Welcome to the special double-length summer issue of Maine PD News. There was so much to cover that we went BIG.

.

Visit us online for a searchable article database, coming events, and a movement disorder doctor directory.

Inside:

Science is spoken here.

Maine PD News

Summer, 2016 - Edition 2 www.mainepdnews.org

Psychosis in Parkinson disease

by Markos Poulopoulos, M.D.

Introduction

James Parkinson, in his pivotal work "An Essay on the Shaking Palsy" (published in 1817), was the first to recognize the condition that later was named after him. He defined it as "involuntary tremulous motion, with lessened muscular power, in parts not in action and even when supported; with a propensity to bend the trunk forwards: the senses and intellects being uninjured." He then noticed that "the sleep becomes much disturbed," "the bowels, in most cases, demand stimulating medicines of very considerable power," and "the urine is passed involuntarily; and at the last, constant sleepiness, with slight delirium, and other marks of extreme exhaustion" (1).

...continued, p 2

An Interview with the neurosurgeon – Anand Rughani, M.D.

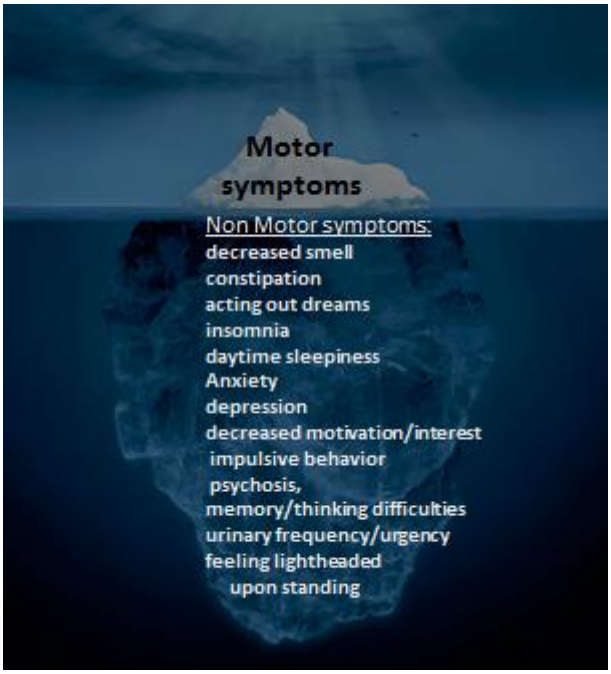
with Bill Stamey, M.D.

As part of a pair of articles on the topic of deep brain stimulation (DBS) in this issue, I met with Dr. Anand Rughani recently to ask a few questions. I hope these articles will give readers a little insight into what it might be like going through the procedure. I also wanted people to know a little about Dr. Rughani, and why we are so lucky to have him here in Maine, not just for the high level of skill he brings with his subspecialty training in functional neurosurgery, but also a bit . about the man and his motivations. This interview is not meant to be exhaustive, but focused on a few relative points.

…continued, p5

Psychosis, continued…

Despite his above observations, little attention was given to the non-motor manifestations of Parkinson disease up until the last 10-15 years. For quite a long time, the scientific focus was heavily on the motor symptoms, which are still used for making the diagnosis (tremor, stiffness/rigidity, slowness/bradykinesia and gait instability). One could say that we were only looking at the tip of the iceberg.



But there is so much more than the physical symptoms, other difficulties that are far more difficult to deal with and cry for more vigorous research and effective treatment development. Psychosis is perhaps the most problematic and intrusive constellation of symptoms, and the subject of this discussion.

What do we mean by psychosis?

Psychosis in Parkinson disease is defined (2) as the presence of at least one of the following symptoms:

- *Illusions* - This is when you mistake a real object for something else.
- *False sense of presence passage (hallucinations)* - This when you think there is somebody behind you, or a vague figure is quickly passing by your side. However, when you turn your head to look at it, there is nothing there.
- *Hallucinations* - This is when you see (less often hear, smell or feel) people or animals typically in front of you that clearly do not exist, and there is no other real object to be mistaken for what you experience.
- *Delusions* - This pertains to unusual, disruptive beliefs or ideas, usually of a paranoid nature, e.g. persecution, theft, infidelity.

We need to highlight the importance of early recognition and treatment of even benign aspects such as illusions. There is about an 80% chance for these to progress to more complex and persistent symptoms such as hallucinations and delusions, which in turn can be very difficult to treat and have a significant impact on your life and that of your family. If psychosis gets out of hand, it is usually a reason for nursing home placement, and results in an increased rate of other complications, including decreased survival. For example, one may lose insight and firmly believe that the hallucinations (people or animals) are real, react by attacking them or running away from them, and subsequently fall and break a hip. These events can lead to a domino effect with potentially serious complications.

Hence, knowing the nature of psychosis will help the doctors, patients, and care givers to ask pertinent questions in search of subtle signs that would otherwise go unnoticed. It is always better to treat early, rather than late.

How frequent is psychosis?

Overall, psychosis can be present in up to 60% of Parkinson disease patients at some point in time. The more fearsome aspect is visual hallucination, which can be present in 7-25% of patients with Parkinson disease. However, if we consider only patients with dementia (so-called Parkinson disease dementia), hallucinations have a frequency from 40 to 80% (3). *…continued, p 4*

Psychosis, continued…

Psychosis is more likely to happen when a patient with Parkinson disease also has dementia. However, it is also well documented in patients with Parkinson disease without dementia. In this situation, psychosis can happen in 20% of patients. In more detailed breakdown, frequency of visual hallucinations is about 13%, auditory hallucinations 7%, illusions 7%, and paranoia 5% (4).

Other non-motor manifestations of Parkinson disease that sometimes predict the development of psychosis are REM sleep behavior disorder (acting out the dream content) and depression/ anxiety.

Do medications play a role? Yes and no.

The answer is yes for most medications, and debatable for carbidopa/levodopa. Although the common practice is to try to reduce the dose or stop medications in hopes of decreasing or stopping the psychosis (see management section), there is evidence that psychosis can happen before the start of any treatment with such medications. It is intriguing that psychosis may happen in up to 40% of drug-naïve patients, as opposed to 5% of individuals without Parkinson disease. In this scenario the good news is that insight is almost always retained, and the type of psychosis is by and large a simple sense of presence or feeling that somebody is passing by (rarely visual hallucination) (5).

What can we do? How can we treat it?

- *Knowledge* - First of all, knowledge of the nature of psychosis, being able to recognize and communicate its existence without guilt or fear, is paramount and the starting point.
- *Search for triggers* - The doctor should work with the caregiver and the patient in an attempt to identify potential triggers such as infections, dehydration, insomnia, malnutrition, new medications/dose escalation, home/environmental changes, bereavement, and exacerbation of depression.
- *Ideally, a team approach* - Behavioral care, case management, and physical, speech, language, and occupational therapy aiming at an individualized treatment plan to relieve distress, provide direction, promote adaptation, and optimize quality of life.
- *Decrease or stop medications for Parkinson disease* (3) - Typically, we wean off, or at least decrease the usual culprits with first and foremost the dopamine agonists (e.g. pramipexole/Mirapex, ropinirole/Requip), anticholinergics (e.g. trihexyphenidyl/Artane, amantadine), and other medications we may use to treat Parkinson disease. Carbidopa/levodopa (Sinemet) is the last medication that should be decreased, and certainly never stopped. There is no clear evidence that this medication can definitely aggravate psychosis. Easily said, but it is usually difficult to implement the above, since decreasing anti-Parkinson medications will lead to worsening of the physical performance (tremor, gait, balance, and overall movement).
- *Add medications that mitigate or stop psychosis* (brand name in parentheses) (3)

➢ rivastigmine (Exelon): In a 24-week, prospective, placebo-controlled trial, this medication, designed as a memory enhancer, both improved memory and decreased hallucinations (6).

