeutic aims, then, are primarily symptomatic with the intention of minimizing or delaying fluctuations.

Since levodopa is the safest and most effective treatment for all the signs and symptoms of PD but may be implicated as a contributing factor to the onset of motor fluctuations, one early treatment option may be to *delay the start of treatment* entirely. Most movement disorder specialists advocate delaying symptomatic treatment until a patient's complaints suggest a functional disability, meaning that there is something important, either socially or at work or both, with which the patient is having significant difficulty. The definition of functional disability varies from patient to patient and may be very different in a young patient compared to an older individual. Younger patients need to work to support themselves and their families and therefore may have functional requirements that elderly retired person do not share. Certain jobs may require fine motor skills, rapid movements, the ability to use complex machinery, and the need to walk long distances. Caring for or just keeping up with young children may require stamina and speed not necessary in retired grandparents. Every patient needs to consider whether he or she can do what is required at work or home despite the deficits of PD, whether things can be changed around to accommodate PD, or whether some form of treatment is needed early after diagnosis. Simple adjustments in daily activities, such as using a computer instead of writing with a pen, or setting up a carpool with co-workers or with other neighborhood children, may be enough to keep patients working efficiently and may be sufficient to avoid the institution of medical therapy. These sorts of accommodations must be considered and discussed with physicians, co-workers, friends, relatives, and support group members.

When a patient feels that some help in the form of medication is necessary, one strategy is to start with medications other than levodopa. There are a number of choices with which to start depending on symptoms and potential side; again, treatment options must be individualized. Some patients and physicians begin treatment with *selegiline* (Eldepryl®). This drug blocks the further breakdown of dopamine in the brain, thereby making each molecule of dopamine produced by brain cells work a little bit longer. It is effective in about 60% of patients and may delay the onset of treatment with levodopa by almost a year. Generally, when used by itself, it may only be useful for very early, mild symptoms.

Some physicians and patients choose to begin treatment with one of a group of drugs called *direct-acting dopamine agonists*. These are "dopamine-like" drugs that act on the dopamine receptor in a similar fashion to dopamine, which is the natural transmitter. The four major drugs in this category are (in order of how long they have been available): *bromocriptine* (Parlodel®), *pergolide* (Permax®), *pramipexole* (Mirapex®), and *ropinirole* (Requip®). The advantage of these agents is that they are usually more effective than selegiline and the benefit may last longer. Studies indicate that agonist benefit may be enough to sustain 35% of PD patients for up to 5 years without levodopa. They also tend to have a long duration of benefit from each dose and patients are much less likely to get motor fluctuations on these drugs.

Eventually, PD will usually progress beyond the ability for agonists alone to treat the signs and symptoms adequately, and ultimately the patient will require *levodopa* (Sinemet), which is the most effective treatment for PD. When initiating levodopa treatment, one needs to consider using the *controlled-release* form of levodopa (Sinemet CR) at the start of therapy rather than the standard i*mmediate-release* preparation. Since it is theorized that intermittent ("pulsed") dosing may play a role in sensitizing the dopamine receptors, *controlled-release* levodopa may be of benefit in the long run by providing more constant, physiologic dopamine receptor stimulation, and may potentially delay or decrease motor fluctuations. Unfortunately, a major study aimed at proving this point failed to show a benefit of controlled-release levodopa over standard immediate-release levodopa in preventing of delaying fluctuations in the first five years. Despite this disappointment, such a strategy still seems reasonable and may be proven to be effective at some future date, or in longer-term follow-up.

The newest class of drugs on the market are the *COMT inhibitors, tolcapone* (Tasmar®) and *entacapone* (Comtan®). COMT stands for catechol-O-methyltransferase, an enzyme that breaks down dopamine in the brain, but also exists in the gut, and it breaks down levodopa into an inert substance called 3-O-methyldopa before the levodopa can get into the bloodstream and cross into the brain. Therefore, inhibiting COMT in the gut allows more levodopa to get to the brain and then be converted to dopamine. Adding tolcapone or entacapone to levodopa stretches out the life of one dose of levodopa. Note that these drugs are only for use in combination with levodopa; they do not do anything by themselves, or even with the agonists. Reports of 3 cases (out of 60,000) of severe liver toxicity with tolcapone has created a policy of tolcapone only being used with frequent blood tests for liver function that can be obtained at any lab. Entacapone does not have this side effect. Nevertheless, tolcapone may be very useful in helping fluctuations in many patients; its action tends to be longer than that of entacapone, which generally must be given with each dose of levodopa. Entacapone is most useful for the early, mild fluctuator; while tolcapone should be reserved for the more severe fluctuators.

Younger patients may be able to tolerate some of the older antiparkinson drugs more easily than older patients and should consider such agents for specific symptoms. Drugs of the anticholinergic class such as *trihexiphenidy*l (Artane®) or *benztropine* (Cogentin®) may specifically help tremor and therefore can be considered early in the treatment of PD when appropriate. *Amantadine* (Symmetrel®) may be a mild drug to start with as well; it may help all of the symptoms, not just tremor. Interestingly, recent studies have shown that, when patients have disturbing dyskinesias, adding amantadine to the existing optimal regimen (usually at least 3 doses daily, sometimes 4) may actually decrease dyskinesias without decreasing levodopa or other drugs.

It is important to note that there are no definitely right or wrong strategies.

Remember: every patient is different, with different needs and different sets of problems. Each person must discuss his or her specific situation with the treating physician and together come up with the best treatment regimen for the individual at any given time. Be aware also that things change over time; fortunately, we have a lot of medications in our arsenal against PD (and more coming every year) to keep up with the symptoms and complications of PD and keep the patient functioning well for a very long time.

CHAPTER 3
SURGICAL TREATMENT OF PD

Jacob I. Sage, MD and Margery H. Mark, MD

Surgery has become a serious option for patients with PD due to the limitations of medical treatment in advanced stages, and because surgical procedures have direct benefits for some of the motor complications of PD. Young patients may, in fact, be better candidates for surgery than older patients because they are better able physically to tolerate long surgical procedures and because they tend to have fewer baseline and potential cognitive problems, both of which predict a good result from the surgery. Having said this, it is important to note that surgery should be considered *only* after all reasonable medical options have been tried and proved inadequate to control the symptoms and complications of PD.

Surgical procedures can be divided into two major categories: *1 - ablative lesions* and *deep brain stimulation;* and 2 - *transplantation.* Transplantation procedures either of human-derived or animal-derived cells are considered experimental and confined to a few research centers. We will therefore focus on ablative lesions and deep brain stimulation.

Ablative lesioning means that the surgeon puts an electrode into a specific part of the brain (see below) and burns out a small part of brain in the immediate area, killing all the cells in that area. These lesions, or destruction of cells, are permanent and, once completed, cannot be undone.

Deep brain stimulation (DBS) means that the surgeon puts an electrode into a specific brain area but leaves that electrode in place. The electrode is then attached to a stimulator underneath the patient's chest skin (very similar to what is done with a heart pacemaker), which then can be turned on or off at will. One can think of such stimulation as temporarily exhausting the cells in the local area of stimulation to the point where they are not functioning. The result, therefore, is the same as an ablative lesion. The stimulator can be turned on all the time, or used only for certain hours during the day when needed. It is a much more complex process than simple ablation, requiring permanent hardware in the brain and body, but is reversible if necessary and can be adjusted if significant benefit is not achieved early on after surgery. DBS is becoming the preferred surgical treatment because it is generally considered safer and more versatile than destructive lesions.

Currently, there are three targets for both ablative lesions and DBS: 1 - the *thalamus*; 2 - the *internal globus pallidus*; and 3 - the *subthalamic nucleus*. Thalamic lesions have the longest history while DBS of the subthalamic nucleus is the latest procedure. Each procedure has its particular indications and uses. Since either DBS or permanent lesioning can be performed in all three of these nuclei, choosing the best location and procedure for an individual patient becomes a very important neurological and surgical decision.

These decisions need to be made by a team consisting of a neurologist and neurosurgeon working together as experts in a PD center.

The thalamus is a good target to relieve tremor. Ablative lesioning *(thalamotomy)* has about a 90% success rate and the effect tends to be long lasting. Unfortunately, the other major symptoms and signs of PD are not significantly improved with thalamic lesions. Rigidity or stiffness may be helped somewhat in 40% of patients, but the major disabling problems associated with slowness (bradykinesia) and walking do not really respond at all. Therefore, thalamotomy or thalamic stimulation are reserved for individuals with disabling tremor only. Significant complications of thalamic lesions include weakness on the opposite side of the body, slurred speech, and confusion; all complications are much worse when done on both sides of the brain.

Lesions of the internal globus pallidus (*pallidotomy* and pallidal stimulation) are useful in diminishing abnormal involuntary movements (dyskinesias) on the opposite side of the body. Patients with significant disability from dyskinesias are therefore good candidates for pallidal surgery. Patients with major disability during their off periods (when the levodopa is not working) may benefit from pallidal lesions with improvement in tremor, slowness and walking. The benefit to tremor is not nearly as good as with thalamic lesions. Interestingly, however, PD symptoms often, but not always, improve considerably on both sides of the body even if only one side of the brain is lesioned. If a patient has undergone pallidotomy on one side of the brain and does require additional surgery to relieve symptoms on the other side of the body, however, it is currently recommended that the second surgery should be DBS of the globus pallidus or subthalamic nucleus rather than a second pallidotomy. Ablative lesions on both sides of the brain are associated with far more side effects and may actually result in less improvement than single-sided lesions.

For all practical purposes, lesioning of the subthalamic nucleus is achieved with DBS only. Except in extraordinary cases, permanent lesioning is considered too risky. The advantages of this target is that lesions here benefit many more of the disabling symptoms of PD: tremor, stiffness, dyskinesias, slowness, and motor fluctuations in general. Unfortunately, complications can also be significant, and include pins and needles sensation, muscular contractions, slurred speech, and mental changes. Here, the current procedure is to do subthalamic DBS on both sides of the brain.

Transplantation procedures are all experimental and the results conflicting. They include transplanting cells from human fetuses and animal cells; in the future, genetically engineered cells in specifically-designed, controlled implantable systems may be more useful and practical than fetal cell implants. The latter has not reached human testing yet at all. All these efforts must be done within the context of an experimental protocol and considered as research rather than accepted treatment.

Finally, it should be remembered that surgery in its present state is not

a cure, nor a final therapeutic endpoint. It should be considered a late therapeutic option for appropriate candidates. As with oral medication treatment, some patients respond very well to surgical intervention, others less well; some respond for months, others for years. Nevertheless, particularly with current deep brain stimulation procedures performed with proper testing and by experienced surgeons, surgical intervention is a valuable addition to the armamentarium in the fight against the progressive symptoms of PD.

CHAPTER 4
GENETICS OF PD

Lawrence I. Golbe, MD

Most people with PD know of no relatives with the disease. Nevertheless, it appears that genetics plays an important role in the cause of PD and could provide the clues necessary to discovering prevention and/or cure of the disease.

Adult children of patients with PD should not feel selfish or contrite in asking about their risk of developing it themselves. The answer is that each child of a person with PD has about a 5% chance of developing the disease some time during life, assuming they live to 80. This compares with a risk of about 2% for the general population. If one has both a parent and a brother or sister with PD, then the risk is much higher, about 20% or 25%. About 20% of people with PD know of at least one parent, brother, sister, aunt, uncle, or grandparent with the disease. For the population without PD, the figure is about 5%.

But "family clustering," as this phenomenon is called, does not prove that PD is genetic. There is evidence that exposure to some sort of toxic substances may contribute to the cause of PD. Perhaps the tendency of PD to occur in families is the result of families sharing the same environment. This possibility is supported by surveys showing that people with PD are about three or four times more likely than those without PD to have been exposed to pesticides or herbicides.

Researchers have tried to tease apart the environmental and the genetic factors shared among family members. The best way to do this is to study twins. By far the largest twin study was published in 1998 by Caroline M. Tanner, MD of the Parkinson's Institute in Sunnyvale, CA and her colleagues. The Federal Government maintains, for purposes of medical research, a list of male twins who served in the military during World War II. This age group was perfect for a study of PD in the 1990's, as the men were old enough to be at risk of developing PD, but young enough that most were still living and able to participate in the research project. Dr. Tanner and a large team of collaborators contacted all the men, asking if they had signs of PD. For each pair in which at least one member answered in the affirmative, both members were examined by neurologists.

The statistical analysis compared identical twin pairs with fraternal twin pairs with regard to the "concordance rate," which is the percentage of pairs in which both members proved to have PD. For a purely genetic disorder that appears early in life, one would expect 100% concordance among the identical twins and 50% among the fraternal twins. For a purely genetic disorder that appears at various times in middle or late life, as PD does, there will be far less than 100% concordance among the identical twin pairs, but their concordance rate will still be twice the rate for the fraternal twin pairs.

The result of Dr. Tanner's study was that 16% of the identical twin pairs and 11% of the fraternal twin pairs were concordant for PD. That these two figures are not very different appears to contradict the genetic theory of the cause of PD. But among those few twin pairs in which the first (or only) member to develop PD did so before age 51, all four identical pairs and two of 12 fraternal pairs were concordant, evidence that younger-onset PD is strongly genetic. Overall, though, this study was widely interpreted as strong evidence against a genetic cause of PD.

But as well-designed as this twin study was, it suffered one important flaw. It relied on neurologists' usual methods of diagnosing PD-listening to symptoms and examining the patient. We now know that the beginnings of PD in the brain occur several years before the symptoms appear. Could it have been possible that more of the twins would have been found concordant if there were some way of detecting mild stages of degeneration of dopamine-producing brain cells? Furthermore, is the difference between the younger-onset and older-onset twins the result of having had more time for PD to develop in the second twin in younger-onset pairs?

There is a way of detecting "presymptomatic" PD, but it's expensive and unwieldy. Positron emission tomography (PET) creates an image of the brain that looks like a blurry CT scan or MRI. But rather than showing the structure of the brain, it shows a map of where a radioactive chemical, injected intravenously, is taken up in the brain. For this and most PET studies of PD, that radioactive chemical, called fluorodopa, is a form of levodopa, a component of Sinemet and the most effective drug for PD. This is taken up by the same cells in the basal ganglia that degenerate in PD. A PET scan showing subnormal uptake of radioactive fluorodopa is evidence of PD.

In 1999, Dr. Paula Piccini and colleagues, from the PET scan group headed by Dr. David Brooks in London, published their PD twin study. They scanned the healthy-appearing members of 18 twin pairs in which one member had clear outward signs of PD and repeated the scans an average of four years later. By the end of the study, they found a 72% rate of concordance among identical twins by PET scan and only an 18% concordance rate among fraternal twins. This study is very strong evidence for a genetic cause of PD, although some scientists don't trust the reliability of PET scanning enough to draw very strong conclusions from this study alone.

Another lesson from the twin studies is that the age at which PD symptoms begin can vary widely even between individuals with identical genes. This may help explain why most people with PD know of no similarly affected relatives. The relatives who would have developed PD may not have lived to the age when the outward signs of the disease would have appeared.

If PD is strongly genetic, what are the specific genes involved? The answer to this question could not only permit diagnostic testing, but more importantly could point scientists toward a chemical pathway in the brain that could be manipulated to provide a cure for PD.

The easiest way to find specific genes contributing to the cause of a disease like PD is to take an educated guess as to a "candidate gene." For

example, knowing that dopamine loss is involved in PD makes candidates of those genes that carry instructions for processing dopamine. The researcher tests for the presence of a variation in this gene a group of typically 50-100 people with PD and a similar group without PD. The variation itself is usually not the disease-causing defect, but is merely a "marker," or a signpost on the gene. If that marker is more frequent among people with PD than without, then there may be a disease-causing defect close to the marker, probably within the same gene.

