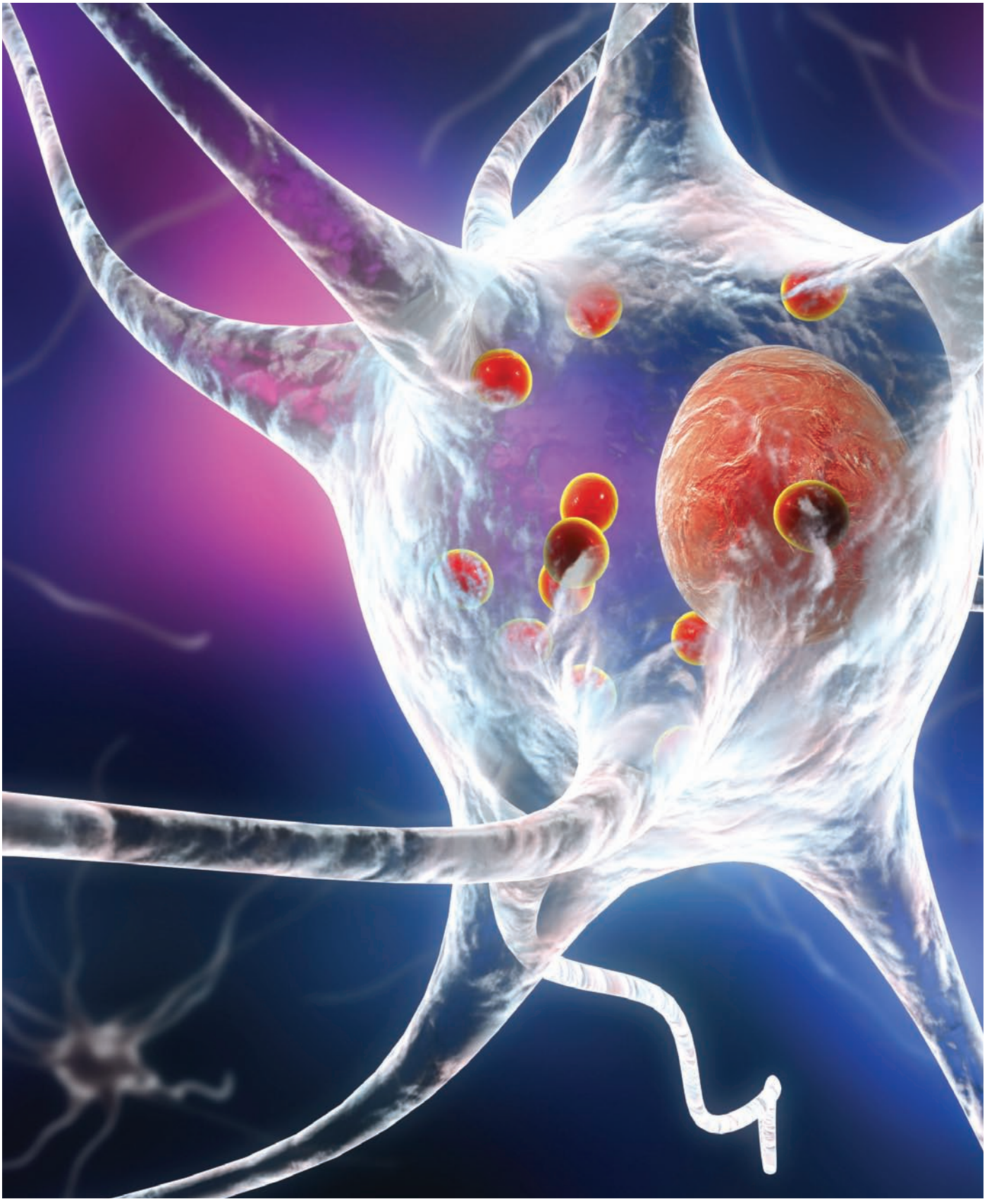

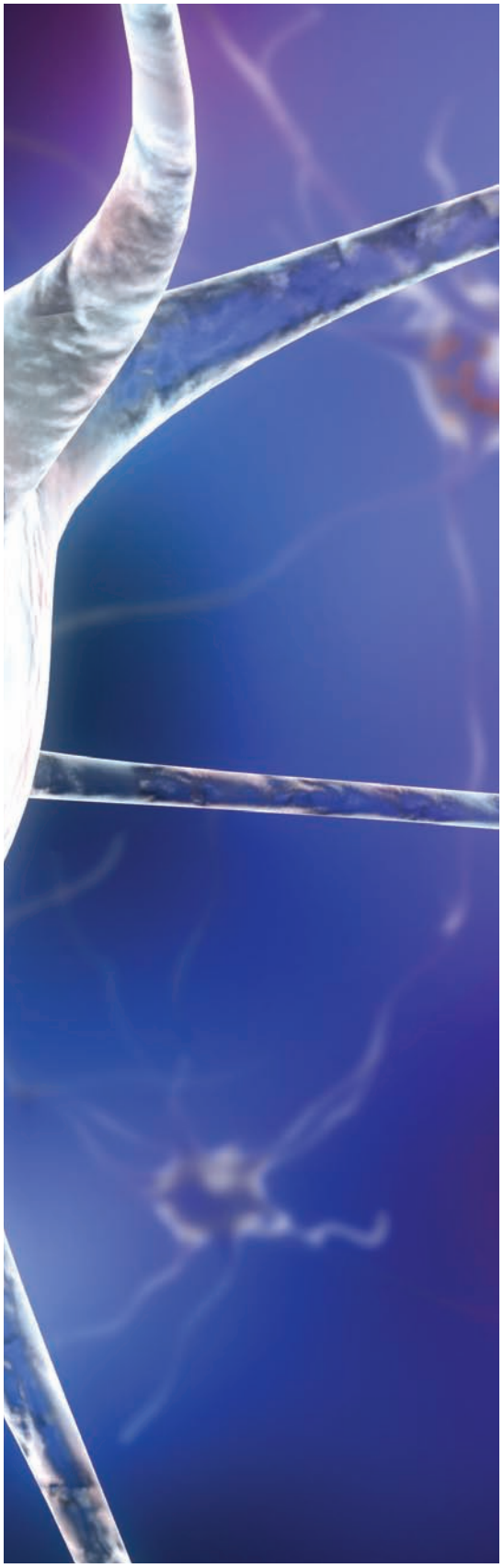

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Understanding parkinsonisms

BY WENDI RANK, MSN, RN, CNRN

Abstract: Parkinsonism describes a group of neurologic disorders associated with signs and symptoms similar to Parkinson disease. This article focuses on four types of parkinsonism: Lewy body dementia, multiple system atrophy, progressive supranuclear palsy, and corticobasal degeneration.

Keywords: corticobasal degeneration, Lewy body dementia, multiple system atrophy, Parkinson disease, parkinsonism, progressive supranuclear palsy

THE TERM *parkinsonism* describes a group of neurologic disorders associated with signs and symptoms similar to Parkinson disease (PD). This article focuses on four types of parkinsonism: Lewy body dementia (LBD), multiple system atrophy (MSA), progressive supranuclear palsy (PSP), and corticobasal degeneration (CBD). Other possible causes of parkinsonism, such as adverse reactions to certain medications or illicit substances, exposure to toxins, and metabolic disorders such as chronic liver failure, are beyond the scope of this article.

Defining parkinsonism