➢ quetiapine (Seroquel): The evidence is rather equivocal in favor of this medication directly improving visual hallucinations based on studies (7). However, it is widely used in clinical practice with good

…continued, p 5

results based on anecdotal and personal experience. The main reasons are the ease of use and titration, and the favorable side effect profile (compared to all the other antipsychotics, it has the least potential for increased mortality).

- clozapine (Clozaril): This medication has, so far, the best evidence with respect to efficacy reducing visual hallucinations (8). However, frequent blood draws, the rare but very significant danger of reducing white blood cells that fight infections, frequent drowsiness, weight gain, and dizziness make it not the first choice for most doctors.

New treatment

Pimavanserin (Nuplazid) is the latest medication tested for treatment of visual hallucinations in Parkinson disease.

Based on a 6-week trial (randomized, double-blind, placebo-controlled) on a total of 200 patients (the largest number tested compared with the other medications listed above, apart from the rivastigmine trial), it did produce a statistically significant reduction in hallucinations, improved night time sleep, and decreased daytime sleepiness (9).

Furthermore, pimavanserin did not aggravate the motor symptoms of Parkinson disease and was overall well tolerated, probably better than quetiapine and clozapine, by extrapolation. This is likely due to a pharmacological action that differs from the other antipsychotic medications. All of the others block the various dopamine receptors in the brain and typically worsen Parkinson disease symptoms, since the levodopa (which converts into dopamine in the brain) cannot act on the dopamine receptors because they are occupied (blocked) by the antipsychotics. Pimavanserin does not act on dopamine receptors. Rather, it acts on serotonin receptors (specifically 5-HT2a). Pimavanserin may cause leg swelling (7%), nausea (7%), confusion (6%), and constipation (4%).

The pressing need for better treatments for psychosis in Parkinson disease led to FDA discussion for early approval on May 1, 2016

In conclusion, it is very important for patients and caregivers to report to the doctor symptoms in keeping with psychosis, so that we can search for triggers (as mentioned above), monitor symptom evolution and prevent/treat, if possible.

References (for full citations, http://mainepdnews.org/2016/06/06/psychosis-in-parkinson-disease/)

1. James Parkinson. *An Essay on the Shaking Palsy*. Neuropsychiatric classics. 1817.
2. Movement Disorders, Vol. 22, No. 8, 2007, pp. 1061–1068.
3. Movement Disorders, Vol. 29, No. 5, 2014. Pages: 651-662.
4. Movement Disorders, Vol. 27, No. 7, 2012. Pages: 858-863.
5. Movement Disorders, Vol. 31, No. 1, 2016. Pages 45-52.
6. Movement Disorders 2006; 21:1899-1907.
7. Movement Disorders 2005; 20:958-63.
8. NEJM 1999;340:757-63.
9. Lancet 2014,(8);383:533-540.

An interview with the neurosurgeon-Anand Rughani M.D., continued from page 1…

BS: I think a good place to start would be in explaining why you are interested in deep brain stimulation. What drew you to this specific area of medicine?

AR: My pathway to neurosurgery started off with an interest in neuroscience and in understanding the mind-brain interaction. The connection between mind and brain is most visible to me on a daily basis in neurosurgery, and specifically in the area of functional neurosurgery, which deals with mapping the functional areas of the brain to understand how different brain structures lead to different experiences and behaviors. We see that through disease, and in treating some of those diseases, we get insight into how the brain works. That's what lead me to neurosurgery to begin with and then specifically, functional neurosurgery and deep brain stimulation. Deep brain stimulation itself is a good example of how you see those interactions in a very real way, through implanting electrodes in an awake patient to treat symptoms from various diseases. So, I gain insight on a daily basis as to how the brain leads us to be who we are, and how we work.

BS: Your training started out with undergraduate college, then medical school, then neurosurgery residency. After this, you had a fellowship in functional neurosurgery. Can you tell us about the fellowship?

AR: My fellowship at the University of Toronto offered exposure to a deep enthusiasm for not just doing surgery or DBS for some of the more conventional indications, but really exploring new indications for surgery. Whereas the FDA currently has indications for Parkinson's disease, tremor, and dystonia, I was also able to be involved in cases for anorexia, for OCD, depression, and for Alzheimer's disease.

BS: What brought you to Maine?

AR: I grew up in Eugene, Oregon, but then spent ten years in Montreal for my Bachelor's degree and medical school, and each year that I spent there I grew more nostalgic for the idea of ending up in a small coastal town. However, the more time I spent in Montreal and in my subsequent training, the more I found myself doing increasingly subspecialized training. First, it was neuroscience, then it became medicine, then it became neurosurgery – and there are only so many places you can do that. Then, even more subspecialized was functional neurosurgery. In the process of that training I got to spend a year at Maine Medical Center, and it turned into the ideal opportunity because I could offer surgeries that weren't previously available in Maine.

BS: Can you walk through what it is like to have the DBS procedure in the operating room?

AR: Deep brain stimulation is a significant undertaking. We do it in a couple different steps, but the most important is the step where we implant the electrodes in the brain. There are a couple of different ways of doing that now. The traditional way of doing that is recording from the neurons in the brain and then stimulating through those electrodes. The way we do that is with the patient awake, and most patients do fantastically well. People are comfortable because we use a local anesthetic, but the surgery itself takes a couple hours. The hardest parts of the operation are probably a couple of things: one is for patients

…continued,p6

An Interview, continued…

with Parkinson's disease being off of their medication. That can be quite challenging. Another part is being stuck in one position for two, or three or four, hours. The local anesthetic can burn quite a bit when it goes in. I think the dentist. The drilling takes about 30 seconds. After, some patients may have a mild headache, but there are no pain receptors in the brain, so no discomfort from the next steps.

BS: Tell me a little about the risk of the procedure.

AR: I categorize the risk of surgery into four areas. First, there is the risk of the operation itself. This includes things like bleeding into the brain, which can happen in about 2% of patients, and can lead to new symptoms in about 1% of patients. The second category of risks are related to having implanted hardware, for example a delayed infection could occur, or a disconnection or other device complication can occur. These types of events probably occur in up to 10-15% of patients over the years. The third category of complications that I think of are the stimulation-related

actual operation where we do the work isn't the biggest source of discomfort, it's more from being stuck in one position for a couple hours and being off of regular medications for those with Parkinson's disease.

complications. We try to minimize these by doing testing in the operating room with patients awake. These depend on the location where the electrode is implanted, but could consist of changes in speech, or mild tingling sensations. The last category of complications are the medical complications, for example things like developing a pneumonia, or blood clot in the leg, or a heart attack.

