# AN UNDERWRITER'S JOURNEY WITH YOUNG ONSET PARKINSON'S DISEASE

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**My Story, as Told by Michael Helms**

Over the course of my underwriting career, I have been exposed to a lot of medical impairments and risks. One thing I have learned in this line of business is there is never a dull moment; just when you think you have seen it all, there is always something that can catch you by surprise. At 44 years old, I'm not exactly in my prime; however, I still have some life to live, things to see, and people to catch up with. I've lived some life and have been around the block a time or two–so it takes a lot to surprise me.

As a life insurance underwriter, when I approach a risk or look at certain impairments, sometimes I can't help but wonder what it would be like to have that impairment or condition. How would I feel? How would my life be different? What other symptoms or co-morbid issues would be associated with this impairment? Sometimes all these thoughts and feelings run through my underwriting brain as I try to deliver a fair and balanced risk assessment with the information I have been provided with. Typically, after the case or risk assessment was completed, I'd move on to the next underwriting risk or impairment, slowly forgetting about the previous thoughts and emotions.

This was especially the case when it came to underwriting a lot of older age diseases such as Parkinson's disease (PD). I would tell myself that this typically only applied to older individuals who usually have associated co-morbid conditions, some form of disability, altered mental status or decreased motor function. I would hear stories from friends or family members talking about how their grandfather or Aunt Loraine had PD for many years, and in all instances they were older. I would briefly think to myself: I hope I never get this when I get older. I would feel sad for individuals who had severe PD. Something like this would never happen to me, or if it does—as I joked with my wife–I'll be so old that I won't even remember it. Oddly enough, usually the first thought of someone with PD always led me to think of the famous actor and PD advocate Michael J. Fox, who was originally diagnosed in 1991 at the age of 29 years old.

If Marty McFly lets me borrow his DeLorean time machine, I could warn myself that it is possible to be diagnosed with PD at the ripe young age of 44 and that I needed to prepare myself for my life to change. Yes, that's right, I was diagnosed with young onset Parkinson's disease (YOPD) last year. To say I was shocked is an understatement. As an underwriter, one of my first thoughts was to look up PD in my company's manual to look at the signs, symptoms and prognosis. I burned up the Google search engine doing all kinds of searches on the topic from A-Z. Perhaps that was a little dramatic, but never in my wildest imagination did I think I could be diagnosed with YOPD.

I have never been a person to complain a lot about aches or pains, or even go to the doctor on a regular basis. I never really had any reason to. It wasn't until about a year ago that the slight tremor in my left hand became more noticeable, especially at night. I had small minor tremors of some sort for years, but thought it was nothing more than benign essential tremors or from drinking too much caffeine. It wasn't until my wife (who luckily works in an orthopedic office) noticed my tremor worsening and consulted with the hand and foot specialist in her office. They referred me immediately to a neurologist, who then diagnosed me with YOPD.

## Epidemiology, History and Definitions

Parkinson's disease is the most common neurodegenerative movement disorder. The prevalence of PD increases by age, with 41 per 100,000 persons in those aged 40-49, 173 per 100,000 in those aged 55-64, and 1903 per 100,000 in those older than age 80, with males slightly more affected than females.[1]

The very first case of YOPD was documented in 1875.[2] In 1987, Quinn et al. described 60 patients with PD who had disease onset under the age of 40[3] and found that all 56 cases with the onset after age 21 had no hereditary factors, while those with onset before age 21 had familial parkinsonism. They proposed the terms "young onset Parkinson's disease" for those who had an unknown cause of PD, and "juvenile parkinsonism" for those who had PD at less than 21 years of age. Currently the International Parkinson and Movement Disorder Society Task Force on Early Onset PD (EOPD) recommends the use of EOPD as PD with age of onset after 21 years but before 50 years.[4] As YOPD is the more widely used term, in this article, YOPD will be the term used to describe PD in those with the age of onset between ages 21-50, while late onset PD (LOPD) will be used to describe onset after the age of 50.