Bradykinesia, resting tremor, muscle rigidity, and other supportive clinical findings are used to identify and diagnose PD (see *Signs and symptoms of PD*). Similar to PD in some ways, parkinsonism is a clinical syndrome that includes many motor signs and symptoms characteristic of PD plus additional signs and symptoms, such as orthostatic hypotension, related to a different underlying cause.[1-3]

While parkinsonism can develop for many reasons, four types share a similar etiology and produce some overlapping symptoms: LBD, MSA, PSP, and CBD. These disorders arise when normal proteins in the brain misfold and accumulate in the brain, causing disease.[1-5]

In CBD and PSP, the misfolded protein is called *tau*, a substance that normally stabilizes cell structures. Because they both result from misfolded tau, CBD and PSP are called *tauopathies*. Interestingly, although not a parkinsonism, Alzheimer disease is also a tauopathy.[2]

LBD and MSA are the result of a different misfolded protein called alpha synuclein. The exact role of alpha synuclein is not well understood, but it may be involved in synaptic and neurotransmitter function. These parkinsonisms are categorized as *synucleinopathies*. PD is also a synucleinopathy.[2]

The reason these proteins misfold, cluster, and cause disease is unknown, although there are many complex theories.[5] Some genetic factors have been identified, but genetic identification is rare. Often, these diseases appear to occur randomly.[5-9]