BS: Who is in the room while the surgery is taking place?

AR: The operating room is busy for this surgery. In addition to the patient and surgeon, there is also the anesthesiologist, or nurse anesthetist, there is an operating room nurse, and there is a scrub tech that passes the sterile instruments. There is also a neurologist, and usually one or two additional people involved in the surgery.

BS: People complain about the sound of the drill a lot too.

AR: I think the drill is probably like being at the dentist, or an aggressive version of being at the

BS: What else would you want people to know about DBS?

AR: It is really important to have a good understanding of the goals of surgery, and the surgery process. Specifically, it's important to know which symptoms are most likely to respond to surgery, and which aren't. With that in mind, and a good understanding of the involvement and risks of surgery, patients and their families can make the best decision about whether or not it is right for them. I am always happy to discuss the surgery in detail with patients who think they might want to consider it.

Dr. Rughani practices with Maine Medical Partners - Neurosurgery & Spine in Scarborough.

The role of the speech pathologist in the treatment of speech, voice, and swallowing disorders associated with PD

by Yonca Berk-Giray, SLP

Eighty-nine percent of individuals with PD have speech and voice difficulties at the onset of the disease. As the disease progresses, this number rises to 100%. Most individuals with PD also experience varying degrees of swallowing difficulties.

Speech and voice difficulties are often overlooked by the patients and their physicians until the problem becomes severe enough that communication is affected. Many patients with PD experience difficulty socializing and being understood by others, and feel left out of conversations. Individuals experience decreases in vocal loudness levels, speech intelligibility, expression in their speech and voice, facial expression, as well as hoarse voice quality. Since these changes happen slowly over many years, most patients are not aware of them. Often it is their loved ones who first become aware of the communication issues.

Yonca Berk-Giray, SLP teaching a workshop at the YMCA, Freeport

Lee Silverman Voice Therapy (LSVT) is a voice and speech therapy program that is specifically designed for treating individuals with PD. It has been scientifically researched for the past 25 years and proven to be effective in treating voice, speech and communication difficulties associated with PD. LSVT follows a standardized treatment protocol that is customized to the needs of the individual. Mid Coast Rehabilitation Services currently has two clinicians who are certified in delivering this program. For those patients who are homebound, the therapy is also available through certified clinicians at CHANS home health services. For more information on LSVT Loud, and a list of certified clinicians, please see www.LSVTGlobal.com.

Swallowing difficulties associated with PD are much more subtle at the onset of disease, and can become serious and life threating as the disease progresses. Patients usually complain of drooling, foods getting stuck in the cheeks or throat, coughing with liquids or solids during meals, and difficulty taking their pills. Two main concerns with swallowing difficulties are the risk of aspiration related pneumonia and weight loss. Recognizing early signs of swallowing difficulties, and receiving treatment for them, is very important in staying ahead of the muscle atrophy that can happen. There are various methods and treatments available that are proven to be effective in treating swallowing difficulties in PD. See a physician or speech pathologist for more information.

It is important and strongly recommended that patients are proactive in receiving treatment early for speech, voice and swallowing difficulties to maximize their functioning for years to come.

Yonca Berk-Giray, SLP practices with Mid Coast Hospital Rehabilitation Services in Brunswick, Maine

Balance in Parkinson disease

by Bill Stamey, M.D.

Balance is a nearly universal issue with PD patients. And, whether it comes along early in the course of disease, or late, as it is more likely to do, it is most often a progressive problem. Bad balance can of course lead to falls. Falls lead to nothing good. This is why I always encourage people with PD to start working on balance early, even if it is not yet a problem. To know why, you should understand a little about what is going on with balance in PD.

We humans are in a precarious situation in the first place. Since we stand on only two limbs, the simple act of walking is one in which we constantly throw ourselves forward in a form of controlled falling. One step after the next propels us into space and we land in what feels like an unbroken motion to keep going. This all requires a very complex onboard system to remain upright. Our eyes give the parietal lobe of the brain a visual map of the three dimensional space. Our inner ears contain semi circular canals in which fluid filled spaces lined with tiny hair cells that click on and off like relay switches, sense the direction our heads move and feed that information back to the brainstem. Tiny nerves in the feet (and the rest of the body for that matter) send signals to the posterior column of the spinal cord about where our body parts are in space. The posterior column climbs to the brainstem and fibers converge on the cerebellum, the true master of balance, which monitors, and in many ways, governs the whole system. Sometime you have tripped over an uneven surface, maybe a root on the ground, and before you knew what happened, your feet corrected themselves in a sudden move. You had no time to react, no time to think about it, and yet reflexively, you stayed upright. You probably gave yourself a little pat on the back for being so cat-like, but that was your cerebellum at work behind the scenes, beneath conscious thought, and in a fraction of a second correcting a footfall to stop disaster. To a neurologist, it is a glimpse behind the curtain of the hidden brain.

There are also automatic procedural patterns of movement that are sometimes referred to as habit learning, such as walking or riding a bike – really any set of movements you had to train yourself to do. These are, for the most part, stored in your brain's striatum, and are unique to you, part of the reason PD is unique to the person. The striatum is particularly important because it is the input of the basal ganglia (BG), into which dopamine is delivered from its source in the midbrain. When there is low dopamine, some automatic patterns fail. This is how people with PD lose their normal gait pattern in what Mayo neurologist Harry Lee Parker, M.D., described in the pre-dopamine drug era of the 1950's this way: "One recognizes the loved one's step before arrival. In this disease all is leveled to a sterile similarity" (1).

One problem with gait and balance can be explained by dopamine tone. If you think about the BG as a store of dopamine, with connections to its own parts and other brain regions, the tone, or flow of dopamine is important. In other words, dopamine may be thought of during wakeful hours as being in a state of flow, similar to water in a pipe, or current in a wire. In this case, the strength or tone of that flow is one way the BG communicates: more dopamine tone may mean "yes, that movement is being done

…continued p9

…Balance, continued

correctly;" whereas less tone may mean "no, that movement was bad." The BG also regulates many of the movements themselves. It is quality control and operator. Thus, the BG might, under normal circumstances, turn down the tone of dopamine messaging to signal a failure of correct movement. If the BG fails to control how high you pick up your feet, how fast you move, or how long your stride is, the abnormal low state of dopamine in the brain forces the basal ganglia to, more or less, tell a lie to the brain. This is how you find yourself walking along, unaware that one foot is not clearing the ground as high as it used to. Your toe makes contact with some uneven or elevated surface in the middle of your stride and your foot abruptly comes to a halt while your body is still in motion. Worse still, your cerebellum does not save you either, because in PD the postural reflex is also not working. Down you go.

The good news is that you may be able to do something about this, and there are some choices. Studies have shown that programmed exercise, dance, physical therapy, yoga, and Tai Chi can all help balance.