The prevalence of YOPD in Europe is 12-20 per 100,000 and up to 45 per 100,000 among those living in Asian countries. It affects approximately 2-10% of the one million people with the diagnosis of PD in the US.[2] In Korea, the incidence of PD under the age of 50 has increased from 1.2 cases per 100,000 in 2010 to 1.7 cases per 100,000 in 2015.[5]

## Pathophysiology and Risk Factors

Parkinsonism is a clinical syndrome presenting with any combination of bradykinesia (slow movement), resting tremor, rigidity and postural instability. The underlying pathophysiology of PD is the progressive degeneration of dopamine-producing neurons, causing decreased production of the neurotransmitter dopamine in the basal ganglia (found in the midbrain). This leads to the overactivation and underactivation of pathways that control movement, with an overall effect of motor cortex suppression, which then manifests as paucity and slowness of movement. Therefore, the primary treatment of PD is to increase dopamine and to stimulate areas of the brain to counteract the abnormal function of these pathways. In addition, as the dopamine depletion progresses, other parts of the brain (the brainstem, thalamus and cerebral cortex) as well as other neurotransmitters (glutamate, GABA and serotonin) are affected, leading to additional neurological signs and symptoms.

Both environmental and hereditary factors are thought to trigger the younger onset of PD, although the definitive etiology of YOPD is not yet known. The higher incidence of YOPD in Asia has been attributed to genetic factors along with environmental exposure. Although causation is not well established, there are studies that have associated rural living, well water drinking, herbicide and pesticide exposure, and proximity to industry as risk factors for the disease. Substantial exposure to environmental toxins leads to rapid degeneration of dopamine neurons and is presumed to be responsible for the development of PD at an early age.[2] These environmental exposures may lead to the development of disease in someone who has a genetic susceptibility. In particular, the dominant genes SNCA, LRRK2, GBA, VPS35 and recessive genes Parkin, PINK1, DJ1 have been implicated.[6] Typically, those with juvenile-onset parkinsonism (less than 21 years old) are more likely to have an underlying familial or genetic cause.

## Diagnosis and Clinical Manifestations

The diagnosis of PD is clinical: classic manifestations of PD consist of resting tremor, rigidity, bradykinesia and gait instability. A positive response to medication trials of PD therapy is also considered supportive of the diagnosis. Ancillary tests, used to rule out alternative diagnoses, include neuroimaging such as magnetic resonance imaging (MRI), DaTscan (an imaging study in which radioactive tracer, ioflupane, is injected into the blood and attaches to the dopamine transporter), positron emission tomography (PET) and transcranial ultrasound.

The symptoms that appear in YOPD resemble the classical symptoms of LOPD; however, YOPD often begins with dystonia, an uncontrollable stiffness or cramping of a muscle group or limb.[2] In addition to dystonia, levodopa-induced dyskinesias are more common in YOPD than in LOPD.[6] Nonmotor symptoms include sleep disturbances, cognitive impairment, and urinary and sexual dysfunction. Cognitive impairment in PD ranges from subtle deficits to frank dementia. In a small study comparing those with YOPD and LOPD, those with YOPD performed better in cognitive assessment testing in comparison

to those with LOPD, with patients with mild motor impairments scoring higher than those with moderate to severe impairments.[7]

Lastly, some studies have shown that depression and anxiety are more prevalent in those with YOPD in comparison to individuals with LOPD. Whether this is causal or a downstream implication of the early onset of the disease is unclear. Societal factors are contributory, as those with YOPD tend to still be working, forming relationships, have young children, or may be considering pregnancy.[6]

### Treatment

Currently there is no known cure for Parkinson's disease. However, certain therapies may help in reducing symptoms. The treatment of YOPD is unique for each individual and can require repeated adjustments of multiple medications. Initially, YOPD patients are most often treated with levodopa or its alternatives. Levodopa (L-dopa or LD) is converted to dopamine and is the main treatment for both LOPD and YOPD. Other treatments for YOPD include MAO-B inhibitors, catechol-O-methyltransferase inhibitors, amantadine and dopamine agonists. MAO-B helps limit the breakdown of dopamine in the brain, while catechol-O-methyltransferase helps extend levodopa's effects on the brain. Amantadine may be added to help with muscle control and stiffness. Dopamine agonists, commonly known under the brand names Requip and Mirapex, help stimulate dopamine receptors and have been shown to be about as effective as levodopa in treating mild-to-moderate PD.[8]