So far, a handful of genes have given positive results with PD in such "association studies." No one of these alone would be the cause of the disease in general. These proteins that these genes code for either break down toxins, are involved in dopamine transmission between brain cells, or maintain the skeletal structure of brain cells. Because association studies can produce falsely positive results, some genes on the present list may drop off with future research. Others will definitely be added in the future as new candidate genes are tested. Perhaps in the next few years, enough genes will become associated with PD that doctors will be able routinely to order a "PD genetics battery" to identify healthy people who should receive a PD prevention that, we hope, will come along soon.

Another way to identify PD genes is by finding unusual families with large numbers of people with PD. Blood samples from about 10 or 11 family members with PD are typically needed to find the causative gene in this way. This has been accomplished so far for only two genes: α *-synuclein* and *parkin*. The first was found in a large Italian-American family, the Contursi kindred, named after the town in Italy where they originated. In that family, 60 people have had PD in the past five generations. They proved to harbor a defect, or mutation, in the gene for α -synuclein, a protein that is involved somehow in preparing packets of dopamine for transmission to other brain cells. Once this gene defect was identified, it was easy to test for its presence in people outside the Contursi kindred. It proved to be present in a few Greek families with PD, but in no one else with PD. A German family, however, proved to have a different defect in the same gene.

Since the identification of an α -synuclein gene defect in the Contursi kindred, it was discovered that a problem with the α -synuclein protein is central to all PD: the pathologic hallmark in the brains of patients with PD, seen under the microscope and called a *Lewy body*, was found in 1998 to be made up primarily of α -synuclein. But in over 99% of patients, the problem isn't in the gene for α -synuclein, but in how that a-synuclein is processed after it is manufactured. So, the discovery of a gene defect peculiar to a few families has provided a valuable clue to the cause of PD in general. Perhaps soon a way will be found to make the α -synuclein protein function properly, and this may constitute a prevention or part of a cure for PD.

Other families are being analyzed in hopes of elucidating similarly valuable clues to the cause of PD in general. The other gene for which a specific defect has been found, the "parkin" gene, is involved in an unusual young-adult-onset form of PD that was first observed in Japan (which the Japanese, as mentioned above, call "juvenile parkinsonism"). Since then, it

has been found in milder form in many other countries around the world. This form differs from typical PD not only in its age at onset and its familial pattern, but very importantly, in the absence of Lewy bodies in the brain cells. Lewy bodies had been considered necessary to the diagnosis of PD. But the parkin discovery is making us rethink that. Subtler defects in the parkin gene than were present in the Japanese families may contribute to the cause of a significant minority of cases of PD worldwide. This, like the role of α -synuclein, is presently an area of intensive research.

PD genes have been sought in several other families with multiple members with PD. In some, the general location of the causative gene has been identified-that is, which of the 23 chromosomes and in which segment of the chromosome the gene is present. But the exact defect in the genetic code has not been worked out. This will surely change over the next few years. When we have that information, we will then be able to test for those defects in other people with PD and their role in the cause of PD overall can be determined.

Another powerful method of identifying genes causing PD, but one that has not yet succeeded, is a "sibling pair" study. Collecting blood samples from 400 pairs of brothers and/or sisters who both have PD permits a "total genome search." This is where genetic markers covering all relevant regions of all the chromosomes are examined systematically for associations with PD. Then, the precise genetic code defect in incriminated regions would be worked out by other means. There are three sibling studies presently collecting samples in the US and one in Europe. People with PD who have a living sibling with, or possibly with, PD should contact the APDA to be referred for participation in one of these studies, which would require only an examination, a questionnaire, and a blood sample.

It will probably turn out that PD is the result of multiple genetic defects present in the same individual. Because these defects will inevitably occur on different chromosomes, they will not be inherited together, accounting for the low probability of passing on the disease to one's children. To make things more complicated, another cell component, the mitochondria, which act as the "lungs" of the cell to use oxygen and produce energy, also contain genes that are separate from the chromosomes; identification of abnormalities in these genes are also under investigation. An exposure to some chemical may also turn out to be a factor. Some researchers feel that there will be so many possible genetic defects that may contribute to the cause of PD that targeting any one or two of them for prevention or cure of the disease would be fruitless. They, rather, feel that all of the genetic defects ultimately produce only one or two modes of brain cell death, and that this process should be studied with an eye to interrupting it. In any case, the first decade of the 21st Century will surely bring major advances in our ability to predict, diagnose, prevent, and possibly even cure PD.

CHAPTER 5
EXERCISE AND PD

Gertrudis G. Sanidad, MHA, PT

INTRODUCTION

Recent studies have found that exercise and early intervention by a physical therapist will greatly improve your ability to perform certain functional activities, thus enabling you to retain your independence longer. Although PD is progressive in nature, the effects of the disease and secondary complications, such as joint deformities, can be minimized or delayed. Starting an exercise routine during the early stages of the disease will prove to be more beneficial than later when you are older and when it may be difficult to perform the exercises due to weakness or deformities.

WHY DO YOU NEED TO EXERCISE?

Exercise will help you achieve the following goals:

1. Maintain or increase mobility in your joints.
2. Reduce rigidity and joint stiffness.
3. Prevent joint deformities.
4. Improve posture.
5. Increase strength.
6. Prevent muscle weakness and wasting.
7. Improve ability to walk.
8. Maintain or improve functional ability.
9. Improve endurance.
10. Improve quality of life.
11. Promote relaxation.
12. Improve circulation.
13. Enhance mood.
14. Improve general well-being.

WHAT EXERCISES CAN YOU DO?

You should ask your physician before you start any exercise program. A physical therapist can plan an individualized exercise program designed specifically for your needs. You can, however, do some basic exercises on your own which can be a combination of the following:

1. Relaxation exercises

What is the purpose of relaxation exercises?

¥These exercises will decrease tension in the muscles and enhance the ability to perform stretching exercises.

What relaxation exercises can you do?

¥ Start with deep breathing exercises:

Sitting or lying down on your back, place your hands on your abdomen and take a deep breath, feeling your hands move out. Exhale fully and feel your hands move in.

¥ Gentle, rocking, and rhythmic motions for generalized relaxation, as in the use of a rocking chair. Slow, rhythmic rolling on a mat may also be done.

¥ Relaxation of lower trunk muscles:

Lay on the back on a flat surface. Bend knees with feet flat on surface. Roll your knees from side to side as far as possible.

¥ Relaxation of upper trunk muscles:

Lay on the back on a flat surface. Slowly rotate your chest and shoulder to the opposite side, holding hips still. Return to center. Repeat to the other side.

¥ Relaxation of neck muscles:

Sitting, turn your head slowly side to side, looking over each shoulder.

¥ Relaxation of shoulder muscles:

Sitting, roll your shoulders forward, making small circles. Then roll your shoulders backward.

¥**Yoga** is also an effective and useful relaxation technique. It promotes relaxation through the combination of deep breathing and slow, steady stretching.

2. Stretching exercises

Why are stretching exercises important?

¥They maintain full range of motion. Mobility and flexibility of the skin, muscles and connective tissue around a joint are necessary for the joint to have full range of motion.

¥They help to maintain the flexibility of the muscles to enable them to work more efficiently. When a muscle loses its flexibility, it shortens and loses strength.

¥They assist in preventing contractions (adaptive shortening) of the soft tissues around a joint, resulting in limitation of joint mobility. Contractions are fixed positions of joints and limbs (essentially getting "stuck"), and cannot be overcome with passive movements.

¥They work against the effects of progressive rigidity.

What stretching exercises can you do?

The following are some exercises you can incorporate in your daily routine:

¥ Full Body Stretch: Lie on your back on a hard surface with body in X form (arms overhead spread as far apart as possible and legs spread apart as far as possible). Inhaling, gently reach for four corners. Exhale and relax.

¥ Head Turns: Sitting, turn head to right as far as possible Return to mid-line. Repeat to left.

¥ Head Tilts: Sitting, bend head to the right, bringing ear towards shoulder. Return to midline. Repeat to left.

¥ Chin Tucks: Sitting, gently pull chin as far as possible.

¥ Shoulder Shrugs: Sitting, bring your shoulders up towards your ears.

¥ Shoulder Squeezes: Sitting, squeeze shoulder blades together, bringing elbows behind you.

¥ Shoulder Circles: Sitting, roll shoulders forward, making small circles. Then roll shoulders back.

¥ Shoulder Stretch: Sitting, place right hand, palm down, on right shoulder blade. Reach behind your back with the left hand, palm up. Try to touch finger tips of the other hand.

¥ Wrist Circles: Make slow circles with your wrists.

¥ Finger to Thumb Pinches: Bring each finger to thumb, making a circle. Start with the index and thumb, progressing to the small finger and thumb.

¥ Trunk Twists: Sitting, place hands on shoulders and gently twist head, neck, and trunk to the right as far as possible. Repeat to left.

¥ Hamstring Stretch: Sitting, place your right foot on a stool or low chair in front of you. Keeping the right knee and your back straight, gently lean forward until stretch is felt at the back of the right leg. Hold for a few seconds. Repeat with the left leg.

¥ Calf Stretch: Standing, holding on to a steady surface for support. Place your right foot forward and bend the right knee, keeping the left heel flat on the floor. You should feel the stretch at the left calf muscles.

Are there any pointers you need to remember when performing stretching exercises?

Yes, keep the following pointers in mind when performing stretching exercises:

¥ Warm up before you stretch to prevent injuries to your muscles and to enable your muscles to perform optimally. You can warm up by either:

1. Taking a warm bath
2. Walking briskly in place for 10 minutes

¥ Stretch to the point of mild discomfort. Hold the stretch for 10 seconds and slowly release. If you experience joint pain or muscle soreness lasting more than 24 hours, you used too much force when stretching.

¥ Inhale at the beginning of the exercise. Exhale while you hold the stretch. Do not hold your breath at any time during the exercise.

¥ Do not stretch when joints are red or swollen.

¥ Do not bounce when you stretch as this can hurt your muscles.

¥ Wear loose clothing.

3. Strengthening exercises

¥ A physical therapist can assist you to establish an individualized strengthening exercise program to target specific weak muscles.

¥ Particular attention must be given to strengthen muscles that maintain proper posture:

- ✥ Neck extensor muscles
- ✥ Back extensor muscles

- ✦ Front thigh muscles
- ✦ Buttocks muscles
- ✦ Shin muscles

Why is maintaining proper posture important?

¥ Maintaining and improving the flexibility of the spine have been shown to improve the physical performance of patients with PD.

¥ Patients with PD have a tendency to developing poor posture, typically a flexed or stooped posture, because of muscle weakness. The flexor muscles (generally, muscles that bend a joint) become stronger than the extensor muscles (generally, muscles that straighten a joint).

¥ Breathing is effortless and easy and muscle energy is expended minimally.

¥ The joints are in good balance. The muscles, bones, and joints are protected from trauma or deformity.

What are some simple strengthening exercises you can do?

¥ Chin Tucks

¥ Pelvic Tilt: Lying, on your back with both knees bent. Tighten stomach and buttocks and press lower back down to surface upon which you are lying.

¥ Prone Press-ups: Lie on stomach with palms by shoulders. Slowly push top half of your body upward using your arms.

¥ Gluteal sets: Recline on your back, supported by your elbows. Keep both legs straight. Squeeze your buttocks together as tightly as possible. Hold for five seconds and relax.

¥ Straight Leg Raise: Lying on your back, with the left leg bent and the right leg straight. Lift the right leg off the surface, keeping your knee straight. Hold for five seconds and relax. Repeat with the left leg.

¥ Knee Lifts: Sitting, with back supported. Straighten right leg. Repeat with the left leg.

¥ Toe Raises: Standing, holding on to a steady surface. Bring the front of the right foot up, keeping the heel on the floor and the knee straight. Repeat with the other foot.

¥ Step Exercises: Using stair or stool, step up with the right foot, then step down with the right foot. Repeat with the left foot.

¥ Face Exercises:

- ✦ Pucker and smile.
- ✦ Open mouth as wide as possible, then close lips as tightly as possible.
- ✦ Raise eyebrows as high as possible, then close eyes as tightly as possible.
- ✦ Stick tongue out as far as possible and move it form side to side.

4. Aerobic exercises

What are aerobic exercises and why are they beneficial?

Aerobic exercises are activities that increase the oxygen available to the body. During exercise, the body's energy requirements increase, which means that the heart has to work harder to pump the blood to meet the increased need for oxygen and other nutrients. They are long in duration and low in intensity.

Aerobic exercises will improve your cardiovascular and muscular fitness and decrease the deconditioning effects of PD.

Aerobic exercises will also increase your endurance. Fatigue is a common complaint of patients with PD, especially in the middle and late stages.

Aerobic exercises you can do.

¥ **Walking:** Conditions your heart and lungs; easy, inexpensive, safe and accessible.

¥ **Swimming:** Good endurance exercise; buoyancy of the water reduces stress on your joints and helps you move your joints through their full range and strengthen your muscles.

¥ **Bicycling:** Also good for cardiovascular fitness; promotes reciprocal fluid movement.

¥ **Dancing:** Promotes trunk rotation and balance.

¥ **T'ai Chi:** Promotes movement control; coordinates breathing, flexibility and balance.

WHAT IS A GOOD EXERCISE PROGRAM?

A good program combines activities that improve flexibility, strength and endurance. You must remember, however, that the only way you will stick to your exercise program is if you enjoy it. So, combine activities that you enjoy and that you can work in your daily routine. You can also attend exercise classes if you want. Remember also, especially for early, mild patients who have been active and healthy prior to their diagnosis of PD, that you may continue to participate in rigorous exercise (for example, tennis or skiing) if you have been doing it all along, and as long as you and your doctor feel you can. A diagnosis of PD is *not* a reason to *stop* doing sports you can do and enjoy doing!

WHAT IS THE WARM-UP AND COOL-DOWN PERIOD?

The warm-up and cool-down periods are essential parts in any exercise routine. They assist the body in adjusting from a period of rest to activity and vice-versa.

Warm-up periods can be done for 5 to 10 minutes at the start of the exercise program. This can be done by walking briskly, swinging the arms or jogging in place.

Cool-down periods are done at the end of the exercise. You can use the same activities you did for warm-up.

Exercise when your medication is "working" ("on").

HOW OFTEN SHOULD YOU EXERCISE?

You can exercise for 30 minutes or more, three to four times a week. You can start with just a few minutes, gradually increasing to that goal. You can also do two 15-minute sessions each day if the 30 minutes is too intense for you.

HOW DO YOU KNOW THAT YOU ARE OVER-EXERCISING?

There are a few ways you can monitor the intensity of your exercise routine:

1. Maximum Heart Rate

   Take your pulse by placing the tips of your index, middle and ring fingers at your wrist below the base of your thumb or at the carotid artery on the neck. Count how many beats you feel in 15 seconds and multiply by 4. This is your resting heart rate.

To determine your maximum heart rate, subtract your age from 220. You should stop exercising if your heart rate is over this number.