While movement disorder neurologists recognize and diagnose gait disorders, physical and occupational therapists are specialists in treating impaired balance and gait dysfunction. In the case of PD, they teach patients to understand the way balance is failing, how to correct, and carry out the movement. If this is practiced enough, it becomes a new pattern, outside of the damaged networks in the striatum. This new neural pathway is reinforced and becomes automatic, similar to learning any new skill. This is why starting early and working on repetition is important. It is not a simple process, but the work pays off. Multiple studies have supported the use of PT for improving balance in PD.

Selina Carey, MS, OTR/L notes that when

seeing a person with PD, "our initial session involves a thorough evaluation. We give the patient a numerical rating on a fall risk, complete at least one other balance assessment such as the Berg or Tinetti balance tests, timed up and go, or several other options. We are able to calculate a numerical score and assess the level of a fall risk that patient may be at pre, during, and post treatment. The great thing is that we are able to see and show the patient how much they have improved from initial measurement." She is also LSVT BIG certified (see the spring issue article "What is LSVT?" for more details), and reports that BIG "works to improve limb and body movement." She and her colleagues tell patients to "think big" and use "big movements," cues to improve the amplitude and speed of movement. "With improved body movement, a person's stability and balance inevitably improves, and therefore carries over in daily routines and patient-centered activities. A patient's balance improves with trunk rotation, stride length, and quality of movement." She opines that "LSVT BIG is highly effective. Patients report and show improvements with balance, stability, and independence completing their chosen functional task. I love seeing how a patient's positive regard improves as the program progresses. Our balance assessments are able to show improvements that are indicated by

…continued, p10

…Balance, continued

higher scores that are taken at initial evaluation, after 10 visits, and at the time of discharge." However, "the LSVT BIG program can be time demanding, and therefore not for everyone." An alternative approach is a structured exercise program tailored to each individual patient. "Therapists can work with a patient to strengthen specific muscles, improve activity tolerance, and endurance as well as improve dynamic and static balance. Specific functional tasks and exercises can be extremely beneficial to a patient who has balance deficits." To prolong the effects of a PT intervention, she urges, "Get exercising now! We really encourage patients to continue exercising and moving. It is highly recommended that patients complete their daily exercises to insure carry over of the progress made with therapy intervention. Incorporating 'bigness' in their everyday life is also very important. A focus in the program is to increase limb movement. We recommend that patients continue to use big movements during their daily routine. Selina Carey is a therapist with Coastal Rehab in Falmouth, Maine, and is the group facilitator for the PD support group at that location.

There are other ways to improve your balance, as has been shown in many studies published in peer-reviewed medical journals. One well-publicized, randomized, controlled trial funded by the National Institute of Neurological Disorders and Stroke, included 195 mild-to-moderate PD patients who were sorted into one of three groups: Tai Chi, resistance training, or stretching. Participants met for an hour, two times a week, over the course of 24 weeks. The Tai Chi group performed consistently better than the resistance-training and stretching groups in terms of maximum excursion, directional control, stride length, and functional reach. Tai Chi lowered the incidence of falls as compared with stretching and was about equal with resistance training. The benefits of Tai Chi training were still present three months after the 24-week period (2). However, a six-month course may not be enough. Generally, the interventions described here should be performed long term. Bill Milan would agree. He has taught Tai Chi at the YMCA in Bath for over eight years, and has been involved in the practice for three decades. He is now also teaching at the Landing YMCA in Brunswick, and notes that in Tai Chi, one has to focus on center of gravity, breathing, centering and making the body posture more upright. This is done through a lot of guidance. Moreover, he notes that it is "not an easy activity. It takes patience, but if one engages, it can really help."

Yoga is another means to better balance. A randomized, controlled, trial of 41 PD patients with an average age of 72, met over a 12-week period to compare the effects of power training and a high-speed yoga program on physical performances, and to test the hypothesis that both training interventions would lessen PD symptoms and improve physical performance. Raters in the study measured standardized PD and balance scales, which were improved significantly, compared with a non-exercise control group (3).

Elizabeth Burd is a Maine Parkinson Society Board member, certified personal and certified Kripalu yoga instructor who has worked with PD patients in Maine since 2006 in various capacities. She finds that high-speed yoga may not

…continued, p11

be ideal for the moderate to advanced PD patient, though in some cases a fast-paced interval training with higher heart rate while using a variety of exercises is helpful. Often, a slower pace is more conducive to good results. Elizabeth has worked with patients at all stages of disease and has found at seminars and PD support group meetings that for patients with advanced disease, especially those on a first foray into yoga, fear of falling may be a major impediment to balance. With a yoga intervention, "the biggest thing I hear is that they feel more confident, which definitely contributes to being able to move around. You're working on balance, core, and leg strength at the same time. When people realize they can practice safely, they are more likely to do it on their own." Still, some patients use a walker or a wheelchair. "I may modify the yoga poses to make them more accessible for everyone. They think, 'I have terrible balance, I can't do that.' Well, here is a way you can." She has produced a 45 minute video, mostly seated home practice, though there are some standing options which help strengthen the legs and core (visit her website for details http://www.pdyogaforme.com/). She also notes, "I also really try to encourage the caregivers, who sometimes don't look after their own health." She is currently a trainer at Natural Fitness in Portland and conducts private yoga training.

Various forms of dance have also been investigated in PD. Though there are several published studies showing improvement of balance with programmed dancing, most are with mild or moderate PD patients. In one study, patients with severe PD who primarily used a wheelchair for transportation were enrolled in a 10-week trial with 20 one-hour tango classes (4) The results included improved balance and balance confidence. Patients also demonstrated increased endurance and reported improved quality of life.

Another popular intervention of late is boxing training. In one study, 31 people with PD were randomly assigned to boxing training or traditional exercise for 24-36 sessions, each lasting 90 minutes, over 12 weeks. Boxing training included stretching, boxing (punching bags but not people), resistance exercises, and aerobic training. Traditional exercise included stretching, resistance exercises, aerobic training, and balance activities. Before and after completion of training, tests were taken of balance, balance confidence, mobility, gait velocity, gait endurance, and quality of life. Only the boxing group demonstrated significant improvements in gait velocity and endurance. Both groups demonstrated significant improvements with the balance, mobility, and quality of life (5).

For more, see the fall issue of Maine PD News, in which Dr. Kleinman will discuss the data on exercise in PD.

References (for full citations, visit http://mainepdnews.org/2016/06/07/balance-in-parkinson-disease/)

1. Clinical Studies in Neurology 1956, p21-24.
2. N Engl J Med. 2012;366(6):511-9.
3. Arch Phys Med Rehabil. 2016;97(3):345-354.
4. Rehabil. 2010;32(8):679-84.
5. NeuroRehabilitation. 2013;32(1):117-24.

What is the risk of melanoma in PD?

by Bill Stamey, M.D.

Melanoma is a dangerous form of skin cancer. The CDC reports that in 2012 almost 68,000 people in the U.S. were diagnosed with melanoma of the skin (1), with a rate of up to 23.6 per 100,000 people in the state of Maine (5). Melanoma has sometimes been associated with other diseases or medications. Increased risk of melanoma in Parkinson disease (PD) was first raised as a concern in 1972 with the report of a PD patient who experienced recurrent bouts of this potentially fatal disease while being treated with levodopa (2). This introduced the question as to whether the case represented a random correlation or an important link between the two diseases. There was also concern of a possible link between levodopa and melanoma because levodopa can convert into melanin, a pigment found both in the dopamine-producing cells of the substantia nigra, and in melanocytes, the cells which can become the tumor of melanoma.