One of the most important treatments for YOPD is daily movement and exercise. Almost all research on YOPD and LOPD indicates that maintaining regular physical activity and exercise improves mobility, flexibility and balance. There may also be a secondary effect at easing non-motor symptoms such as depression or constipation.[9] Some of the most popular exercises include exaggerated movements to help with hand-eye and brain coordination, strength training, aerobic exercise, stretching and non-contact boxing.

Because a person with YOPD is likely still working, occupational therapy is also considered important and often put into the same classification of treatment as exercise. It is important that someone with YOPD learn daily techniques and tips to help them continue to contribute and be productive in their field of work. Occupational therapy combined with daily movement and exercise is highly recommended to prevent progression of the disease.

In terms of surgical intervention, a well-known treatment for YOPD is deep brain stimulation (DBS). Initially approved by the US Food and Drug Administration (FDA) in 1997 to treat Parkinson's tremor, it was subsequently used as treatment in 2002 for more advanced Parkinson's symptoms. In 2016 it was presented as a surgical option for YOPD patients who have been diagnosed for at least 4 years and who continue to be symptomatic despite medication.[9] DBS is most effective for people who experience disabling tremors, wearing-off spells (i.e., when the effectiveness of levodopa wears off prior to the next scheduled dose, resulting in the return of symptoms) and medication-induced dyskinesias. Electrodes are inserted into a targeted area of the brain using MRI and a second procedure is performed to implant an impulse generator battery (IPG), similar to a heart pacemaker and approximately the size of a stopwatch.[9] A less invasive surgical option is called Duopa therapy. It is a form of carbidopa-levodopa delivered directly into the intestine in gel form rather than a pill. This is designed to improve absorption by delivering the drug directly to the small intestine.[9] Other treatment options for YOPD include the non-invasive focused ultrasound which emits sound waves into the brain to create high energy and heat, thus destroying specific areas in the brain that are associated with the tremor.[9] Less common procedures include thalamotomy, subthalamotomy and pallidotomy, in which a lesion is performed surgically on the thalamus, globus pallidus and subthalamus, respectively, to reduce symptoms.

### Prognosis and Mortality/Morbidity Implications

A study using the Swedish National Registry for PD found that a lower age of onset was associated with slower progression of motor symptoms.[10] In general, individuals with YOPD have less comorbidity, slower disease progression, less frequent gait disturbances, as well as delayed falls, freezing and delayed cognitive decline compared to those with LOPD.[11]

Cognitive dysfunction has been found to progress more slowly in those with YOPD.[12] A study of older PD patients (mean age of 72.1) found that survival in those with normal cognition at baseline was 11.6 years, not significantly different from the general population, in comparison to 8.6 years in those with a diagnosis of mild cognitive impairment.[13]

Mortality data reveal varying results in those with YOPD. Early research of 54 patients found that younger age of onset was associated with a more favorable prognosis than an older age of presentation, with the YOPD group showing the most favorable observed-to-expected mortality ratio of 1.82, as compared to 2.17 for those with an onset of symptoms at between the ages of 50 to 59, and 2.20 for those who were 60 and older. However, the difference was not statistically significant.[14] Another study of 129 patients with YOPD revealed a mortality risk that was double the general population and comparable to the overall PD population, with poor response to L-dopa identified as a risk factor for early death. Cognitive impairment was found in only 19% of YOPD patients as compared to 43% in the LOPD patients.[15]