2. Borg Scale

This is the Borg Scale of Rating of Perceived Exertion (RPE). You will rate how hard you are working on a scale of 0 to 10; zero is doing no work at all and 10 is working very hard that you could only do it for a few seconds. The table below describes the Borg Scale. A good level for an aerobic exercise program is between 3 and 6 on the scale

0	Nothing at all
0.5	Very, very weak
1	Very Weak
2	Weak
3	Moderate
4	Somewhat strong
5	Strong
6	
7	Very strong
8	
9	
10	Very, very strong Maximal

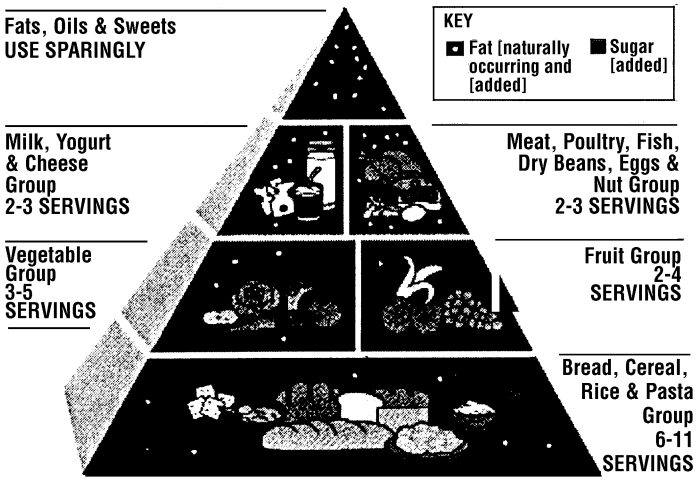
CHAPTER 6
NUTRITION AND PD

Julie H. Glass, RD, CNSD

General Nutrition

A well-balanced diet is the key to proper and adequate nutrition for all individuals throughout their lifetime. What is considered a well-balanced diet? The Food Guide Pyramid was created to help people determine which foods to eat and how often they should eat them to meet their basic nutrition needs. The general recommendations are the same for those with PD. By following the pyramid and eating the recommended servings from each group, most individuals will meet their calorie, protein, vitamin, and mineral needs.

FOOD GUIDE PYRAMID
A Guide to Daily Food Choices



Source: U.S. Department of Agriculture and the U.S. Department of Health and Human Services

Is a multivitamin necessary? Most people should be able to meet their vitamin and mineral needs by eating a balanced diet. If, however, you feel that you are not able to eat the recommended number of servings from each group, a standard multivitamin may be beneficial. The multivitamin should provide close to 100% of the daily recommended allowance (RDA) of most vitamins and minerals. For those individuals taking levodopa, do not take an extra vitamin B_6 supplement in addition to a multivitamin. This vitamin in very

large doses can reverse the action of levodopa. In the preparation with carbidopa (as in Sinemet), however, this is not really a concern, since the carbidopa protects levodopa from the effects of vitamin B_6.

Preventing Constipation

Constipation is a common side effect of PD and PD medications. Although a result of the disease process, constipation may also be caused by an inadequate intake of fiber containing foods, insufficient fluid intake, and/or decreased activity. Several helpful hints for constipation are listed below. Aim to consume, at a minimum, approximately 25 grams of fiber per day. Even if you do not suffer from this uncomfortable condition, it may be helpful to incorporate some of these principles for both overall bowel health and for prevention of constipation.

To prevent constipation or to relieve it, try some of these useful ideas:

- ¥ Consume at least 2-3 servings of fruit a day, preferably raw with the skins and seeds.
- ¥ Aim for 2-3 servings of vegetables per day. Try to have at least one serving of a raw vegetable. Include the skin whenever possible; i.e., baked potatoes. Vegetables with seeds will provide slightly more fiber.
- ¥ Choose whole grain bread and cereal products over those made with refined, highly milled flour. When baking, try using one part whole wheat flour and one part all purpose flour. Also, add wheat germ to your baked goods or sprinkled on top of yogurt or cereal for additional fiber.
- ¥ Include more dried beans and legumes in your daily diet.
- ¥ Snack on high fiber snacks such as nuts, seeds, popcorn, and dried fruit.
- ¥ Include prunes or prune juice in your diet.
- ¥ Make sure you are drinking adequate fluids. Drink at least eight glasses of fluid each day.
- ¥ Try to get some form of exercise daily.
- ¥ If you have difficulty chewing or swallowing and constipation is a problem, try using a fiber supplement that can be mixed with juice or water, such as Metamucil¤. If you need extra calories and protein, try a supplement such as Ensure¤ Fiber.
- ¥ Do not ignore the urge to have a bowel movement. Allow yourself time to relax following a meal where you have easy access to a bathroom.

Sidebox for Constipation:

An "all natural" recipe (i.e., one that is made from ingredients from the supermarket rather than the pharmacy) that many patients find helpful is:

2 parts wheat bran, unprocessed
1 part apple sauce
1 part prune juice
Take 2 tablespoons once or twice daily.
Refrigerate remainder.

Preventing Weight Loss

Weight loss is a frequent side effect of PD. Contributing factors include loss of appetite, decreased interest in food, and dyskinesias (abnormal involuntary movements from medication fluctuations). In more advanced PD, weight loss is further complicated by difficulty in feeding oneself and difficulty swallowing (dysphagia). Weight loss due to inadequate intake consists of both fat and lean body mass (muscle). Not eating adequate calories to maintain your weight will make you feel tired and will put you at risk of developing malnutrition. It is important to keep an eye on your weight by either weighing yourself on a scale or by paying attention to how your clothes are fitting. If you notice weight loss, whether it be a gradual decline or a sudden drop, you should bring it to the attention of your physician and dietitian so that they can help you prevent further weight loss.

If weight loss is a problem for you or your loved one, try following some of these calorie-boosting suggestions:

- ¥ Add butter or margarine to hot foods such as soups, vegetables, mashed potatoes, cooked cereal, pasta, and rice. A teaspoon will add 45 calories. Serve bread hot-more butter is used when it melts into it.
- ¥ Increase calories in food by using gravies, cream or cheese sauces, salad dressing, or mayonnaise on meats, vegetables, and starches.
- ¥ Try light or heavy cream in place of milk when baking, making soups, or making desserts such as pudding or custard.
- ¥ Use milk in place of water in hot cereals and instant beverages such as hot chocolate. Increase the protein content by adding 2-4 tablespoons of powdered milk per cup of milk.
- ¥ Add whipped cream generously to pies, fruit, puddings, hot chocolate, Jell-O, and other desserts.
- ¥ Powdered coffee creamers add calories without volume-add them to gravy, soup, milk shakes, and hot cereals.
- ¥ Add extra calories to food by using sugar, jelly, jam, honey, molasses, or maple syrup.
- ¥ Drink fluids with calories and nutrients such as juice and milk. Water, black tea, and black coffee have no calories; regular soda is high in calories but is lacking in all nutrients.
- ¥ Try adding canned dried beans to meals; they are high in calories, protein, and fiber. You can puree them with your favorite seasoning to make spreads for crackers or sandwiches, or add them to soup and pasta dishes to boost the nutritional content.

¥ Carry convenience foods with you such as canned supplements (e.g. Carnation Instant Breakfast¤, Ensure¤, Boost¤, Resource¤), yogurt, pudding, applesauce, or instant soup packets so you never miss a meal.
¥ Help stimulate the appetite by preparing the meals and setting the table in the most visually appealing manner possible. Use garnish to accent the foods.
¥ Eat three meals with three snacks each day.
¥ When preparing soft or pureed foods, remember to season them as you would any other meal.

Is there special diet for PD?

A diet commonly known as the "PD Diet" is the result of both observation and research examining the effects of dietary protein intake on patients taking levodopa. A high protein meal following levodopa intake has been shown to have an effect on motor function. Protein digestion begins in the stomach with the small intestine being the primary site of absorption. As protein passes through the stomach into the small intestine, it is broken down into amino acids. Levodopa is a type of modified amino acid that is also absorbed in the small intestine. The amino acids are absorbed into the bloodstream and are carried to the brain. At any given point, only a certain number of amino acids may enter into the brain, leading to competition for absorption between levodopa and the amino acids from dietary protein. This competition can create variable levels of absorption, which may cause fluctuations in motor function.

The typical American diet contains more than the RDA for protein, which is 0.8 grams per kilogram of body weight (0.36 grams per pound). When modifying the diet to limit motor fluctuations, the dietary protein intake is adjusted to the RDA, 0.8 grams per kilogram. The diet is also organized with the majority of the protein intake during the evening meal and limited protein intake during the day. Several factors should be considered prior to changing your diet, such as severity of your disease, nutritional status, and level of activity. Should you be interested in trying a moderate protein diet, speak with your physician and dietitian for the best method of incorporating it into your lifestyle. Remember, however, that patients not yet on levodopa, and individuals with early, mild, stable PD need not be overly concerned with levodopa competition with protein. Good nutrition is paramount and the "PD Diet" should be reserved only for those patients with significant fluctuations and protein effects on their disease.

Other nutritional considerations with levodopa

As discussed, levodopa is absorbed in the small intestine; therefore, any factors that slow the passage of food from the stomach into the intestine

will have an effect on the consistency of levodopa absorption. Foods that slow the rate of digestion include those that are high in fiber and fat. In an effort to reduce fluctuations in motor function, it is recommended that you limit the fat and fiber content in your meals that are consumed after taking levodopa. Adequate fiber intake is very important for those with PD as discussed earlier; therefore, plan your meals or snacks that are high in fiber away from the times when you take your medication.

Nutrition is one of the factors that requires consideration in PD. Figuring how and when to plan your meals will take time and effort; therefore, refer to both your physician and dietitian for assistance on the best plan of action for you.

CHAPTER 7
SPEECH AND SWALLOWING PROBLEMS ASSOCIATED WITH PD

Kate Towlen, MS, CCC-SLP
&
Christine Bove, MA, CCC-SLP

PD is a progressive neurological disorder that causes stiffness or slowness in muscles. Speaking and/or swallowing difficulties may develop because the muscles of the throat, mouth and muscles that aid in breathing can be affected. These problems usually occur in more advanced PD, but occasionally may be encountered earlier. Awareness of the issues and their solutions is the best way to deal with them.

Swallowing Disorders Associated With PD

Do I have a swallowing disorder?

PD is a type of movement disorder that makes it difficult to initiate muscle movement. Since swallowing foods and liquids requires muscle use, swallowing problems are associated with PD. Many patients have difficulty swallowing foods, liquids, saliva and/or pills. A swallowing disorder is called *dysphagia*. These difficulties may occur because the movement of the throat muscles becomes weak. Some patients are not aware that they are having swallowing difficulties. Weight loss is often another result of dysphagia.

Swallowing difficulties can occur in various phases of eating. Difficulties may include reduced lip movement, increased fatigue when chewing, limited tongue movement, food sticking to the top of the mouth, forward/backward tongue movement or "pumping," a sensation of food sticking in the middle and/or sides of the throat and sensations of food sticking in the upper middle chest area. Some people experience coughing, choking, throat clearing, a change in the sound of their voice or shortness of breath during meals. If any of these symptoms are frequently happening when eating or drinking, the patient should inform his/her physician. A referral can then be made to a speech pathologist for a swallowing evaluation and a dietitian for nutritional needs.

How can a speech pathologist help me?

A speech pathologist is a person who can diagnose and treat people with swallowing disorders. The speech pathologist will examine the lips,

tongue and throat. He or she will observe the patient eating and drinking and look for signs of swallowing problems. The speech pathologist may recommend changing the texture or consistency of the food or liquids to make swallowing easier (regular, soft or puree/smooth foods). Patients maybe told to hold their head in different positions when eating or drinking to improve their swallowing. If needed, an X-ray swallowing examination may be performed to determine if food or liquid is being swallowed safely. Commonly, this test is called a *modified barium swallow study*. This test will also help to determine which food consistency and swallowing strategies are needed for the patient to eat safely. A condition called *aspiration* can also be diagnosed using this test. Aspiration is when food or liquid is directed into the airway instead of the stomach. Aspiration can lead to pneumonia and other breathing problems.

If appropriate, in severe cases, a feeding tube may be discussed by the speech pathologist and physician with the patient. This tube may be needed if eating is not safe or the patient needs to gain more weight. Feeding tubes may be placed for short term or long term needs. Usually a tube is placed in the nose (called a nasogastric tube) for short term feeding problems. A tube may be placed into the stomach (called *percutaneous endoscopy gastrostomy* and/or *gastrostomy*) for those people requiring long term nutrition. A stomach-feeding tube is shaped like a button and easily covered with clothing. It is important to note that a person can still eat by mouth despite having a feeding tube in the nose or stomach.

Suggestions to improve swallowing problems:

Some strategies to improve swallowing will require training from a Speech Pathologist; however, a few suggestions are provided below: adapted from the American Parkinson Disease Association, Inc. "Speech Problems and Swallowing problems in Parkinson's Disease."

If you feel that food is collecting in your mouth after you have already swallowed:

- Keep your lips closed, and teeth together. Once the food is on your tongue, lift the tongue up, then move the food back and swallow. Think "up-back-swallow" for each bite/sip.

- Eat and drink slowly. Take small bites and chew well.

- Chew hard and move the food around your mouth with your tongue. Chew on one side of the mouth and then the other.

- Do not put more food into your mouth until you have swallowed the previous mouthful.

Other suggestions include:

- The patient and family should have knowledge of the Heimlich maneuver.

- Talk to your doctor about taking your medications near mealtimes to improve mouth and throat muscle movement.

Suggestions for saliva control:

Many people with PD complain of excessive saliva build up in their mouths. Normally, saliva is automatically swallowed. PD causes the muscles in the mouth and throat to work slower which lets saliva collect in the mouth. Sometimes, this saliva will spill or drool from their mouth. Often, patients will cough on this saliva. Extra saliva can also make speech less clear. The amount of saliva may change from time to time. In particular cases, physicians may prescribe a type of medication to decrease drooling. This type of medication is referred to as an *anticholinergic drug* and may cause dry mouth, nausea, vomiting, and worsen swallowing problems. The following is a list of temporary methods to alleviate the discomfort.

- Make a conscious effort to swallow your saliva often. Think, "up-back-swallow."

- Try to keep your head in an upright position. Saliva will collect in the back of your throat and cause an automatic swallow.

- Swallow any excess saliva before you begin talking.

- Drink papaya juice to reduce the thickness of your saliva.

- Sleep with two pillows under your head or raise the head of the bed to reduce collection of saliva in the throat.

Suggestions for relieving dry mouth:

People with PD may experience a very dry mouth (called xerostomia). which may be caused by certain medications. The following is a list of temporary methods will help reduce dryness in the mouth:

- Use a humidifier in main room/ bedroom

- Clean your mouth with lemon glycerin swabs, (which are available at most pharmacies)

- Use synthetic saliva drops which will help produce saliva

- Eat Lemon/Italian ice

- Drink more water and other non-caffeinated beverages
- Suck on a lollipop or piece of hard candy, if approved by your healthcare team.

- Apply petroleum jelly (i.e., Vaseline) to the outside of your teeth and lips

Speech Problems Associated With Parkinson's Disease

Approximately 75% of people with PD develop problems with speech production and/or voice quality. This is due to problems coordinating the muscles that control breathing, voice and speaking. It may be hard to control the rate and rhythm of speech, as the muscles become stiff. The most common difficulties that a person with PD experiences are:

- Softer voice
- Hoarse voice
- Slurred speech
- Vocal tremor or unsteady voice
- Short rushes of speech
- Monotone voice

What can be done to help?

Speech or voice therapy, with a licensed speech-language pathologist, can be very effective in treating the above mentioned areas of difficulty. The overall goal of speech therapy is to improve functional communication. A speech pathologist will evaluate speech/voice production and provide exercises to increase volume, reduce hoarseness and improve overall speech production. Good breathing techniques and exercises to improve breath support will also be provided to improve your speech.

One type of voice therapy is the "Lee Silverman Voice Treatment" approach, which was developed by Lorraine Olson-Ramig PhD, CCC-SLP and Carolyn Mead MA, CCC-SLP. This is an intensive form of voice therapy (i.e., 16 sessions in one month) which focuses on increasing vocal effort. Therapy consists of vocal exercises, which improves both speech production and vocal quality.

Alternative forms of communication, such as augmentative devices or voice amplification devices, are also available. A speech pathologist will help decide which device is best for your individual needs.