The risk is not clear however. Not all cases of either condition are reported. The best we can do is study the available reports, epidemiology, and disease databases. One case-control study evaluated 862 malignant melanoma cases, compared with 862 age- and sex-matched controls to detect the incidence of PD among melanoma sufferers (3). Among the melanoma patients, 25 (2.9%) had PD. Among the controls, 11 had PD (1.3%). Thus, the odds of having PD was over twofold greater if one had melanoma versus those who did not. A meta-analysis (the combination of multiple studies) using the National Institute of Health (NIH) Medline search engine found that between 1972 and 2010 just over 50 cases of melanoma in PD had been reported in the worldwide medical literature (4). One chart review of 409 PD patients found two with melanoma where 0.3 would have been expected based upon incidence among the same age group (5).

Does levodopa raise the risk of melanoma?

Some authors included information about whether or not people were taking levodopa. A cross-sectional survey (6) to determine the frequency of skin cancers at 12 medical centers in Israel found 9 out of 1,395 patients had biopsy-proven melanoma, 14 had melanoma in their medical history (6 occurring prior to, 8 after PD diagnosis). Twenty patients total (1.4%) had a current or prior melanoma, with an overall rate of melanoma 4.4 times greater than expected for these patients. Melanoma did not correlate with PD duration, PD stage, or the duration of levodopa treatment. In North America, 2,106 PD patients underwent full skin examination with a dermatologist and 346 patients had biopsy of suspicious-looking pigmented skin lesions (7). Twenty biopsies confirmed in situ melanoma (0.95%), and four had invasive melanoma (0.19%). There was no observation of a relationship between levodopa use and melanoma. The DATATOP patient cohort (800 patents entered between 1987 and 1988), followed until 1994, yielded five melanoma cases (1.5 would have been expected, considering the age and gender of the group). Malignant melanoma was diagnosed in two of the patients prior to starting levodopa. The evaluators could not come to any conclusion regarding association between levodopa use and melanoma incidence (8).

…continued, p13

…Melanoma, continued

Although there have been case reports of patients who started levodopa prior to onset of melanoma, there have also been cases in which patients who remained on levodopa had no recurrence or exacerbation of melanoma. And, no formal relationship between the two has been established in the lab. One epidemiological study attempted to determine if levodopa was causative (9). In this prospective study of 1,099 patients from the Melanoma Clinical Cooperative Group, a single patient was taking levodopa at time of diagnosis. The authors found no role for the induction of melanoma. The link is still not clear (10) and levodopa has not been found to be carcinogenic (cancer-causing) otherwise.

A prospective study of 157,036 people without PD at baseline as part of the Health Professional Follow-up Study and the Nurses' Health Study took place over a 14-20 year follow-up (11). 616 cases of PD developed. A history of melanoma in a first-degree relative revealed relative risk of 1.85. The authors concluded that PD and melanoma "share common genetic components," though genes were not identified.

In 2007, some experts considered the ongoing questions and cited the absence of convincing proof of an interaction between levodopa and melanoma, though still offering the following advice: "it would seem prudent not to treat with levodopa if other anti-Parkinson agents remain effective (12)."

In 2009, investigators reviewed five published studies exploring the associations between melanoma, PD, and levodopa (13). They noted the increased risk of melanoma is already present before PD is diagnosed, that as it is unlikely that levodopa plays any role in this phenomenon. The authors noted that while there is a need for further investigation, they also recommended removal of the warning from the drug insert leaflet, noting it might "lead to unnecessary fear on the part of the patients and physician resistance to prescribing this medication."

Still, the current prescribing information for Sinemet from Merck Pharmaceuticals indicates in regards to reported cases of melanoma in PD, "whether the increased risk observed was due to Parkinson's disease or other factors, such as drugs used to treat Parkinson's disease, is unclear….patients and providers are advised to monitor for melanomas frequently and on a regular basis when using SINEMET for any indication. Ideally, periodic skin examinations should be performed by appropriately qualified individuals (e.g., dermatologists)."

Other anti-Parkinson medications

Azilect (rasagiline) is a monoamine oxidase B inhibitor, which boosts dopamine in the brain. The Azilect prescribing information notes, "The increased incidence of melanoma in the Azilect development program was comparable to the increased risk observed in the Parkinson's disease populations examined in epidemiological studies." In the lab, rasagiline has been found to actually decrease melanoma growth, and has been tested as a therapy in lab animals (13).

Requip (ropinirole), Mirapex (pramipexole), Neupro (rotigotine) are dopamine agonists. A PubMed search for each generic name and the term "melanoma" yielded zero citations. Prescribing information of ropinirole is silent regarding melanoma; whereas prescribing information of rotigotine and pramipexole note

...continued, p 14

…Melanoma, continued

that there is a general two- to six-fold higher risk of melanoma in PD but the connection with drugs is unclear.

Summary

The incidence melanoma among PD patients is at least twice that of the general population, though still in the low single digits. And, if one has melanoma, the risk of developing PD is double that of the general population also. Thus, it would seem that there is a common risk or genetic cause for the two diseases, though the link has not been identified. In spite of the fact that levodopa can covert to melanin, the pigment of melanocytes and melanoma cells, there is no proof that levodopa causes or stimulates melanoma growth. The growth of melanoma seems dependent on genes, not the amount of melanin present. Still, some experts suggest it might be prudent to avoid levodopa in the melanoma patient if possible, and some recommend discarding the warning about levodopa and the risk of melanoma altogether.

Dr. Stamey practices at Mid Coast Medical Group Neurology in Brunswick, Maine

References (see full citations at http://mainepdnews.org/2016/06/08/what-is-the-risk-of-melanoma-in-pd/)

1. http://www.cdc.gov/cancer/skin/statistics/
2. Arch Pahol 1972;93:556-61.
3. Movement Disorders 2006;21:S58.
4. Movement Disorders 2010;25(2);139-48.
5. Ann Neurol. 1985;17(5);505-09.
6. J Neural Transm 2011;118(8):1199-207.
7. Ann Neurol 2006;60(Suppl 3):S71-72).
8. Mov Disord 2007;22:720-22.
9. JAMA 1978;240:554-555.
10. Dermatol Surg 2016;42:(2)11-46.
11. Neurology. 2009;73(16):1286-91.
12. Principles and Practice of Movement Disorders 2007, chapter 6,131.
13. Parkinsonism Relat Disord. 2009;15(8):551-3.
14. Cutan Ocul Toxico 2012;31(4):312-17.

Maine PD News is an online quarterly newsletter for Parkinson disease patients and their caregivers in Maine, though we welcome members of the PD community around the globe.
Visit us online for current and prior issues at www.mainepdnews.org
-Bill Stamey, M.D., editor
-Liz Stamey, BSN, RN stylistic and copy editor

The New England Parkinson's Ride

by Bill Stamey, M.D.