In contrast, a longitudinal study from Norway followed a cohort of 587 patients with both LOPD and YOPD from 1997 to 2020.[16] The overall cohort mortality was more than two times higher than expected in comparison to whole population mortality. It was found that those with normal cognition had improved survival. The YOPD group, accounting for 12% of the whole cohort, lived for a longer period of time in absolute terms than older patients; however, the mortality in the PD cohort with age of onset of 20-39 years (17 patients) was more than five times higher than expected. Cause of death information was available for 51 people in the YOPD group. PD was identified as the underlying cause of death in 39%, while the rest were due to cardiovascular and cerebrovascular disease, neoplasms, pneumonia, external factors and other causes.[16]

Studies have shown that individuals with YOPD exhibit a slower progression of both motor and cognitive impairment than those with LOPD, and that cognitive impairment is strongly associated with mortality. Overall mortality for those with PD across all ages has been found to be approximately double the general population, with a recent study demonstrating that mortality to be far higher than expected in those who have YOPD. Given the lack of large-scale, long-term data on the mortality of YOPD, further research is needed to provide a deeper understanding of disease course and prognosis.

### Underwriting Considerations

When underwriting YOPD it is important to distinguish YOPD from LOPD. Simply, PD can be broken down into age classifications. Typically, the younger the age of onset and the more severe the disease, the higher the mortality risk. One must also consider how progressive the disease is in the applicant. Does the applicant have very slow, moderate or rapid progression? Are there any cognitive symptoms or mental disorders present? If so, are they currently being treated? What is the duration and severity of the applicant's cognitive or mental condition? Paying attention to any signs, symptoms or features of disability also plays a role in the overall consideration and risk assessment of the individual. Treatment (and response to treatment) is also an important factor, as typically the longer a person with YOPD has been on treatment, especially levodopa, the more likely they are to have dystonia and levodopa-induced dyskinesia.

The following table demonstrates the differences between YOPD and LOPD and the various underwriting considerations, highlighting the importance of paying close attention to the overall age, symptoms, and commonalities and differences between the two.

| | YOPD | LOPD |
|---|---|---|
| **Age of onset** | Ages 21 to 50 | Mean age is early- to mid-60s |
| **Dystonia and levodopa-induced dyskinesia** | More common | Less common |
| **Cognitive symptoms** | Less common | More common |
| **Family history** | More likely | Less likely |
| **Progression** | Tends to progress slower | Tends to progress faster |
| **Frequency in population** | Less common | More common |

*Source: Adapted from T. Cooper, 2022.*[17]

## My Story, Continued

Almost a year has passed since being diagnosed with YOPD. After seeing my neurologist and going over the results of my DaTscan, I soon realized that I basically had no dopamine at all and that I was facing a diagnosis of an incurable, rare disease. It took me a while to process and to really let the overall diagnosis sink into my now "no dopamine underwriter brain," but I soon realized that I'm still healthy enough to control this disease. I'm in the very early process of YOPD and I'm able to exercise, work and connect with great people, family and friends for support. Not all is lost. I'm still semi-young, and the longer I stay involved and active, the better it will be for me in the long run.

There are many great resources out there for YOPD as well as LOPD. The Michael J. Fox organization has raised over $1 billion in funds since he has gone public and does tremendous work for Parkinson's awareness and the YOPD community. The Parkinson's Foundation is also a great place for updated resources, and for advancing research, as the main drive for this foundation is to one day find a cure. Just like the American Heart Association, PD has its own association, called the American Parkinson's Disease Association (APDA), which benefits both LOPD and YOPD. Finally, the Young Onset Parkinson's Network, which is operated by grants given to the organization by the Parkinson's Foundation, is a great resource for YOPD individuals and their family members.[18] I am fortunate that there are many great organizations at the national and local levels, even for a lesser known and rare disease.

We hope that this information at least finds its way to someone you may know or can help you become more aware of someone going through a similar issue. Being diagnosed at an early age with a disease thought to only affect older individuals is tough. Through awareness, support at local and national levels, and continued vital research, my hope is that one day this rare disease will be something that the medical community can easily treat and cure in a safe and efficient manner.

## Notes


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