Helpful hints to improve communication:

Some of these strategies will require training from a speech pathologist.

Adapted from The American Parkinson Disease Association, Inc. "Speech Problems and Swallowing Problems in Parkinson's Disease."

- Speaking clearly requires conscious effort.

- Take a breath before you speak, try to maintain good posture while speaking and pause between every few words.

- Try to use a louder, more projected voice. Encourage your family and friends to remind you to use a louder voice.

- Use short phrases. Long and complicated sentences may be difficult for your listener to understand.

- Always face your listener. It will be easier for him/her to understand your speech.

- Talk for yourself. Try not to let others do the talking for you.

How can I get help for my speech or swallowing problems?

1. Talk to your physician. Your doctor will give you a referral and other information that your insurance company may need for a speech and/or swallowing evaluation.

2. Call local hospitals, outpatient facilities or home care departments to make an appointment for a speech and/or swallowing evaluation.

3. You may contact the American Speech-Language-Hearing Association (ASHA) to find a speech pathologist in your area. ASHA can be reached by calling 1-800-498-2071.

4. You may contact the Lee Silverman Voice Treatment Foundation (LSVT) at 1-800-606-5788 for a listing of local LSVT-certified clinicians.

A speech pathologist is one of the many professionals involved in the overall care of an individual with PD. Speech and swallowing therapy can be very effective in improving or managing speech and swallowing problems. Recent research and education in the field of speech pathology has allowed for continuous changes and new developments in the management of speech and swallowing disorders associated with PD.

CHAPTER 8
PSYCHOSOCIAL AND PSYCHIATRIC ASPECTS OF PD

Kenneth R. Kaufman, MD MRCPsych

The existence of any illness, no matter the duration or severity, if it causes any degree of incapacity, results in associated psychosocial changes. In addition, the illness process and/or the medications used to treat the illness may be the reasons for the development of different psychiatric features. Such is clearly the case with PD.

Psychosocial Issues:

1. PD affects motor functioning that is visible not only to the patient but to the patient's family, co-workers, and others resulting in a changed attitude towards self or a real or perceived change in attitude by others.
2. The need to take medications for PD and the physical limitations secondary to PD may make the patient feel dependent, an issue of significant importance to those individuals who have never had a serious illness in their lives or who have never felt dependent upon others before.
3. The degree of motor impairment may preclude normal activities if tremor, stiffness, and slowness or the dyskinetic movements are not adequately controlled.

Primary Effects of PD:

4. PD patients may present initially with depression or anxiety, including full-blown panic attacks, as a chief complaint, with PD motor symptoms only becoming apparent later.
5. As PD progresses, depression or anxiety may be a prominent component of the illness because of biochemical factors, not just psychosocial factors. In late stages of PD in older individuals, significant cognitive dysfunction or even dementia may develop, with decreased memory, confusion, dementia, hallucinations, and delusions.

Medication Side Effects:

6. Medications used to treat PD, primarily in older patients with advanced disease, may result in psychiatric symptoms such as confusion, hallucinations, delusions, and paranoia.

Psychosocial Issues: Understanding and Acceptance

A first step is understanding and accepting the illness process: initial symptoms, progression, neurologic treatment, long-term consequences, and commitment to compliance with supportive family interventions. PD is a neu-

rologic illness that usually begins with minor motor movement abnormalities (for example, tremors), progresses to further motor impairment (slowness and stiffness), and can lead in more advanced stages to more disabling signs and symptoms requiring significant support systems. It is important to realize that the rate of progression is usually quite slow and in some patients may not progress into the most severe stages. Young-onset patients, in fact, tend to have a very long durationf of PD and do well for a long time. Despite the likely genetic contribution, the chance of passing on PD to the next generation is very low, and one should not worry about this. PD should be viewed as any other medical illness that can be treated, much like high blood pressure or diabetes. As with many chronic medical illnesses, a degree of denial and anger at the diagnosis may be present, but should be followed by acceptance, since the illness can be effectively treated; and despite long-term complications of therapy, multiple modern treatment options mean that the symptoms of PD can be controlled throughout the course of the condition.

PD should be viewed from a positive perspective ("I have been diagnosed so let's get it treated and continue on with my life"). Initially few complications from PD or the early therapeutic interventions occur. As disease progresses, long-term complications such as motor fluctuations, dyskinesias, on-off phenomena, and gait/ balance/ speech problems may need to be addressed. Even during these stages, medication can be effectively used to control symptoms and to maintain the patient in the work force with appropriate social interactions.

Depending on one's life and career situation, an initial mild rest tremor may have no work or social issues for one patient, while for another, such tremor could be disabling. For example, airline pilots would not receive FAA clearance in such circumstances; hand craftsmen might not be able to complete projects; the handwriting of calligraphers would not meet professional standards. But the vast majority of workers would be able to continue in their professions, usually with only minor concessions to adjustments at one's job. Psychosocial issues that individuals must consider include what patients think of themselves once they have been diagnosed with PD and how they believe that others regard them.

If family, friends, co-workers, or others are uncomfortable with a mild tremor or even more exaggerated movements, then it is wisest to discuss the illness process in an open manner. Not being honest regarding the condition can result in misperceptions by others. Most people, including at the work site, are very understanding regarding this condition. It is especially important to address work issues, since PD may affect safety in certain jobs. If employers, co-workers, or even family remain uncomfortable after your explanations, then it is sadly *their problem and not yours*. Often, people question if they can be good or successful if they have this condition. Patients should appreciate that no stigma is attached to PD, that many well known people have this illness and function at a high level (including the US Attorney General and the

Pope), and finally, that who and what you are is not defined by illness but rather by character. In other words, you ***have*** PD, you ***are not*** PD.

Doctor, patient, and family form a health trio; each one provides different and necessary information and each one is vital to the therapeutic choices. If there is to be successful treatment, PD may require a team of professionals in addition to input from the patient and family. The neurologist, or movement disorder specialist, addresses the physical symptoms of PD; sometimes, a psychiatrist or mental health provider must address both coping with the illness and medicating psychiatric symptoms associated with PD or secondary to medications used to treat the illness. Although PD is progressive, the patient's quality of life can be markedly improved when neurologic, psychiatric, and psychosocial aspects are treated. The initial symptoms of PD may not bother the patient sufficiently to seek medical care, but when they become functionally limiting, it is important to treat and minimize these symptoms as directed by your neurologist. Progressive symptoms can lead to impairment in physical functioning and an associated dissatisfaction with one's performance. Compliance with medical care (timing and dosage of medication) is mandatory, given the nature of PD, especially in the setting of motor fluctuations. The lack of compliance may make it difficult for the treating neurologist to determine whether the dosage of medications prescribed is sufficient and may result in excessive antiparkinson medications, which, in turn, may lead to significant side effects. Compliance reflects acceptance of one's illness and a pro-active contribution to controlling it.

In a family setting, PD affects not only the patient but also the spouse. In general, all chronic illnesses require the support of the spouse and other family members. In the later stages of PD, activities that were considered commonplace in the past may become difficult secondary to movement impairment and require the spouse's assistance. There may be a potential role reversal if the spouse is required to return to or seek employment because the patient can no longer work; or if the spouse must take over the duties of check writing, handling bills, household books, taxes, etc; the patient's self-esteem may diminish accordingly. A patient with PD may not feel that he or she is as attractive to the spouse as before and a withdrawal from the spouse may occur because of fear of rejection. There may be decreased libido secondary to the illness or to medications, further threatening one's sex life. If these issues are addressed, the patient will invariably find the spouse and family to be very supportive. It is crucial to deal with these issues before they get out of hand. No one can help if you do not *communicate* your concerns. Ultimately, the patient with PD must remember that all supportive spouses need time off to relax-a tired spouse cannot assist the patient adequately, may become depressed at the situation, and may even show irritability and frustration. Addressing psychosocial issues together is of paramount importance.

Finally, in addition to other interventions, the patient/spouse/family would also benefit from involvement in support groups. Many of these sup-

port groups are associated with the American Parkinson Disease Association (APDA). The APDA National Office and/or the APDA Information and Referral Centers can give referrals to groups in the patient's neighborhood.

Primary Psychiatric Manifestations of PD

Although psychiatric symptoms are not the most frequent initial signs of PD, the development of depression, anxiety, and panic attacks is not uncommon. As PD progresses, about 50% of patients become depressed, although severe depression is not common. Symptoms of depression include irregularities in sleep, appetite, and energy; decreased concentration, memory, interest level (lack of motivation), and sex drive; feelings of guilt (such as that PD is a punishment), hopelessness, helplessness, worthlessness; and even suicidal thoughts (although suicide is *extremely* rare in PD patients). Panic attacks have a precipitous onset and can include palpitations, shortness of breath, hyperventilation, sweating, GI symptoms, numbness and tingling, and a "feeling of impending doom." These symptoms can and should be treated with appropriate psychotherapy and psychiatric medications. For depression, the class of drugs known as the serotonin specific reuptake inhibitors (SSRIs) may be most useful; these include paroxetine (Paxil®), sertraline (Zoloft®), citalopram (Celexa®), and fluvoxamine (Luvox®). The first drug in this class, fluoxetine (Prozac®), may make PD patients more agitated and so should be reserved as a later choice if others fail. Other agents, mirtazepine (Remeron®) and venlafaxine (Effexor®), may also be helpful (in fact, Remeron has recently been reported to help tremor in PD as well as depression). The oldest class of antidepressants, the tricyclics (TCAs), may be less effective for depression than the SSRIs, but may be more helpful for sleep (see below). These include amitriptyline (Elavil®), nortriptyline (Pamelor®), imipramine (Tofranil®), and desipramine (Norpramin®). These medications must be initiated at low dose and slowly increased, as they may initially be ineffective and require an increased dosage; or, a particular medication may not be effective for a specific PD patient. If the depression is severe and does not respond to multiple medications, and if there are no other potential causes of depression (for example, decreased thyroid functioning can result in depression), then electroconvulsive therapy (ECT) should be considered. ECT has been shown not only to decrease depression but also to decrease PD symptoms for months up to a couple of years. Modern ECT techniques have proven to be safe as well as effective. Panic attacks and anxiety can be controlled with SSRIs, but often low dosage drugs in the Valium® family, called benzodiazepines, are very helpful. They may be used alone or in combination with the SSRIs. Newer benzodiazepines like lorazepam (Ativan®) and alprazolam (Xanax®) tend to cause less drowsiness than the older drugs like diazepam (Valium®).

Sleep disturbances are very common in PD. Often a reduction in nighttime antiparkinson medication will eliminate nightmares. Difficulty falling asleep is not as prevalent as difficulty staying asleep, with multiple awaken-

ings through the night. Selegiline (Eldepryl®) may be associated with insomnia (problems falling asleep); this may be corrected by taking selegiline no later than noon, or just once early in the morning. If falling asleep is a problem, short-acting benzodiazepines like lorazepam (Ativan®) or alprazolam (Xanax®), or a different compound like zolpidem (Ambien®) may be useful; these won't cause a "hangover" effect as is seen with older sedative drugs like temazepam (Restoril®) or flurazepam (Dalmane®), and these latter medications should be avoided in PD.

The usual scenario in PD is that an individual falls asleep just fine, but can't stay asleep. Needing to urinate is usually blamed, and although that may frequently be a real consideration, awakening 3 or 4 times a night to urinate is probably not necessary. Instead, the more likely case is that the sleep cycle is disrupted and when one awakens, one realizes that one could do with emptying one's bladder, even if it's not very full. This sleep pattern probably results from biochemical abnormalities in PD. Just as a deficiency of dopamine is responsible for most of the motor symptoms of PD, there is also a loss of related neurotransmitter chemicals called norepinephrine and serotonin-not as low as dopamine, but also impaired. Norepinephrine and serotonin are important in controlling both mood and the sleep cycle. It therefore makes sense that insomnia is often associated with depression, and that the drugs used to treat depression are more effective for this typical parkinsonian sleep cycle than old-fashioned sedatives. As noted above, the TCAs are often very effective for treating the sleep cycle abnormality; SSRIs may also be helpful. A good night's sleep is important for everyone, but especially so for individuals with PD; unquestionably, Improving sleep patterns results in improved daytime functioning.

In very advanced PD in older patients, significant cognitive dysfunction (mental disablitiy resulting in problems with thinking, memory, planning, and organizing) or dementia may be present. Confusion may be very prominent. Sundowning, or worsening of psychiatric symptoms in the late afternoon/evening, may be noted. At this point, the condition is also called diffuse Lewy body disease, or dementia with Lewy bodies, indicating that the hallmark of PD seen under the microscope, the Lewy body, now is seen not only in the substantia nigra where dopamine is produced, but throughout the brain, which is why more than just motor function is impaired. All the antiparkinson medications may worsen these features; one may be "caught between a rock and a hard place" with balancing motor and mental function, but a "balancing act" may be the best option. With the dementia often come inappropriate behaviors and psychotic symptoms. Treatment of these features is discussed in the next section.

Medication Side Effects

As PD progresses to later stages, there may be a requirement for higher doses and multiple medications that work on the dopamine system. Just about all of these medications have side effects, and in advanced patients,

particularly older individuals, the most disturbing side effects are those of psychosis, which include *hallucinations, delusions*, and *paranoia*. The hallucinations (false perception of something that is not really there) in PD are usually visual in nature, but rarely patients may have auditory (hearing things or voices) or tactile (feeling things, like bugs crawling) hallucinations. Delusions are fixed false beliefs, and paranoia is an irrational belief that others are "out to get" someone in a variety of ways. If possible, the antiparkinson medications should be reduced, or drugs with a poor risk:benefit ratio (such as anticholinergic drugs, amantadine, selegiline) should be eliminated, which may effectively treat the drug-induced psychosis. When motor function is so severely compromised by reduction in levodopa or dopamine agonists, it may be necessary to start antipsychotic medications that will treat the hallucinations. The older, traditional antipsychotic medications are pure dopamine-receptor blocking drugs, and medications such as haloperidol (Haldol®), chlorpromazine (Thorazine®), fluphenazine (Prolixin®), perphenazine (Trilafon®), thiothixene (Navane®), and thioridazine (Mellaril®) must never be used in PD, since it can worsen the symptoms to the point of immobilization, with potentially serious medical side effects. Instead the newer "atypical" antipsychotics that affect both serotonin and dopamine systems have a decreased occurrence of parkinsonian features. One of them, however, risperidone (Risperdal®), has a similar dopamine receptor activity to haloperidol and should be avoided in PD. Another, olanzapine (Zyprexa®), although helpful in low doses in some PD patients, has unfortunately been seen to worsen PD and should be low on the list. The safest and most effective anitpsychotic agents for use in PD are clozapine (Clozaril®) and quetiapine (Seroquel®). Quetiapine is now the treatment of first choice for PD-related hallucinations since it is the safest and easiest to use. Clozapine is about twice as strong as quetiapine but has more side effects in high doses, and, because of a rare but potentially serious problem with the white blood cell count, can only be prescribed in one week amounts after weekly blood tests.

CHAPTER 9
SEX AND PD

Thomas R. Zimmerman, Jr., MD

Introduction

Sexual function is a concern of all individuals of all ages. Patients with PD are frequently concerned with how the disease affects sexual function and also how sexual activity affects their PD. Patients do not routinely ask these questions, so the physician and nurse must often take the initiative to ask about sexual function. It is important to remember that aging does not exhaust the sexual drive; no patient should be embarrassed to ask about it, and no physician should ignore these concerns, whether the patient is 32 or 92.

The following chapter discusses basic information about sexual function, changes with age and disease, and how to address certain problems when they arise.

Normal Function

In both men and women, there are at least four stages of sexual excitement: desire (libido), excitement, orgasm, and resolution.