Chris Woods was 41 years old when he was diagnosed with PD. His sister, Cindy Woods Theberge, tells me Chris was one of the people who felt a call to action when Hurricane Katrina struck New Orleans in 2005. He left his business and his family, and wound up living in a tent city, where as a volunteer with the Red Cross, he tried to help people find housing, fill out paperwork, and do the things they needed to do to get going. "At the time he was standing around for 14 hours a day on cement or in mud, and his sneakers were always wet."

Chris, whom I talked with later, notes "I can pinpoint the very first symptom. I remember the ball of my right foot cramping and my right arm not swinging when I walked. I thought it was from just being on concrete all the time, living in tents. I was down there almost a month. That would have been late October '05. After I came home, I noticed some other things. My right hand wasn't moving as much as my left when I washed my hair, for example." Cindy did not know at first, because Chris told no one, not his sister, his mother, or even his wife. But, he knew something was wrong, and in January 2006, started looking to doctors for an answer.

Early PD can sometimes be difficult to diagnose, and Cindy tells me that now she knows "at first they thought it was a stroke, or maybe a brain tumor. He was checked for Lou Gehrig's and MS. Then, they called it dystonia." In June 2006, Chris met Dr. Marie Saint-Hilaire, a movement disorders neurologist at Boston University. "She made the diagnosis in 20 minutes."

Only then did he let family know there was a problem. Cindy recalls, "He called me and said, 'Are you sitting down? They said I have Parkinson's.' We didn't know anything about it back then, nothing." They found out as much as they could, and being the fighters they are, the Woods family pulled together to help Chris, and ultimately, to find a cure.

At the time, Chris had been riding in fundraisers for diabetes and multiple sclerosis, diseases which touched other family members. They wanted to participate in a cycling fundraiser for PD. "We looked around and couldn't find anything. I contacted both directors from the MS and diabetes rides and they told me 'don't even think about starting a ride, there is no way in this economy.' There was a guy doing a bike ride from Boston to Maine, and we got together and joined him. That first year there were 37 cyclists. It went one way from Boston to Old Orchard Beach, and there was no way to get back. A family member would have to come get you. Still, we did it again the second year, and we had about 85 riders. The next year the ride didn't happen. We ended up starting all over in 2008, and instead of a one-way we made it a loop around Old Orchard Beach, so riders can get back to their own car or whatever they want." That was how the New England Parkinson's Ride began.

The family all wanted to be a part of this. "Here's the problem, the rest of us don't ride," Cindy said. To this, Chris replied, "well, find a bike." At the time, they were based in New Hampshire, but according to Cindy, "we already loved Old Orchard Beach, and it is much more bike friendly." So they

..continued, p16

...New England Parkinson's Ride, continued

began the process. The first year, 2008, their mother Edna, who was 73, went door-to-door asking Mainers if it would be okay to put in a rest stop at their home, and "no one could say 'no,' how could you?"

From the beginning, the ride was linked to the Michael J. Fox Foundation. Thirty-five riders in 2008 raised over $27,000. Cindy notes they have grown by about 30% each year, and in 2015, 800 riders raised $546,000, the biggest single-day fundraising event for the MJFF to date. Since starting the ride, the group reports over $1,750,000 in cumulative donations to MJFF.

Per the New England Parkinson's Ride website, "all routes begin and end at The Ballpark in Old Orchard Beach, Maine and travel through the countryside and shoreline."

Riders can choose to cover distances of 10, 30, 50, or 100 miles and each section starts at a different time, longer distance riders starting at 7:30am, and the shortest distance riders at noon.

Chris and Terry Woods

Cindy stresses, it is hard work to ride that far, but it is a really fun event. "Everybody has a great time. And, last year we had 42 riders with Parkinson's." Chris is still riding. "I will do the 100 mile ride as long as I possibly can."

Randall Curtis, a 59 year-old resident of Belfast, Maine has PD, and has ridden for each of the last five years. Last year his group of friends and family totaled 11 riders. He notes, "they stagger the starts so everybody gets done at roughly the same time for food and music. It's a fun, fun, day." Randall did the 30 mile ride last year and tells me it is also about fighting PD. "I just think that people need to move it or lose it. You have to exercise, and I can tell it helps me."

There is a dinner on the Friday night prior to the race, but space is limited, and early registration is key for that. Based on projections, they think they will hit 1,000 riders this year. There is a lot that goes into preparing for this, and typically about 300 people sign up during the last two weeks before the deadline – not much notice for the organizers.

There is also the issue of taking enough time for riders to raise funds. Riders are committed to raise (or contribute) at least $100 each to the Michael J. Fox Foundation. The registration fee of $45 covers the cost of food, rest stops, a modest meal after the ride, T-shirt, porta-potties, and even a

…continued, p17

…New England Parkinson's Ride, continued

band after the ride. There is more overhead involved in running an organization like this, and that is why they have become a 501(c) (3). Aside from Cindy, who gave up her career to become the full time director, the staff is all-volunteer. Some funding comes from sponsors.

The New England Parkinson's Ride will be Saturday, September 10, 2016, starting at The Ball Park, 7 Ball Park Way, Old Orchard Beach, Maine.

The schedule is:
100-Mile – 7:30am
50-Mile – 9:00am
30-Mile – 10:30am
10-Mile – 12:00pm

To register or learn a little more, go to http://neparkinsonsride.com/

PD Pearls

Excessive stomach acidity delays stomach emptying, thus delaying the absorption of levodopa. Patients with gastritis, reflux, or chronic heartburn are at risk of other medical conditions as well, and should seek medical help.

Iron salts (such as in multivitamin tablets or ferrous sulfate tablets) may reduce the amount of levodopa available to the body. Iron salts can chelate levodopa and carbidopa, and therefore reduce the bioavailability and effectiveness of carbidopa/levodopa (Sinemet). If iron must be taken, it should not be given at the same time as carbidopa/levodopa.

Restless Leg syndrome (RLS) is a little more common among PD patients than the general population and tends to be worse at night. Symptoms typically include a deep, uncomfortable sensation in the legs (not the feet), the urge to move, stretch, or walk, and at least partial relief with movement. Some patients confuse these symptoms with wearing off of PD medications. Adding to the confusion, PD medications can relieve the symptoms, and therefore, wearing off might exacerbate RLS.

The **incidence of PD**: 1% over age 60, 4% over age 80. In 2012, the U.S. Census Report estimated about 43,000,000 people over 65 in the U.S. The Parkinson Disease Foundation estimates up to a million U.S. citizens have PD.

Constipation in PD

by Bill Stamey, M.D.

Unexplained and persistent constipation in adults is associated with an increased risk of PD. Nearly one third of patients will have been diagnosed with a GI disturbance within the year prior to PD diagnosis (1), and many more will have noticed a change in bowel habits without having received a formal diagnosis. There is a preponderance of data to support this.