Men

In men, *desire* or *libido* is linked to testosterone levels; low levels lead to reduced desire. *Excitement* occurs as a result of either psychic stimulation (pictures, movies, thoughts) or touching, or tactile stimulation (caressing, masturbation), which leads to penile erection, lubrication of the urethra with prostatic fluid, nipple erection, and enlargement of the testes. This is followed by *orgasm*, at which time the testes elevate toward the pelvis, and there are deep intense contractions leading to ejaculation of seminal fluid. The *resolution* phase occurs next, in which the penis loses its rigidity. Afterward, there is a *refractory* phase during which the desire for sex is abated. This may last 30 minutes to several hours.

The response described above is controlled by at least two separate but linked mechanisms: *hormonal mechanisms* and *neurologic mechanisms*. The hormone, *testosterone*, is correlated with desire (libido) as well as morning penile erections; it does not necessarily correlate, however, with the ability to have erections from tactile stimulation. Sexual response to touching (tactile stimulation) is controlled by *neurologic mechanisms*. Penile function is governed by two sets of nerves, direct (*parasympathetic nerves*) and indirect (*sympathetic nerves*), which make up the *autonomic nervous system*. The *parasympathetic nervous system* is of primary importance in erection, and appears to be responsible for both tactile and psychic stimulation. The

sympathetic nervous system may be capable of producing erection by controlling blood supply (vascular function), and is responsible for ejaculation. Therefore, many factors play a role in the male sexual function.

Women

Desire (*libido*) in women is controlled by estrogen as well as low levels of male hormones (androgens). *Excitement* occurs when a woman focuses on psychic clues such as books or movies, or tactile cues such as caressing or self-stimulation. At this stage, there is widespread vascular (blood vessel) congestion in the pelvic area, leading to vaginal lubrication, enlargement of the clitoris, and vascular engorgement of the uterus. The uterus rises, the vagina "balloons," becoming deeper and wider and creating an "orgasmic platform" in the lower one-third of the vagina. Blood pressure and heart rate increase and there is also nipple erection and breast engorgement. When *orgasm* occurs, there is rhythmic contraction of the uterus, orgasmic platform, and rectal sphincter resulting in intense pleasure. Afterwards, there is a resolution phase in which the body returns to its pre-excited state. If orgasm were not achieved, resolution would take longer. These activities, like those described above for men, are all under control of the sympathetic and parasympathetic nervous systems and hormonal influences.

Normal Changes with Age

Interest in sex does not necessarily decrease with age. An individual's decrease in sexual function may be the result of illness, lack of a partner, depression, or frustration at the changes in the bodies functions to have or maintain sexual excitement. In men, age alone causes only a minimal decrease in testosterone levels, but waning interest in sex may result from decreased testosterone and is often the result of chronic illness. Normal age-related changes, however, do occur, such as less frequent morning erections and less sustained erections. In women, on the other hand, there are substantial decreases in estrogen (and androgen) production with age, which account for the normal changes in the body (for example, vaginal lubrication and elasticity are diminished as a normal course of events). In both older men and women, however, reduced sexual activity appears to be a function of previous activity: those who were more active when they were younger will remain active; those who were less active will have further decline in activity.

The body's lubrication by vaginal or prostatic fluid is an important part of sexual function that is often overlooked, and more a consequence of age than PD. Lack of vaginal lubrication can cause painful intercourse. It is therefore often necessary to use tactile stimulation before penile penetration, or to use estrogen creams to restore suppleness to the vaginal wall.

Effects of diseases on sexual function

Before considering that PD is the reason for a problem with sexual func-

tion, consider other more common causes. Many conditions affect sexual function either directly or indirectly, they include:

1. Drugs: Certain drugs, including some used for PD such as the anticholinergics (trihexiphenidyl is one), may interfere with sexual function.
2. Vascular: Problems involving the blood supply, which generally result from heart disease, high blood pressure, and diabetes, may reduce blood supply to the genitals and interfere with sexual function. Poor circulation may lead to decrease in libido. Diabetes may also affect the blood supply to the sexual organs as well.
3. Prostate problems: An enlarged prostate may not affect sexual function directly, but can cause urinary problems that can make intercourse uncomfortable. A man who has undergone transurethral prostate surgery (TURP) will still be able to have an erection and orgasm, but may have *retrograde ejaculation* in which the semen is propelled backwards into the bladder instead of out of the penis.
4. Depression: Mood dysfunction can result in loss of libido.

Effects of Parkinson's disease on sexual function

Some patients with PD who have problems with sexual function also have other medical problems such as heart failure or diabetes that explain the reason for the dysfunction. PD does not cause peripheral nerve damage or vascular damage, but it may have an effect on the autonomic nervous system, which can result directly in impotence and other problems. There are other aspects of PD that can interfere both directly and indirectly with sexual function. For instance, loss of dopamine may result in loss of ability to have erections, so the loss of dopamine in the brains of PD patients may have a direct effect on sexual function, although this effect may be minor.

Many patients with PD also have depression as part of the condition. Depression causes a loss of interest in sex, as well as an inability to maintain an erection in men or achieve sexual excitement in women. When the depression and PD are adequately treated, adequate sexual function may return.

The three cardinal manifestations of PD are slowness, stiffness, and tremor. All of these may interfere with the agility that is needed during sexual performance. These symptoms also contribute to the longer time needed to achieve sexual arousal and excitement.

ON-OFF and Dyskinesia

A patient who is severely "off" or wildly dyskinetic will have difficulty having sex, even though the desire for sex may be undiminished. It is important to remember that patients in the "off" state, the period identified with extreme slowness and rigidity, often have mental changes, such as depression. When they go to the "on" state, not only does their motor function improve, but their mental function improves, too. Patients who have dyskinesias represent the other extreme: they have many abnormal involuntary movements

that may interfere with sexual activity without diminishing their libido or ability to get aroused. But the patient may be uncomfortable, or the dyskinesias may actually injure the sexual partner. In both, it is important to time one's sexual activity to the medication to limit "off" time, and remain "on" with minimal dyskinesias.

Effects of sexual function on Parkinson's disease

There is no evidence that sexual activity decreases dopamine or causes long-term changes or worsening of PD. There may, however, be temporary changes in PD symptoms at the time of sexual activity.

The physical activity associated with sexual activity is often likened to climbing a flight of stairs. Furthermore, when timing sexual activity, it is important to consider that if one is tired, one may not be able to perform sexually. This is true in healthy young adults, as well as people with PD. In the same way that you must time your daily activities such as eating, shopping, or exercising to the medication, so must sexual activity. If you are "off" or if you are having bad dyskinesias, sexual activity will be difficult.

Patients have indicated that they can not predict what short-term effect sex activity will have on their PD. They may emerge after sex with more dyskinesias, more *"off"* time, or be just the same.This may differ from day to day. If worse, it may be necessary for you to take an extra levodopa dose *before* having sexual activity. If dyskinesias are worse, then the medication may either have to be reduced, or the timing changed. It is suggested that you experiment on your own and then discuss medication changes with your doctor.

Effects of medications on sexual function

Many medications effect sexual function. A review of the Physicians Desk Reference (2000) revealed that over 300 medications were reported to be associated with some form of sexual dysfunction. It is important to understand what this means. It does not mean that the medications are associated with a "cause-and-effect" relationship, but that patients taking these medicines have reported symptoms of sexual dysfunction. When discussing problems with the physician or nurse, it is important first to define what area of sexual function is affected, then explore any medications that you are taking. Is libido or desire effected? Ability to have or maintain erection? Always know ALL the medications you are taking, as something you may be taking for another condition may be causing unacceptable side effects.

Of drugs that may be used in PD, those that have "anticholinergic" effects (primary tremor drugs like trihexiphenidyl, or antidepressants like amitriptyline (Elavil¤), or even diphenhydramine (Benadryl¤) can affect the function of the nerves and may make erection or ejaculation difficult. These drugs can also slow the heart rate as well.

Carbidopa/levodopa (Sinemet¤) is associated with increased libido. The dopamine agonists such as pergolide (Permax¤), pramipexole

(Mirapex®), and ropinirole (Requip®) are associated with both impotence as well as increased libido. The COMT inhibitor, tolcapone (Tasmar®), is also associated with both impotence and increased libido. Antidepressant medications may increase libido (as they treat depression) but may also have a negative effect on desire. They may also be associated with problems with erections and ejaculation.

Approaches to problems

As discussed above, there is no one single effect of PD medications on sexual function. In fact, some medications such as the agonists are associated with several different and opposite effects. Thus, any discussion of sexual function in PD must consider the following:

- ¥ Are there any coincident diseases that can cause sexual dysfunction?
- ¥ Are there any medications, either prescription or over-the-counter, that may contribute to sexual dysfunction?
- ¥ Is the patient depressed?
- ¥ Is the PD medication timed properly to allow for sexual function?
- ¥ What are the effects of sexual excitement on your medication and PD symptoms? Can you alter the timing or dose of your medication? Nighttime is not the best time for many PD patients, so you may want to have sex in the morning or during the day.

Assuming that one has explored the possibilities above, and "optimized" everything, then it is time to consider other treatments.

Men

Oral Medications

For patients with erectile dysfunction due to vascular disease, there are two marketed drugs, sildenafil (Viagra®) and yohimbine (Aphrodyne®). Sildenafil may be used in PD but there are several contraindications. First, it should not be used if you have significant heart disease or are taking nitrates (for example, nitroglycerin, Isordil®, or even a nitrate patch). Second, because sildenafil causes dilation of blood vessels, it lowers blood pressure. This effect can be additive to PD patients taking antiparkinson medications who have low blood pressure when standing. Yohimbine has similar warnings, but, although it may also be used to treat erectile dysfunction, no formal studies have been performed.

Apomorphine is currently being presented to the FDA for use in erectile dysfunction. Apomorphine is available in Europe and Canada as an injection for the treatment of PD, but is also known to improve penile erections.

Other Treatments

Injectable medications: In the last few years, several agents have become available that can be injected into the penis in order to enhance erections. These drugs lead to improved blood flow. When injected, these drugs cause an erection that may last several minutes to several hours,

depending on one's sensitivity. The three drugs in this category are papaverine, phentolamine, and prostaglandin, all of which may be mixed together, if needed. The patient self-administers these drugs by a relatively painless injection directly into the penis. The sensation and orgasm will be nearly normal. A new drug delivery system known as MUSE has become available in which Aloprisidil® is administered by depositing a pellet in the urethra. The pellet dissolves and the medication is absorbed into the penis, resulting in an erection about 65% of the time.

***Vacuum devices*:** If the patient can not take or tolerate the above oral or injectable medications, the vacuum devices are commercially available. These act on the principle of drawing blood into the penis. The blood is retained in the penis by placing a retention ring at the base of the penis (similar to a tourniquet). Patients indicate that the erection appears and feels normal.

Prostheses: There are two types of prostheses, semi-rigid-moveable and inflatable. The semi-rigid prosthesis is surgically implanted into the penis and gives one a permanent state of erection. It is moveable so that the penis can be moved out of the way to wear clothes. With this device, the penis does not get larger with sexual excitement, but allows for intercourse. Inflatable devices allow one to chose when he is going to have an erection. Two cylinders are implanted in the penis; a reservoir is placed in the abdomen and a pump in the scrotum. All of the parts are concealed. When an erection is desired, the pump is used to inflate the cylinders and hence the penis. Sensation is quite normal for both prostheses. Orgasm may be achieved, but not necessarily ejaculation. These prostheses carry the risks of surgery, including infection, but these risks are very small. Pump failure is less than 10%.

These treatments are reserved for significant problems with sexual function. There are specific indications and contraindications. A consultation with a urologist is critical to determine if one is a candidate for one of these invasive approaches.

Women

None of the oral medications such as sildenafil, yohimbine, or apomorphine are approved for women yet. Physiologically, one might expect them to help because they increase blood supply to the pelvis, but safety and effectiveness are not proven.

Lack of adequate vaginal lubrication may be caused either by insufficient stimulation or because of dry, narrowed vaginal tissues. Lack of sufficient lubrication may also cause pain during intercourse. One approach to improve the situation is to have the sexual partner provide a longer time of stimulation of the genitals before intercourse. If this fails, the women should undergo an examination of the vagina to determine whether she might benefit from oral estrogens or estrogen creams in order to increase the suppleness of the vaginal tissue and improve lubrication. In addition, water-soluble lubricants are also helpful. If the vagina is narrowed, the use of lubricants and a vaginal dilator (15 minutes per day) may also be required.

CHAPTER 10
CAREPARTNERS

Hedy Weinstein, RN, MS

Psychological and Social Support

When a person receives a diagnosis of PD, they need some time to adjust to this new information. The spouse and any other close relatives and friends must also be given an opportunity to accept this diagnosis. For some people, this acceptance comes easily; for others, it is more difficult and a slower process. Once an individual comes to terms with a new self-image, he/she still may have difficult days in which they respond to the disease with anger and frustration. If significant despair persists, the patient and his or her significant other should consider medical intervention. Frequently, the spouses of the PD patient will find themselves helping their partner and other members of the family cope with the disease. You may be the one that everyone relies on to answer questions and to keep them informed. If the carepartner is not a spouse, then a parent or an adult child may be called upon to support the patient.

The young parkinsonian will have many relationships with those around him. Colleagues will be found in the workplace, socially, and within one's own family. Most patients will have one individual that they have chosen to become their *carepartner.* This is the individual who assists the patient with tasks and activities of daily living. These tasks vary depending on the patient's needs. The carepartner takes an active role in learning about the disease process and is involved with the treatment plan. Frequently, this individual is a spouse, but can be any relative or friend, as long as the individual is committed to working with the patient in all issues related to his or her PD.

As the disease continues to evolve, the symptoms of the disease may change. The role of the carepartner is also changing. The carepartner focuses a great deal of energy on the needs of the patient with PD. If the patient continues to work, some patients choose not discuss their diagnosis at work and therefore it remains a secret. Some feel they will be terminated and others are afraid they will be treated differently. Whether or not to reveal one's diagnosis is a private decision. The carepartner can play a key role in supporting the patient as they tackle this issue and helping make these decisions.

One of the most significant qualities a carepartner must possess is that of being a good listener. Sometimes the patient will need to talk to someone who will not judge them. They will be frightened and may need someone to comfort them. In these situations, the carepartner takes on the role of "cheerleader." They encourage the patient to remain positive and focused,

while allowing them to communicate their fears and concerns.

The role of the carepartner can be very challenging at times. Most carepartners of young PD patients are also working. They may have distractions from their children and from their own parents. They may feel as though they are being pulled in many different directions, and the subsequent stress can make them very vulnerable. The carepartners need to have a mechanism for support as well. Both the patient and carepartner must keep lines of communication open. Building bridges in their relationship is a wonderful way to maintain effective communication. They both need an opportunity to verbalize their feelings.

Anger can be a very common emotion for carepartners. They often express feelings of anger in learning about the diagnosis. They now have to incorporate a disabled person into their life! This anger can be a result of accepting the reality that their vision for the future will not be as they planned. Both the patient and those significant others around them must mourn their previous relationship and the patient's good health. It is only then that they can learn to dismiss these destructive angry thoughts and replace them with positive motivation. The diagnosis can be the foundation of a stronger bond between the patient and spouse (or other carepartner) as they work together to deal with this progressive and chronic disorder.

Frustration with the unpredictable aspects of PD can be a burden to a carepartnership. The constant need for medication and the unpredictable response to therapy can make planning a daily routine a difficult task. The anticipation of what may lie ahead (what sacrifices and adjustments may be necessary to cope with the progression of the disease) does not promote a sense of future security. It is important to remember, however, that each case of PD is different for every individual. For some young-onset patients, the progression of the disease is generally slow. Many young patients will be able to function with only slight limitations and most daily plans can be adjusted to compensate for times when medications are not working as well.

The carepartner should maintain social activities with and without the PD patient. Making time to socialize with family, friends, and coworkers can help minimize feelings of isolation and loneliness. PD patients may be reluctant to socialize if they are fatigued or are concerned about their symptoms. It is the responsibility of the carepartner to reach out and help plan social engagements.

Physical Support

A young PD patient can be very independent and enjoy an active lifestyle, whereas another patient may be more dependent on their carepartner, requiring increased emotional and physical support. The carepartner must try to avoid "taking over." It is very easy to get in the habit of helping the patient. They may need assistance with dressing, for example, but don't forget to encourage independence. You can foster this by helping the patient

to identify areas of difficulty.

Once again the patient and carepartner must communicate carefully to alleviate any anxiety here. Compile a list of activities of daily living that the patient finds difficult. Then take each issue and determine what he or she may need assistance with. Adjustments can be made to the bathroom to promote safety. In the kitchen, cabinets can be re-organized to place the most frequently used items in easy reach. Assessing the home with the patient can help nurture independence and security.

As the disease progresses, reassess what the patient's needs are. Work as a team to promote independence and a positive self-image. Patients want to maintain their dignity. Careful observation combined with communication can achieve the highest level of freedom.

Care for the Caregiver

The caregiver must take time to care for themselves. A proper, well-balanced diet and daily exercise is essential. Every effort should be made to develop outlets to relieve stress. Keeping a sense of humor is also a pre-requisite for a healthy lifestyle.

The carepartner who is also an adult child of the patient may also be a parent of his or her own children, and this can become very stressful. As an adult child, they need to balance the needs of their spouse, children, and now the parent with PD. They will need to learn to function as the support person for many different individuals. Remaining socially active and having close personal relationships often helps the daughter or son of the patient to have some down time and to relax.

If the carepartner is a spouse, they will most likely be living with the patient and will be with the patient on a daily basis. Participating in social activities that are important to the carepartner will foster some independence for the spouse who is in the carepartner role. Maintaining friendships and carrying on one's regular routines will also help.

Whether or not the carepartner is an adult child or a spouse, their needs are very similar. They must recognize that their expectations for the future have now changed. The carepartner must acknowledge that their feelings of anger, frustration, and fatigue are normal. Have someone other than the patient to talk to. This person must be able to listen carefully and not monopolize the conversation. You may also find it helpful to speak to others who are also dealing with similar circumstances.

A caregiver must possess the ability to say "NO!" When responsibilities or issues prevent you from doing something, learn that saying no is the best response. Frequently, the matter can be postponed until it is convenient for the individual. If it is something that will not be able to be accomplished in the future, then that must be determined quicly and one should feel confident with one's decision.

Weapons Against Parkinson's Disease

Knowledge is a powerful weapon to use in living with a progressive and chronic disease. The carepartner and patient should understand the disease as well as the treatment plan. They should question the physician if either of them are uncertain about a particular medication or if the symptoms have changed. Knowledge about the disease will help alleviate the fear of the unknown. Most physicians encourage patients and their families to take an active role in the treatment plan. Mutual trust and respect must be established with the practitioner and the family at the beginning. This will be a crucial building block for the future.

PD is manifested differently in every patient. There are no absolute guarantees of which symptoms will develop. Through reading, attending educational seminars, and support groups, the patient, the carepartner and the entire family can better understand what the disease may have in store for them. Then they can take it one day at a time and manage the current symptoms. This will help everyone to focus on the present symptoms and not worry about what is to come. Some patients decide to do fundraising or to work with political advocacy groups to raise awareness and funds for research.

The patient and their family can begin to plan their assault on this condition by keeping a diary (handwritten or audiotaped) of symptoms and a list of questions so they are prepared when meeting with their physician. This will also allow them to observe subtle changes and participate in choosing alternatives in their treatment plan. They should also be familiar with all the medications, the dosages, and the schedule. This will help everyone to focus on the present situation and not worry about the distant future (which no one is able to foretell!).

APDA Resources

The American Parkinson Disease Association Inc. has dedicated an information and referral center which focuses solely on the Young Onset Parkinsonian. This site has a large resource of literature on many unique issues facing the young patient and family members. The Center can also connect young patients with other young patients so they can talk and share the experience as only they can understand. There is also a newsletter, computer networks, and trained personnel to speak with.

A Final Thought

Navigating through the years with a diagnosis of PD will have its effect on the entire family. With education, patience, faith, love, and support, patients and their carepartners can support one another.

Resources

The following is a list of resources that may be helpful as you build the arsenal of educational and support weapons in your fight against PD:

* American Parkinson Disease Association
  1250 Hylan Blvd. Suite 4B, Staten Island, NY, 10305
  Phone: 800-223-2732
  Email: info@apdaparkinson.com

* Arlette Johnson APDA Young Information & Referral Center,
  Glenbrook Hospital
  2100 Phingsten Road, Glenview, IL 60025
  Phone: 800-223-9776
  Email: apdaypd@aol.com

CHAPTER 11
FINANCIAL PLANNING AND PROVING DISABILITY TO THE SOCIAL SECURITY ADMINISTRATION

Jan Powell, BA, MA, MDiv
and
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What comes to mind when someone says the words "financial planning?" Do you think of a wealthy individual in search of tax shelters, or planning for your child's Ivy League education? Are you tempted to skip over this section because you think that financial planning is boring and complicated? Do you have trouble seeing how it will help you as a PD patient?

Actually, financial planning is for everyone, young and old. It's especially important to those facing a challenging illness like PD. The good news is that *smart financial planning is simply putting your own creativity, experience, and common sense to good use.*

Ideally, of course, a financial plan should be designed and maintained by a personal financial planner. The planner should preferably be an attorney with a strong practice in Medicaid law, as well as estate and trust planning. But even individuals with limited financial resources can help create a realistic plan of action to help them obtain needed benefits and conserve both their income stream and hard-earned assets.

This chapter will show you some of the financial tools available to construct and maintain your financial plan...a plan that now includes dealing with the effects of PD. Information on these and other resources is available from The American Parkinson Disease Association, Inc., your insurance company, governmental agencies, local hospitals, and many other groups.

Disability Benefits Overview

As a PD patient, there are several types of disability benefits you could receive. Understanding what they are, how to get them, and how they fit into your overall financial plan is essential.

(1) Mr. Smith contributed the section on Social Security Disability

Employer Group or Individual Disability Insurance benefits replace a portion of your income lost due to disability. If you had disability insurance through your employer, or an individual plan, prior to the onset of PD, the benefits will be supplemented by Social Security Disability benefits. The amount depends on the plan that covers you. Some disability insurance-especially group plans through your employer-offer special services to help you get benefits. This is especially important when it comes to Social Security, since only 35% of claim applications are approved in the initial decision. An insurer's Social Security consultant can explain the appeals process if you are initially denied, help you contact advocates, and assist you with the necessary paperwork. Your insurer also has information on how Social Security and other benefits affect taxes and retirement income. Many offer vocational rehabilitation assistance and will work with your employer on a specific return-to-work and stay-at-work program for you.

People usually think of ***Social Security Disability (SSD)*** insurance and the ***Supplemental Security Income (SSI)*** program first when it comes to disability benefits. Administered by the US government, benefits are contingent upon whether your illness meets certain criteria that make it a "disability." The medical requirements for establishment of disability are the same under both programs. Eligibility for SSD benefits is based on your FICA contributions, while SSI benefits are determined by financial need.

SSD: Social Security Disability Benefit

The Social Security Administration (SSA) has a number of criteria that, once met, define you as "disabled" and qualify you for benefits. You are considered disabled if you are unable to do any kind of work for which you are suited, if your inability to work is expected to last at least one full year, or if it is expected to result in death. Benefits will continue for as long as you are disabled. You can get SSD benefits at any age, provided the requirements are met.

Your work history determines your SSD benefits eligibility. Social Security benefits are based on "credits" earned through work. "Credits" are based on wages or salary. The amount that must be earned for each "credit" increases every year (as general wage levels in the US increase), but one can earn a maximum of only four credits in each year worked. The number of credits needed to obtain SSD benefits is based on your age when you become disabled.

According to the 1999 edition of "Disability," published by the Social Security Administration, you need the following credits to be eligible for SSD benefits:

- Before Age 24 You need six credits in the 3-year period ending when your disability starts.

- Age 24 to 31 You need credits for having worked half the time between 21 and the time you become disabled.

- Age 31 or older You need to have the number of work credits shown in the chart below. *At least 20 of the credits must have been earned in the ten years immediately before you became disabled.*

Born after 1929 and became disabled at age	Credits you need to qualify
31 through 42	20
44	22
46	24
48	26
50	28
52	30
54	32
56	34
58	36
60	38
62 or older	40

Applying for SSD Benefits

The most important thing about applying for SSD benefits has nothing to do with credits, forms, or age. It has to do with ***time***. You should apply immediately after becoming disabled. You can apply in person at any Social Security Office, apply by phone, or apply by mail.

Why apply so quickly? First, it can take SSD offices 60-90 days to process your disability benefit claim, while they gather pertinent medical information and evaluate your potential disability. Second, you may need to go back to your doctor(s) for more information or contact your insurance company. Promptness assures that any retroactive benefits are protected, that disability insurance coverage does not lapse, and that evidence does not disappear or go into storage. The longer you wait there is more chance of misplaced forms, additional requests for information, slower responses, and other delays. *The bottom line is under the best circumstances, once your application has been approved, you probably will not receive SSD benefits until the sixth full month of your disability.*

You can help shorten the process by furnishing the following information *in full* when you apply:

1. Social Security Number and Proof of Age,

2. Correct names, addresses, and phone numbers-of your doctors, the hospitals, institutions, and clinics where you've been treated, and the treatment dates,

3. A clear, detailed summary of your work history-include names, addresses, and phone numbers of employers and a description of the work you did for the last 15 years, and

4. A copy of your W-2 Form. If you are self-employed, submit your Federal Tax return for the most recent year.

Most Social Security offices are staffed by people who are glad to help you complete the application and answer any questions you might have. Remember: you have a right to receive help with your application-and you deserve courteous service. Don't hesitate to ask for what you need.

After You Apply for Benefits

The Social Security office staff will review your application for basic information. They will look at the nature of your potential disability, whether you've worked long enough and when you last worked, and check to see that you've included all of the requested data such as Proof of Age and Social Security Number.

Assuming you've included all the necessary information, your application will be sent to the Disability Determination Services office in your state. In this office, a physician or psychologist (depending on the nature of your claim), together with a disability evaluation expert, will review your application. The process can slow here if the team needs to send for more documentation, such as medical records. That's why it's important to give your local Social Security office all of the pertinent medical reports available.

What kind of medical documentation are the application evaluators looking for? Your medical reports should include a medical history of your condition, what exactly is wrong and when it started, and how your condition limits your ability to work and/or your general activities. They should include what medical tests have been taken and their results, along with what treatment has been provided to you. Even with all of this, the evaluation team may require a "consultative examination." This exam is given by a doctor recommended by Social Security, and is paid for by Social Security.

Your Disability Determination Services team will base their decision on five basic questions:

1. Are you working?
   If you work and earn more than $700 a month, generally your application will be denied.

2. Is your condition severe?
   It must be found that your illness or symptoms interfere with your basic work-related activities.

3. Is your condition on the list of disabling impairments maintained by Social Security?
   Very few "diseases" are on this list; "parkinsonian syndromes" is one, although it does not spell out PD per se. Nevertheless, PD is considered a parkinsonian syndrome and should therefore make this step a little easier.

4. Can you work as you did previously?

5. Can you do another kind of work?
   Age, education, past work experience, transferable skills and your medical reports are tools used to assess your ability to do another kind of work.

CLAIMANTS MUST PROVE THEIR CLAIMS

Proving disability is not easy. The federal statute says:

"An individual shall not be considered to be under a disability unless he furnishes such medical and other evidence of the existence thereof as the Commissioner of Social Security may require."

A claimant must provide substantial evidence that answers three basic questions in the statute:

1. Can he or she do substantial gainful work?

2. Do the limitations on work capacity result from a medically determinable physical or mental impairment or combination of impairments?

3. Is impairment expected to last at least 12 continuous months (from the time it made the claimant unable to work), or result in death?

SSA typically denies claims if the evidence leaves doubt about answers to any of the above questions.

PROVE YOUR CASE

Disability is proved the same way under both Social Security Disability Insurance (SSDI) and Supplemental Security Income (SSI) programs. The best way to prove a disability claim is to simultaneously present SSA with the application for benefits and all the medical evidence available. This shows that the claim is ready-or nearly ready-for decision, and allows the SSA decision maker to anticipate a quick, clean job.

A. List information before filing a claim.

To assure getting all the available medical data, claimants should create five lists. The lists include:

1. Impairments. Claimants should list every impairment that hinders work significantly-not just the most recent or most severe impairment.

2. Medical sources. List doctors, hospitals, clinics, chiropractors, therapists, and other health care providers, including their names and most recent addresses and phone numbers. When claimants furnish these details, SSA does not waste scarce resources getting routine information.

3. Medications/therapy. Also describe current medication and therapy and the good and bad results (and also discontinued medication and therapy if it illustrates disability at an earlier time). SSA looks at medication and therapy as an indication of impairment severity.

4. Daily living activities. SSA needs to know what claimants do, do not do, and what others do for them because of their disability. So, one should think of a typical month and write a short summary describing the number of "good", "fair," and "bad" days. A "good" day is one when most necessary things get done; on a "fair day" some but not all necessary things get done; and on a "bad" day nothing gets done.

5. Jobs of the past 15 years. Claimants must identify each job and describe its essential physical and mental demands. Part of this should be an explanation of the most demanding physical and mental tasks, and whether one can perform such activities now.

Attach a copy of each list to the SSA Disability Report form. Also give a copy of the daily activities information to your doctor. Not only does receiving the lists help SSA understand the claim, but making the lists organizes a claimant's thought process and promotes effective communication with SSA personnel.

B. Get a thorough medical report

A thorough medical report addresses the three statutory questions listed at the beginning of this chapter. It helps SSA decision makers (who are not doctors) understand how incapacity to work results from the claimant's medical condition. It focuses on both the diagnosed medical conditions, and on how they affect the patient's daily activities. Photocopies of applicable medical test results should be stapled to the report.

To assure that a thorough medical report reaches the disability decision maker, claimants should not only tell SSA about sources of medical evidence, but also should contact at least one of their treating doctors themselves. They should ask for a report that explains the diagnosis and illustrates the disability by giving examples of how the illness or injury impairs the ability to function. Illustrations may be based on what the doctor observes or on what the patient tells the doctor. If the doctor repeats what the patient says, the doctor should explain why the patient's statement is considered credible.

APPLY IN PERSON

Claimants should go to SSA offices (or if necessary ask SSA representatives to come to them) for disability interviews. SSA encourages people to file papers by mail and have interviews by telephone, but government studies show that more errors occur in cases where claimants are not interviewed in person.

A face-to-face interview enables the claimant to get advice from a trained SSA representative as well as a review of the claim papers. This assures correction of any errors before the papers go to the decision maker.

KEEP COPIES OF EVERYTHING

SSA often loses claim folders, and documents out of folders. Making notes and keeping copies of everything allows claimants to replace items that become lost, and to prove they followed proper procedures if that becomes necessary. So, when talking to SSA, note the name of the representative, and the date, time, and substance of the conversation.

Referring back to notes and copies of documents previously filed, also can prevent one from making conflicting statements.

CHECK UP ON CLAIMS

After filing disability papers at the local SSA office, claimants should

monitor the progress of their claims. This assures that all medical sources provide the necessary information. About two weeks after filing their papers, the claimant should telephone the state Disability Determination Services (DDS) office evaluating the claim, ask for the disability examiner assigned to the case, and confirm that the folder has arrived. An SSA office can furnish the DDS telephone number.