In 2001, the Honolulu Heart Program (2), a large population-based prospective study, showed a 2.7-fold risk of developing PD for men with less than one bowel movement daily, when compared with men having one or more daily. In 2011, a large prospective study of over 100,000 people also showed that over the next six years, relative risk of developing PD was was nearly five times greater for men who had one bowel movement every three days or less (3), compared with men having a daily BM. A 16-year prospective study of 8,166 people with PD and 46,755 without PD in the United Kingdom reported that, in the two years prior to diagnosis of PD, 32% had constipation (4).

Unfortunately, the prevalence of GI disturbances in PD increases with age and longer duration of disease. Ultimately, constipation is reported by almost 60% of PD patients (5).

The reasons for constipation in PD are complex. As discussed in the Spring 2016 issue of this newsletter, in the article "What's so bad about alpha-synuclein?," disease-specific pathology may be seen in the gut years prior to the development of motor symptoms of PD, and that pathology may be part of the culprit leading to disease (see also the slides from the April talk on alpha-synuclein).

The problem seems to arise from a slowing down of the gut in PD. There may be a delayed gastric emptying, followed by slow transit of stool. This allows the over-production of bacteria which results in gas, and sometimes, colic. Another problem is that stool is supposed to dry out as it moves through the colon. The slower it goes, the drier it gets, and in a self-perpetuating cycle, the drier it gets, the slower it goes. Some patients with PD have such slow transit they develop a hard stool called a fecalith, or "feces rock." In rare cases, all movement of stool may stop, a medical urgency/emergency known as impaction.

So, what can a person with PD do about constipation?

First, educate yourself. Drugs used to treat many conditions, including those meant to help PD, can cause constipation. Some of these drugs include anticholinergics such as trihexyphenidyl (Artane), benztropine (Cogentin), and even carbidopa/levodopa (Sinemet). Other medication-induced causes of constipation include narcotics, antihistamines, tricyclic antidepressants such as amitriptyline, certain antipsychotics, some antihypertensives, lipid-lowering drugs, calcium supplements, iron tablets, and even antacids (APDA). Before stopping any of these medications, check with your prescribing doctor.

Daily exercise is helpful, and you should check with your doctor about any recommendations or limitations you may have.

…continued, p19

…Constipation, continued

There are certain foods and beverages that may lead to, or enhance, constipation, for example, cheeses, red meats, dairy products, processed foods, fast food, fried food, and "junk" food, are all culprits (your mother was right). Caffeine, while stimulating bowel movements in some, is actually a diuretic, which may dry the stool out even more. The same is true of alcohol. If you drink either caffeine or alcohol, compensate with water. People with constipation should stay hydrated, and eat foods which aid in digestion, such as raw vegetables and fruits, dried prunes, bran, and high-fiber foods. To learn more about diet and find a recipe for a high fiber mix that may help, see the APDA handout on constipation (6).

Generally, one should eat at regular times, and try to have a bowel movement about 30 minutes following the morning meal. This naturally occurs due to a phenomenon called the gastrocolic reflex, in which the brain sends messages to the GI tract to defecate when the stomach is distended.

The Guidelines

In 2006, the American Academy of Neurology (AAN) wrote in their guidelines for the treatment of non-motor symptoms of PD that increased water and dietary fiber intake have shown clinical benefit in relieving constipation (7). The AAN found only limited, or "weak" evidence regarding medications at the time, but supported isosmotic macrogol (polyethylene glycol), available as Miralax over the counter, which "may be considered to treat constipation in PD." The benefit of Miralax is that it is not systemically absorbed, and instead, pulls water like a sponge into the gut, where it can help hydrate stools. Soluble fiber has the same effect and is not absorbed into the bloodstream. However, using Miralax or fiber alone is often inadequate.

Calin Stoicov, M.D., a gastroenterologist at Mid Coast Medical Group in Brunswick, points out that we should be careful in how we define constipation. The definition is not solely based on the frequency of bowel movements, though less than three weekly is a place to start the description. Also, there is straining at lumpy, hard stools, a sensation of incomplete evacuation, and a sensation of rectal obstruction or blockage with 25% of bowel movements.

Dr. Stoicov noted that, broadly, the constipation in PD is treated similarly to other forms. "The agents are divided into four groups: bulking agents such as dietary fiber or artificial fiber (Benefiber, Citrucel), osmotic agents (Miralax), stimulants (senna), and pharmacological agents (Amitiza or Linzess)." He noted that constipation is usually not painful. If there is pain associated with constipation, one should think of another culprit, such as irritable bowel syndrome (IBS). That patient should be evaluated by a physician. For patients with PD who have simple constipation, he recommends starting with Miralax, and to use it liberally, as it is not absorbed. Some patients will try lactulose, but should be careful, as it may be more likely to cause bloating. When patients are prescribed Amitiza or Linzess, they may find more success, but should be careful as these medications may be a little too successful, and result in diarrhea.

People should not expect immediate results, either. …continued, I often hear that this or

…continued, p20

…Constipation, p20
that approach did not help after a few days, so it was abandoned. Or, some people will note that they may have a loose stool on some days, and a hard stool once a week. In this case, there is too much dried out stool in the colon, and loose stool is moving around it. It is better in that case to have a regular, daily bowel regimen, to keep everything moving. Per Dr. Stoicov, medications are usually effective within about a week. He notes that stimulants, such as the herbal laxative senna, have a tendency to stain the colon black, which may be seen during colonoscopy. Some data suggest that this condition, known as melanosis coli, might predispose to polyps.

Finally, it is not good to wait until constipation becomes a problem. Be proactive, educate yourself, and promote good bowel health. This really is a situation wherein you get out what you put in.

References (full citations at http://mainepdnews.org/2016/06/12/constipation-in-pd/)
1. J Parkinsons Dis. 2011;1(1):65–74.
2. Neurology. 2001;57:456–62.
3. Am J Epidemiol. 2011;174:546–51.
4. The Lancet Neurology, 2014;14(1):57-64.
5. Clin Auton Res. 2005;15(2):76–82.
6. http://www.apdaparkinson.org/uploads/files/Constipation-8-25-15-er0.pdf
7. https://www.aan.com/Guidelines/Home/GetGuidelineContent/409

What is deep brain stimulation?

by Bill Stamey, M.D.

Your brain is the most complicated machine known, a mass of billions of interconnected neurons, each of which may form an average of about 7,000 synapses (connections with other neurons), forming your personal and highly unique connectome – the map of all of those connections. At each of these synapses, electrochemical signaling fires in patterns that may be individual or in networks, which generate a signal strong enough to detect with electrodes on the scalp. This is what allows us to study brain waves on an electroencephalogram (EEG). Brain electrical activity can also be changed in a way that helps patients with neurologic diseases, and is the basis of deep brain stimulation (DBS).

However, the story of how we got here goes back farther than most would guess. For example, in the first century A.D. the Roman physician Scribonius Largo described using the electric ray fish, torpedo, to shock the heads of headache sufferers (1). Since the common ray can deliver a charge of up to 200 volts to immobilize prey, one can imagine the effect on a person might be less than pleasant. Electric fishes were actually used in European medicine at least into the eighteenth century. There were many other experimental uses of electricity, on living and expired subjects, in centuries past. Thus it was not so unusual that in the early twentieth century electroconvulsive therapy was established in psychiatric medicine.
…continued, p 21

…What is DBS?, continued

Work with electricity also led to a greater understanding of how our brains operate. In the 1930s, Canadian neurosurgeon Wilder Penfield famously mapped functional regions of the brains of awake patients with electrical stimulation during surgery (2). He was not alone in using electricity to study or modulate the brain. These early pioneers led the way to the formation of DBS.