When telephoning, ask the disability examiner which medical sources have provided evidence, and which have not. If all medical sources haven't responded, claimants should ask the disability examiner for permission to check back in about two weeks. (Permission isn't needed, but it is a courtesy to ask.) Always write down the examiner's name and telephone number for later use.

After contacting the disability examiner, claimants should check with any non-responding medical sources to assure that they supply evidence. Follow-up calls to doctors, clinics, and hospitals should be courteous, but must stress the vital importance of getting the documentation quickly. Sometimes it helps when a patient offers to come by and pick up the documents.

It is wise to recheck with the DDS about every two weeks until all medical sources have supplied the evidence.

If Your Claim Is Approved

If you are one of the fortunate 35% of Americans approved for Social Security Benefits in the initial decision, your first SSD check may (but not always) date back to the sixth full month from the onset of your disability. The amount you will receive each month is based on your FICA contributions. (Your Social Security office can furnish you with an estimate of what you are likely to receive.) If you receive other disability benefits payments from workers' compensation, Federal, state or local governments, Civil Service or private insurance, the amount you will receive from SSD could be reduced. As a rule of thumb, total disability payments *may not exceed 80% of your average current earnings* before you were disabled.

Trial Work Period

After you are approved for SSD benefits, you also become entitled to a nine-month Trial Work Period. This provision permits you to work up to at least nine months and-regardless of the amount of your earnings-still receive full Social Security benefits. This allows you to attempt to return to work (if you want) without loss of Social Security benefits. If you are able to continue working, benefits could eventually terminate after the 12th month of work.

But don't try working until you and your doctor agree you are likely to be able to work indefinitely. Any month you earn $200 or more is counted as a trial work month and deducted from the 9 months to which you are entitled. Do not waste trial work months by working sporadically for small sums of money.

If Your Claim is Denied

Two-thirds of all SSD claims are denied in the initial Social Security decision. That fact may not make you feel any better if you are denied, but it will help you understand that you are not alone, and that you have several more opportunities to get benefits by appealing.

There are a number of reasons why claims-especially first-time claims-are denied. Perhaps there was an incomplete description of your disability by your doctors and medical providers. Missing paperwork, wrong dates, unexplained gaps in medical or work history, and a number of other factors lead to denial of benefits. When SSA disapproves a claim, the claimant should not give up. Many people appeal successfully and overcome the initial disapproval of their claims.

For 60 days after SSA issues an unfavorable decision, it is nothing more than a notice that the agency is not yet convinced of the disability. Appealing the decision within 60 days causes the agency to consider any additional evidence supplied and to make a new decision.

But not appealing within 60 days makes the unfavorable decision final and usually irrevocable. Therefore, claimants should appeal unfavorable decisions promptly-within 60 days-if they and their treating physicians agree they are still disabled. Each time an appeal is made, claimants should gather and submit any newly obtained evidence to SSA, including updated medical reports. Doing so within a week to ten days after a notice of denial may bring about a quicker favorable decision.

If you are denied, you may appeal the decision. There are a number of appeal levels:

- Level 1 is a **"Reconsideration."** A team other than the team that denied your claim will review all data.

- Level 2 is a **"Hearing."** If the reconsideration is still not in your favor, you may apply for a hearing before a judge.

- Level 3 is an **"Appeals Council."** If the judge supports the denial, an Appeals Council can review your case. At this point you can appeal to a Federal Civil Court if your case is denied.

¥ Level 4 is an appeal to a United States District Court. You may appeal for a hearing at US District Court and, if the claim denial is supported, you have 60 days to appeal to a Federal Court.

Keep Appealing

Many claimants currently must make more than one appeal to win benefits. But higher level decision-makers often are more thorough and attentive than those making the initial decisions, meaning that even though lower level decision makers approved only 35% of claims in 1998, administrative law judges approved 53.3% percent of the claims appealed to them.

SSA TESTING NEW DECISION PROCESS IN TEN STATES

Claimants for disability benefits in ten states now have an opportunity to submit additional evidence to Social Security decision makers when the agency is about to disapprove their claims. This is part of the new "disability redesign prototype" decision process initiated in October 1999 in Alabama; Alaska; California (Los Angeles North and West areas); Colorado; Louisiana; Michigan; Missouri; New Hampshire; New York (Brooklyn and Albany areas); and Pennsylvania. Here is how it works.

In a "prototype state" the claimant who is about to be denied benefits receives what SSA calls a "claimant conference letter." Though vaguely worded, the letter is a warning to submit more evidence or lose the case. To discover the problem that threatens the case, the claimant contacts the government disability "adjudicator" (decision maker) and asks for a claimant conference. In the conference the claimant should: (1) ask which medical sources have failed to provide evidence; and (2) ask which sources have provided evidence that failed to address disability issues appropriately. With this information, the claimant gets the non-responding medical sources to provide the necessary evidence quickly.

If, however, SSA denies benefits, claimants can appeal. In prototype states there is no conventional "reconsideration" step. Instead, an unsuccessful claimant appeals directly to an administrative law judge, requesting a hearing. Aside from these procedural differences, the claimant has the same role in both prototype states and states that follow the conventional decision process; that is, to document the claim with evidence that proves disability.

Consider Using a Lawyer

Does a claimant need a lawyer? Claimants are not legally required to have lawyers to pursue Social Security disability claims or appeals, and most claimants file their claims without lawyers. But when SSA disapproves a claim, claimants should consider getting a lawyer to help with appeals. Many claimants handle their own reconsideration appeals because reconsideration generally is a matter of obtaining all missing medical evidence and supplying it to SSA. Ideally, everyone should have a lawyer during a hearing before an administrative law judge.

At the administrative law judge hearing level, a lawyer is needed because both the administrative law judges and many staff members are lawyers, and it helps to speak the language. Legal rules and precedents may need to be argued. Also, administrative law judges may summon medical and vocational experts who often must be cross examined. This is better done by experienced lawyers than by claimants.

To find experienced disability lawyers people may call the National Organization of Social Security Claimants' Representatives (NOSSCR), telephone 1-800-431-2804. NOSSCR has about 2000 members nationally, mostly lawyers. People also may call their state or local lawyer referral services, but they should specifically ask for lawyers with extensive Social Security disability experience.

Medicare

You will automatically be enrolled in Medicare at the end of your first two years on disability.

Do SSD Benefits Ever Stop?

Your benefits will continue for as long as you are disabled, but your claim of disability will be reviewed periodically. According to the Social Security Administration, your review will typically take place in:

¥ 6-18 months if medical improvement is "expected,"
¥ 3 years if medical improvement is "possible," or
¥ 7 years if medical improvement is not expected.

SSI: Supplemental Security Income

The most important thing to remember about this section is to talk to your local Social Security office. You may locate the nearest office or

make an appointment by calling 1-800-772-1213. Because the amounts payable for SSI differ from state to state, your Social Security office can be a rich resource of information, advice and help.

As defined for SSD, SSI also considers a person disabled if that person has a physical or mental problem that keeps him or her from working, is expected to last at least a year, or to result in death.

How much one can receive in SSI benefit payments is determined, in part, by where one lives. The basic SSI check is, according to the Social Security Administration's 1993 "SSI Supplemental Security Income" publication, the same across the country. Many states, however, add money to the basic check. To find out the amount of the SSI "basic" check and the amount that might be added by your state of residence, call 1-800-772-1213.

Qualifying for SSI

Basics first! To be eligible for SSI benefits you must:

¥ Live in the US or the Northern Mariana Islands,

¥ Be a US citizen or be in the US legally,

¥ Apply for regular Social Security or any other benefits for which you are eligible. You can receive both SSD and SSI checks if you're eligible for both. and

¥ Accept vocational rehabilitation services if they are offered

Whether or not you qualify for SSI depends on what you own and on your income. If you're married, your spouse's income and possessions will be considered, too.

How much income you can have each month and still be eligible for SSI benefits is also determined, in part, on where you live, since each state imposes its own limits. Again, the best place to get the most recent information is 1-800-772-1213.

The Social Security Administration *doesn't count all of your income* when determining SSI eligibility. For example, they don't count:

¥ The first $20 of most income received during any month,
¥ The first $65 you earn each month from working and half the amount over $65,
¥ Food stamps

- ¥ Most food, clothing or shelter you receive from a non-profit agency,
- ¥ Most Home Energy Assistance, or
- ¥ Salary or wages spent to pay for items/services you need to help you work.

Subtract any or all of the above, and whatever income you receive after these deductions is considered "countable" income.

The Social Security Administration will also let you *make exceptions for your possessions*. Real estate owned, personal belongings, bank accounts, cash, stocks, and bonds are considered your possessions, but:

- ¥ A single person with possessions valued at up to $2000 can qualify for SSI.

- ¥ A married couple can have possessions valued up to $3000.

- ¥ The home you live in and the property it's on don't count in SSI considerations of ownership.

- ¥ Depending on the value, personal and household goods and life insurance may not count.

- ¥ Your car doesn't count.

- ¥ Burial plots for you and your immediate family don't count. In addition, up to $1500 in burial funds for you and your spouse may not count.

- ¥ Items you use for work or to generate income may not count.

In addition, if you're disabled but still working, there are special rules to help you continue some benefits from SSI. But remember, as your earnings increase, SSI benefits may decrease.

SSI rules may also allow you to set aside some of your earnings to achieve a work goal or to go to school. Besides the 800 number, Social Securities' booklet "Working While Disabled How Social Security Can Help" (Publication #05-10095) can give you valuable information.

Enrolling for SSI

To sign up for SSI benefits, visit your local Social Security office or call 1-800-772-1213 for an appointment. It's extremely important to ***apply as soon as possible***. The faster you apply, the faster you may begin receiving benefits!

When you go to your Social Security office, take the following items with you.

1. Your Birth Certificate or other proof of age;

2. A summary of information about your home (mortgage information, loan number, mortgage holder(s), monthly payments, etc.) or rental information (lease, landlord, monthly payments, etc.);

3. Payroll stubs, bankbooks, insurance policies, car registration, burial fund information, and all possible information on your earnings and possessions;

4. Names, addresses, and phone numbers of doctors who have treated you, as well as clinics or hospitals where you've been treated.

Other SSI Benefits

If you qualify for SSI benefits, you may qualify for additional help from your state or county. You may, for example, be able to get Medicaid and/or Food Stamps. For more information, call your local Social Service or Public Welfare office.

Social Security Benefits: Summary

SSA's two disability programs, SSD and SSI, provide valuable benefits to people who learn the disability criteria, organize their cases, and supply to SSA the evidence that proves their claims. But people who merely "sign up" for disability often are disappointed. The disability application process challenges claimants to exert their best efforts to prove their claims, and usually rewards people who persevere.

Beyond The Benefits

You can see how important disability benefits are to your overall financial plan. Now that you know what they are and how to get them, it's time to add two legal components to your plan. Remember just because they're "legal" doesn't make them complicated or expensive. In fact, ***every responsible adult who has a family and/or who holds property should have these legal documents.*** Regardless of your current health, you should have a legal Power of Attorney and a Living Will.

Why You Need A Power of Attorney

Someone with a "Power of Attorney" to act in your behalf means that you are giving someone the authority to handle your financial affairs. This includes access to your bank account, selling stocks for you, handling debts, etc.

A **regular** Power of Attorney gives specific and limited authority to someone you choose, but that authority ends if you become incapacitated. A **durable** Power of Attorney still limits the person's authority, but it remains in effect if you become incapacitated.

You can see how important and helpful a Durable Power of Attorney would be to someone, especially someone who is single. It's important, of course, to choose someone in whom you have complete confidence. If you don't designate someone, and you become incapacitated, the state can step in and set up a Conservatorship for you. This would take the choice of who runs your affairs out of your hands-and could end up costing you money.

Why You Need a Living Will

Could you picture yourself being kept alive by machines and feeding tubes for years? What would that do to you to your family and friends to your financial situation?

Unfortunately, people find themselves in that sad situation because they made one wrong assumption. Many people believe that a spouse or other family member is legally entitled to decide whether they are kept alive by artificial means. *You* are the only person who can make that decision and, if you become incapacitated, your life or death wishes may not matter. The State could deem you incapable of making a clear judgment, thereby taking matters into its own hands-and out of yours.

A Living Will *does not* direct someone to end your life. It gives someone the power to make sure that *your wishes* are *honored*. Decisions about what to do if your heart stops, how long you want to be on a respirator, and issues around feeding and hydration are difficult. It's better if you make them, however, and not someone who doesn't know or understand how you wish to live.

Living Wills are binding in some states and not in others. But even in non-binding states, a Living Will gives your loved ones some leverage with doctors and hospitals about you and your medical treatment. Most importantly, you need to discuss your Living Will with those family members who will be involved in the decision-making process. Make sure that everyone understands and respects your wishes. Having your family battle each other in court over how you will live or die makes a painful situation even worse.

The Importance of Investments

As a young-onset PD patient, you can probably look forward to many productive years. During those years you'll remain a productive wage earner with many opportunities to invest for your future. Investment counseling, either through a broker or on-line, is easily accessible.

For good or for bad, investments can play a key role in the health of your overall financial plan. Do your homework. Stay away from highly speculative ventures. Concentrate more on investments with solid returns over time. Stocks, bonds, mutual funds, C.D.'s, money market accounts, and real estate are all possible choices. Basically, there are just two things to remember: ***Know your own financial situation and how much risk you can take, and know your financial advisor***. A good advisor will take a hard look at your financial situation up front, and may suggest ways to get your finances in good shape before they suggest specific investments. For example, they might recommend that you consolidate your debts and lower your monthly payments before you invest.

A Final Word Plan

Once it's too late, it's too late. *Now* is the best time to put together a financial plan you can live with for years to come. To begin, here's a list of things to consider-and to do.

- ¥ Take a careful look at where you are today financially. Look at your income, your debts, and your assets.

- ¥ Imagine how your income and assets will change in the next five years? Is there a way to lower or better manage your debts?

- ¥ Wills and trusts-the most important tool for protecting your family and assets. You probably should have a will (or perhaps a trust) even if your estate is small.

- ¥ Financial planning-make sure the correct person receives the proceeds from your pension, annuity, or IRA.

- ¥ Do you have a Power of Attorney?

- ¥ Do you have a Living Will?

- ¥ Do you have disability insurance? What benefits and services does it provide if you become disabled?

- ¥ Do you have life insurance? Make sure you have enough assets to provide adequately for those who are financially dependent on you.

- ¥ Documentation is essential if you need to apply for SSD and SSI benefits. Do you have a safe, central place for your important papers? (Birth Certificate/proof of age, mortgage or renters information, bank documents, tax forms, names and addresses of your doctors and

other healthcare providers, a brief written employment history.)

- ¥ Do you know the location of the Social Security office near you?

- ¥ Housing alternatives-make planning for your housing needs as you grow older part of your estate plan.

Getting a financial plan together is a way of getting your life together. Now more than ever, you need to think about keeping-and growing-your income. After all, income is much more than money. It supports your lifestyle, gives you and your loved ones security, and (best of all) helps make your dreams come true.

BEYOND DISABILITY BENEFITS

People applying for disability benefits also should consider related issues that can be grouped under the heading of "estate planning." What is estate planning? The Colorado Bar Association says it is: "a written expression of how you want your assets to be owned, managed and preserved during your lifetime and how you want them disposed of upon your death. Your attorney often designs it to incur the least possible taxes and other costs."