DBS as we know it today begins in the early 1950s, when Spanish neuroscientist Jose Delgado reported implantation of electrodes in humans (3). A decade later he described radio-equipped electrodes which he had placed in various animals, and in 25 people. In 1963, he demonstrated he could stop a bull from charging (or at least veer off-course) with the device. Over the following decades, investigators around the world experimented with brain electrodes. Two groups in 1991 ushered in modern technology when they successfully demonstrated that DBS could treat tremor (4,5).

DBS was FDA-approved in 1997 for tremor associated with essential tremor and Parkinson disease (6). In 2002, the indications were expanded to include other symptoms of Parkinson disease. Previously, patients may have undergone surgical procedures such as thalamotomy or pallidotomy, which affected or diminished motor symptoms by surgically destroying a specific brain region. After DBS was approved, it became the preferred, or much more commonly administered, treatment. There are several reasons for this, such as the facts that DBS is not intended to destroy brain tissue, is adjustable and programmable, and is at least as equally effective as the older surgical procedures. DBS is also reversible. It may be turned off and sometimes will be removed, though this is uncommon. It has been implanted a great deal since 1997. According to the International Neuromodulation website, between 1997 and 2012 there were over 80,000 DBS implants around the world for a variety of indications (7).

Who should have DBS?

In Parkinson disease, the procedure is usually intended to treat disabling motor complications, tremor, and dystonia refractory to medical treatment. For example, patients may want DBS when dyskinesias become intolerable, or when medications fail to work in a predictable way. Some patients complain of sudden, unpredictable off time when medications should be working. DBS may help, as it is a continuous delivery system instead of the pulsatile approach of medication dosing. There are also PD patients whose tremors respond inadequately, or not at all, to medications. Typically, if other symptoms of PD such as stiffness and slowness respond to medication, then DBS may be a good option for tremor control in these patients. DBS does not cure PD, but helps to diminish some of the motor symptoms. Because of this, patients are usually able to cut back on medications, and thus, medication side effects.

And, who should not have DBS?

Medtronic, the manufacturer of the first, and still most widely used DBS platform, offers the following exclusion criteria: no longer responsive to medications; severely disabled, even in the best “on” state; medical conditions that prevent surgery; and onset of dementia. It may be hard to define exactly when a person has reached each of these states, and this is where having a team-based approach to

…continued, p22

…What is DBS?, continued

assessing the patient can be helpful. In Maine, a team composed of a neurosurgeon, movement disorder neurologists, neuropsychologists, and specialized nurses meet to discuss potential cases and outcomes.

What are the risks?

Risks of surgery can be serious and fortunately occur in only a small minority of patients. These can include stroke, paralysis, coma, death, bleeding in the brain, cerebrospinal fluid leak, seizure, infection, allergic reaction to implanted material, confusion, pain at surgery sites, and headache. These risks should not be taken lightly and should be discussed at length with your doctor prior to making a decision to proceed. Since no two patients are the same, there may be other risks unique to that person, and sometimes the risks are too great.

Programming of DBS is meant to stop motor symptoms of PD, but may also cause unwanted side effects, such as tingling, temporary worsening of symptoms, speech problems (for example, slurred speech or trouble producing words), double vision, dizziness, weakness of the face or limbs, temporary worsening of dyskinesia, coordination problems, feelings of shocks or jolts, numbness, and very rarely, behavioral disturbance. Usually, programming changes can eliminate these issues. In the rare circumstance where relief from side effects cannot be found, DBS can be turned off.

Who has had the procedure in Maine?

Lots of people. Before 2013, patients were implanted in other states. Since 2013, dozens of implants have been done in Maine with Dr. Rughani. I asked a few of my patients about their experience as I was writing this article.

Barbara Keezer of Augusta, Maine, told me that she had suffered with PD for over 14 years

when, in 2005, the development of uncontrolled dyskinesias led to deep brain stimulation with Dr. Van Horne in Boston. She states she "could tell a difference right away, dyskinesias stopped. I couldn't walk before the surgery because I never stopped moving. I was in a wheelchair, but after DBS I could walk by myself." Her husband Gordon notes that she had several good years. Still, with a 25 year course, the disease has progressed. In the last 2 -3 years she has needed a wheelchair again, and a lot of help from him. "She has gone downhill gradually with the Parkinson's, you can't change that. But, she still wants to do things, to be independent." Barbara states her husband is always reminding her "don't do that by yourself." He says, "I have to watch her, and make sure she doesn't fall." Yet, Barbara reports that DBS is still helpful and keeps her doing what she can. "If you turn the DBS off, I can't move at all. So, it's doing something good." For her it was, and still is, a definite benefit.

….continued, p23

…What is DBS?, continued

Forty-three year-old Melinda Jewell has young-onset PD, and in 2013 had right brain DBS in Boston. In 2015 the left brain was implanted by Dr. Rughani in Portland. She is happy with the results and tells me the local experience was ”really good, with a great team.” She was a little nervous about going through it a second time in a different place. At the procedure,”Dr. Rughani talked to me and I think he was distracting me a little,” she smiles. “And, the anesthesiologist was funny. He made jokes and I listened to country music. I forgot to be nervous and I am glad I did it.” As to whether or not it has been helpful, she says, “Yes! I still have some issues, but I probably wouldn’t be working now if I hadn’t had DBS.”

Patricia Joyce has had PD since 2005, and underwent DBS surgery in 2014 with Dr. Rughani. She notes, "I didn't think the surgery was bad at all. It was pretty cool actually. They warned me that the worst part was staying still for so long. I did get a little itchy, but other than that, it was fine. I'm happy I did it, and I'd do it again."

Still, as Dr. Rughani notes in our interview, the risk and benefit should be taken very seriously, and carefully considered before proceeding with surgery.

References (full citations at http://mainepdnews.org/2016/06/12/what-is-deep-brain-stimulation/)

1. Soc. Biol. Fil. 2006;329: 298–302.
2. Brain 1937; 60, 389–443.
3. Confin. Neurol. 1952;12:315-19.
4. Lancet 1991; 337:403–406.
5. Acta Neurochir. 1991; S52: 109–111.
6. http://www.fda.gov/newsevents/newsroom/pressannouncements/ucm451152.htm
7. http://www.neuromodulation.com/deep-brain-stimulation

Parting thoughts

I want to extend another thanks to all those who contributed in articles, interviews, or insights. Also, I would like to thank those who are reading. I have heard from many of you personally, and it seems like this is a good resource. And, as I am new to building websites, please forgive any online clumsiness.

In the fall we will look forward to an article on exercise in PD by Dr. Kleinman. I believe Dr. Dodwell will be contributing, and I am hoping to entice Dr. Drasby and various PT/OT/ST authors.

Be well, and stay out of trouble,

-Bill Stamey, M.D.