How do you find an estate planning lawyer? A good way is to contact the lawyer referral service of a state or local bar association in your area. Ask for an experienced estate planning lawyer. Bar association names and addresses normally are found in the "yellow pages" of telephone directories. Also, Internet users can go to the American Bar Association web site, which includes a national directory of lawyer referral services: www.abanet.org/referral/home.html

See also the Parsons Internet web site, a legal software publisher: *www.itslegal.com/infonet/estates/estatemain.html.*

APPENDIX A
PD MEDICATIONS COMMONLY USED IN THE USA

Generic Name	Trade Name¤
Dopamine Replacement:	
carbidopa/levodopa	Sinemet
controlled-release arbidopa/levodopa	Sinemet CR
Dopamine Receptor Agonists:	
bromocriptine	Parlodel
pergolide	Permax
pramipexole	Mirapex
ropinirole	Requip
COMT Inhibitors:	
entacapone	Comtan
tolcapone	Tasmar
MAO-B Inhibitors:	
selegiline	Eldepryl
Anticholinergic Drugs:	
benztropine	Cogentin
trihexiphenidyl	Artane
Other Drugs:	
amantadine	Symmetrel

APPENDIX B
GLOSSARY

Italicized words are cross-referenced in the Glossary.

acetylcholine - a *neurotransmitter.*

agonist - see *dopamine receptor agonists*.

amantadine - an antiparkinson medication; it may be used early in the disease or added to *levodopa*; has recently been found to be helpful in treating *dyskinesias.*

anticholinergics - a class of antiparkinson medications that are mostly useful for *tremor.*

atypical parkinsonisms - disorders related to PD in that they are characterized by *bradykinesia* and sometimes *rigidity, tremor,* and balance problems, but have other clinical features and other *pathology.*

autonomic nervous system - a part of the nervous system responsible for control of bodily functions that are not consciously directed; for example, heart beat, blood pressure, sweating, intestinal movements, temperature control.

basal ganglia - The interconnected cluster of nerve cells that coordinate normal movement, made up in part by the *substantia nigra, striatum, globus pallidus,* and *subthalamic nucleus.*

blepharospasm - forced closure of the eyelids.

bradykinesia - literally, "slow movement"; one of the main symptoms of PD.

bromocriptine - a *dopamine receptor agonist*.

carbidopa - a drug, used with *levodopa*, to block the breakdown of *levodopa* to *dopamine* in the intestinal tract and in the blood.

carepartner - caregiver; spouse, child, sibling, or other family member or friend who participates in the care of and for a patient with PD.

catechol-O-methyltransferase (COMT) - an enzyme that breaks down *dopamine* at the *dopamine receptor* in the brain and that breaks down *levodopa* in the intestinal tract.

catechol-O-methyltransferase (COMT) inhibitors - a new class of antiparkinson drugs that blocks the enzyme COMT preventing the breakdown of *levodopa* in the intestinal tract by blocking intestinal COMT, thus allowing more levodopa to cross into the blood and then into the brain; examples are *tolcapone* and *entacapone*.

chorea - jerky, random, dance-like, involuntary movements, usually seen in PD from too much medication (see *dyskinesias).*

cognitive function - the ability to think, to remember, to plan, and to organize information.

COMT - see *catechol-O-methyltransferase*.

controlled - release levodopa-a formulation of levodopa that is released more slowly in the gut than the standard preparation, and thus lasts almost (but not quite) twice as long as the standard preparation does for any given patient.

deep brain stimulation - electrical stimulation of cells in the basal ganglia instead of destroying as a treatment for tremor (see *thalamic stimulation*) or other signs and symptoms of PD (see *pallidal stimulation* and *subthalamic stimulation) .*

delusions - erroneous beliefs that cannot be altered by rational argument.

dementia - a progressive decline in mental functions.

diffuse Lewy body disease - PD that has spread to include many parts of the brain and usually is characterized by *parkinsonism, dementia,* and *hallucinations.*

dopamine - the primary chemical messenger of the *basal ganglia*; it is reduced in PD.

dopamine receptor - the area of the nerve cell in the *striatum* that receives the *dopamine* message from the *substantia nigra.*

dopamine receptor agonists - synthetic compounds that mimic the action of dopamine at the dopamine receptor in the *striatum*; examples are *bromocriptine, pergolide, pramipexole*, and *ropinirole.*

dysautonomia - abnormalities of the *autonomic nervous system*, which include such automatic functions as sweating, temperature regulation, blood pressure, urination, bowel movements, and penile erection.

dyskinesias - abnormal involuntary movements, usually associated with too high levels of antiparkinson medication.

dysphagia - difficulty with or abnormality of swallowing.

dystonia - in PD, tightness, spasm, or cramping of muscles; may also involve twisting or posturing of muscles.

end-of-dose failure - a loss of benefit from a dose of *levodopa*, typically at the end of a few hours.

etiology - the cause of a disease, or how it is acquired.

entacapone - a *catechol-O-methyltransferase (COMT) inhibitor.*

enzyme - a protein or chemical tool that speeds up the rate of a biological reaction; *MAO-B* and *COMT* are enzymes that break down *dopamine.*

festination - slow, small, shuffling steps.

freezing - inability to move or getting "stuck," as with the feet appearing to be glued to the floor.

gait - the manner in which a person walks.

globus pallidus - a part of the basal ganglia; the ***internal*** part of the globus pallidus is what is targeted by *pallidotomy* to treat PD.

hallucinations - false perception of something that is not really there. In PD, they are usually things or people patients see *(visual hallucinations*), but occasionally things they may hear (*auditory hallucinations*) or feel (*tactile hallucinations*).

high-dopa dyskinesias - abnormal movements that occur when the *levodopa* in the blood is at its highest level.

hypomimia - the mask-like expression typical of PD.

levodopa - the chemical precursor of *dopamine* and the most effective treatment for PD.

Lewy body - the spherical marker seen in the dopamine-producing nerve cells of the *substantia nigra* indicating a damaged and dying cell; the pathologic hallmark of PD.

low-dopa dyskinesias - abnormal movements that occur when doses of *levodopa* are wearing off, or when the *levodopa* in the blood is at a low or falling level.

mentation - mental or *cognitive function*.

micrographia - the very small handwriting seen in PD.

monoamine oxidase - B (MAO-B)-an enzyme that breaks down *dopamine* in the area of the *dopamine receptor*.

monoamine oxidase-B (MAO-B) inhibitors - a class of antiparkinson drugs (for example, *selegiline*) that blocks the enzyme *MAO-B*, preventing the breakdown of *dopamine* in the area of the *dopamine receptor*.

motor fluctuations - the complications of the treatment of PD affecting ability to move; examples are *wearing-off of dose, on-off phenomena*, and *dyskinesias.*

neurotransmitter - a chemical messenger; *dopamine* is a *neurotransmitter*.

norepinephrine - a neurotransmitter.

off - the state of re-emergence of parkinsonian signs and symptoms when the medication's effect has waned.

on - improvement in parkinsonian signs and symptoms when the medication is working optimally.

on-off phenomenon - unpredictable, usually abrupt oscillations in motor state.

palilalia - stuttering or stammering speech in PD.

pallidal stimulation - electrical stimulation of cells in the *internal globus pallidus* instead of destroying them as a treatment for the symptoms of PD and *dyskinesias*.

pallidotomy - surgical destruction of a small group of cells in the *internal globus pallidus*, the major area from which information leaves the *basal ganglia*, most effective in relieving *dyskinesias* and other symptoms of advanced PD.

paranoia - an irrational belief that others are "out to get" an individual, making the patient suspicious and untrusting.

parkinsonian syndromes - disorders related to PD in that they are characterized by *bradykinesia* and sometimes *rigidity, tremor*, and balance problems, but have other clinical features and other pathology. They are sometimes called "Parkinson plus" or "atypical parkinsonisms."

parkinsonism - the motor picture that makes up PD: **bradykinesia, rigidity, tremor**, balance and *gait* problems.

pathogenesis - the abnormal processes in the body that produce the signs and symptoms of a disease.

pathology - the study of a disease process, including what is affected and what it looks like under a microscope.

pergolide - a *dopamine receptor agonist*.

pramipexole - a *dopamine receptor agonist*.

propulsion - propelling forward as the patient accelerates with rapid, short steps.

psychosis - a mental syndrome in which the patient loses contact with reality; psychotic manifestations include *delusions, hallucinations*, and *paranoia*.

retropulsion - stumbling or falling backwards.

rigidity - a tightness or increase in muscle tone at rest or throughout the entire range of motion of a limb; it may be felt as a stiffness by the patient.

ropinirole - a *dopamine receptor agonist.*

seborrhea - excessive oily secretions of the skin, particularly on the forehead and scalp, causing a flaky, red, itchy condition.

selegiline (deprenyl®) - an antiparkinson medication, it inhibits one of the enzymes (*monoamine oxidase*, or *MAO-B*) that breaks down *dopamine*; it may be used alone as a first-line treatment or in addition to *levodopa*.

serotonin - a *neurotransmitter.*

sialorrhea - drooling.

striatum - part of the *basal ganglia* circuit; it receives connections from the *substantia nigra* and contains the *dopamine receptors*.

substantia nigra - meaning "dark substance," the part of the *brainstem* that

produces *dopamine* and that degenerates in PD.

subthalamic nucleus - a part of the *basal ganglia*; it is targeted by *subthalamic stimulation* to treat PD.

subthalamic stimulation - electrical stimulation of cells in *subthalamic nucleus* instead of destroying them as a treatment for the symptoms of PD and *dyskinesias*.

thalamic stimulation - electrical stimulation of cells in the *thalamus* instead of destroying them to treat *tremor*.

thalamotomy - surgical destruction of a small group of cells in the *thalamus*, a major area of the brain that receives information from the *basal ganglia*, to abolish *tremor* on the side of the body opposite the surgery.

thalamus - a part of the brain that receives information from the *basal ganglia*.

tolcapone - a *catechol-O-methyltransferase (COMT) inhibitor.*

tremor - rhythmic shaking, usually of the hand (but also may affect the leg, lips, or jaw), that occurs at rest in PD. In PD, It may occur less commonly on holding up the hands (*postural or sustention tremor*) or when moving a limb (*action tremor*).

wearing off - a loss of benefit from a dose of levodopa, typically at the end of a few hours.

Please contact the nearest I & R Center for information regarding Support Groups and Chapters or call the National Office at 1-800-223-2732 You can also dial the toll free number 1-888-400-2732 to contact the I & R Center closest to you.

APDA Information and Referral Centers

Alabama, Birmingham
University. of Alabama at Birmingham
205-934-9100

Arizona, Tucson
University of Arizona
520-326-5400
800-541-4960

Arkansas, Hot Springs
St. Joseph's Regional Health Center
800-407-9295
501-318-1690

California, Fountain Valley
Orange Coast Memorial Medical Center.
714-378-5022
877-610-2732

California, Laguna Hills
Saddleback Memorial Medical Center
1-877-610-2732
1-714-378-5022

California, Long Beach
Long Beach Memorial Medical Center
1-877-610-2732
1-714-378-5022

California, Los Angeles
Cedars-Sinai Health System
310-423-7933

California, Los Angeles (U.C.L.A.)
Reed Neurological Research Center
310-206-9799

California, San Diego
Information & Referral Center
858-273-6763

California, Stanford
Stanford University Medical Center
650-724-6090

Connecticut, New Haven
Hospital of Saint Raphael
203-789-3936

Florida, Jacksonville
Mayo Clinic, Jacksonville
904-953-7030

Florida, Pompano Beach
North Broward Medical Center
800-825-2732
954-7344, 954-786-7316

Florida, St. Petersburg
Edward White Hospital
727-898-2732

Georgia, Atlanta
Emory University School of Medicine
404-728-6552

Idaho, Boise
St. Alphonsus Medical Center
208-367-6570

Illinois, Chicago
Glenbrook Hospital
847-657-5787

***The Arlefte Johnson Young Parkinson Info. & Referral Center**
Glenbrook Hospital
800-223-9776 (Out of IL.)
847-657-5787

Louisiana, New Orleans
School of Medicine, LSU
504-568-6554

Maine, Scarborough
Maine Medical Center
207-885-7560

Maryland, Baltimore
John Hopkins Outpatient Center
410-955-8795

Massachusetts, Boston
Boston University School of Medicine
617-638-8466

Minnesota, Minneapolis
Abbott Northwestern Hospital
Minneapolis Neuroscience Institute
612-863-5850
888-302-7762

Missouri, St. Louis
Washington University Medical. Center
314-362-3299

Montana, Great Falls
Benefis Health Care
406-455-2964
800-233-9040

Nebraska, Omaha
Information & Referral Center
402-397-2766

Nevada, Las Vegas
University of Nevada School of Medicine
702-464-3132

Nevada, Reno
V.A. Hospital
775-328-1715

New Jersey, New Brunswick
Robert Wood Johnson University Hospital
732-745-7520

New Mexico, Albuquerque
HEALTHSOUTH Rehabilitation Hospital
800-278-5386
505-344-9478 Ext. 5099

New York, Albany
The Albany Medical College
518-452-2749

New York, Far Rockaway
Peninsula Hospital
718-734-2876

New York, Manhattan
New York University
212-983-1379

New York, Old Westbury
NY College of Osteopathic Medicine
516-626-6114

New York, Smithtown
St. Catherine's of Siena Hospital
631-862-3560

New York, Staten Island
Staten Island University Hospital
718-226-6129

North Carolina, Durham
Duke University Medical Center
919-668-2938

Ohio Cincinnati
University of Cincinnati Medical Center
800-840-2732
513-558-6770

Ohio Cleveland
The Cleveland Clinic Foundation
216-445-8480

Ohio Cincinnati
University of Cincinnati Medical Center
513-558-6770
800-840-2732

Oklahoma, Tulsa
Hillcrest Medical Center System
918-747-3747
800-364-4450

Pennsylvania, Philadelphia
Crozer-Chester Medical Center.
610-447-2911

Pennsylvania, Pittsburgh
Allegheny General Hospital
412-441-4100

Rhode Island, Pawtucket
Memorial Hospital of RI
401-729-3165

Tennessee, Memphis
Methodist Hospital
901-726-8141

Tennessee, Nashville
Centennial Medical Center
615-342-4635
800-493-2842

Texas, Bryan
St. Joseph Regional Rehab. Center
979-821-7523

Texas, Dallas
Presbyterian Hospital of Dallas
214-345-4224
800-725-2732

Texas, Lubbock
Covenant Hospital
806-785-2732
800-687-5498

Texas, San Antonio
The University of Texas HSC
210-567-6688

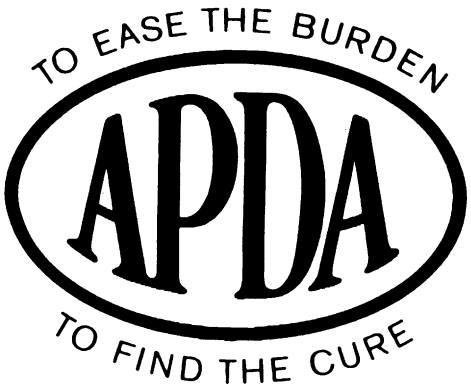
Utah, Salt Lake City
University of Utah, School of Medicine
801-585-2354

Vermont, Burlington
University of Vermont
802-847-3366
888-763-3366

Virginia, Charlottesville
University. of Virginia Medical Center
804-982-4482

Washington, Seattle
University of Washington
206-543-5369

Wisconsin, Neenah
The Neuroscience Group of
Northeast Wisconsin
920-725-9373
888-797-2732



The American Parkinson Disease Association, Inc.
1250 Hylan Boulevard - Suite 4B
Staten Island, New York 10305-1946
800-223-2732

www.apdaparkinson.org

APDA West Coast Office
10850 Wilshire Boulevard, Suite 730
Los Angeles, CA 90024
800-908-2732