Difficult to diagnose

The four types of parkinsonism discussed here are relatively rare and difficult to diagnose (see *Incidence of parkinsonism*). The clinical presenta-

Signs and symptoms of PD[19,20]

Cardinal manifestations	Supportive manifestations	
bradykinesia	**Motor**	**Nonmotor**
resting tremor	hypomimia	anosmia
rigidity	freezing	sialorrhea
impaired gait	festination	depression
	decreased arm swing	fatigue
	hypophonia	nocturia
	micrographia	
	dysphagia	

tions are not uniform, and the various signs and symptoms that help identify a particular parkinsonism may take years to develop. Some, such as PSP and CBD, have similar signs and symptoms. Further complicating diagnosis is the lack of specific testing for parkinsonism. Diagnosis is often based upon signs and symptoms and the ruling out of other disorders.[6-10]

One assessment tool that can be helpful is the active transmitter dopamine scan (DaTscan), which uses single-emission photon computed tomography (SPECT) to assess dopamine activity. Dopamine activity on a DaTscan will be impaired in all four of these diseases. While the DaTscan can help differentiate the patient's condition from other diseases, it cannot discriminate between PD and parkinsonism or identify a specific parkinsonism.[2,11]

Despite exhibiting impaired dopamine on DaTscan, these parkinsonisms tend to respond poorly to carbidopa-levodopa, a mainstay of PD treatment. This poor response can sometimes help distinguish a parkinsonism from PD. However, carbidopa-levodopa may be continued after a parkinsonism diagnosis is made if it provides some symptom relief.

No curative treatment exists for parkinsonism, so symptom relief with dopaminergic agents and nonpharmacologic interventions are the prevailing management strategies. See *Select medications used to treat parkinsonisms*.[2]

The following discussion examines each parkinsonism in more detail.

Lewy body dementia (LBD)

Abnormal protein deposits in the brain called Lewy bodies cause cognitive impairment and other symptoms. Because the same protein (alpha synuclein) is abnormally folded in both LBD and PD, many patients with LBD start with a diagnosis of PD or exhibit PD signs such as tremor, rigidity, or bradykinesia. However, patients with LBD exhibit several hallmark signs and symptoms that distinguish it from PD. In addition to the classic cognitive impairment observed in other dementias, disparities in the patient's level of alertness may be reported by caregivers. Periods of attentiveness are contrasted with daytime sleepiness, staring, or poor concentration.[3]

Another feature of LBD is visual hallucinations. Often comprised of children, animals, or dead family members, these hallucinations are usually not frightening, but the patient and caregiver may be distressed that they are having the hallucination. Visual hallucinations associated with LBD are more common in poor lighting.[3,6]

LBD can present similarly to PD dementia (PDD). The distinction is the timing of cognitive decline. In LBD, it usually presents before the motor signs of parkinsonism are seen. In PDD, parkinsonism is present before cognitive changes are seen.[6,13] For a diagnosis of PDD, parkinsonian signs and symptoms must develop at least 1 year before the onset of cognitive impairment.[6]

Rapid eye movement (REM) sleep disturbances are the final hallmark symptom indicative for LBD. Patients may act out their dreams, often violently.[3,6]

Parkinsonisms tend to develop with or after the above symptoms. Because parkinsonisms are so variable in LBD, only one parkinsonism is required for diagnosis. Bradykinesia and impaired gait are seen most frequently and tend to occur bilaterally.[3,6]

Treatment of LBD can be complex. Medications that relieve one symptom can exacerbate others.[13] Research suggests that patients may decline less rapidly if they take a cholinesterase inhibitor, even if their signs and symptoms do not improve, so use of a cholinesterase inhibitor such as donepezil or rivastigmine is common.[3] Nonpharmacologic interventions include environmental modifications, such as altering sleeping quarters for safety when REM dysfunction is present or adjusting lighting to minimize hallucinations.[6]

Multiple system atrophy (MSA)

Progressive autonomic dysfunction is the most significant characteristic of MSA. The autonomic nervous system controls basic bodily functions such as heart and respiratory rates, body temperature, and sensation. Autonomic dysfunction is present early in the course of the disease and is very pronounced.[7]

Cardiovascular dysautonomia is very common in MSA and results in

significant BP changes when the patient moves from a resting to a standing position. Systolic BP can drop up to 30 mm Hg, while the diastolic can drop up to 15 mm Hg. It can take up to 3 minutes for BP to drop.[7] Patients may complain of dizziness or frequent syncopal episodes related to orthostatic hypotension, and falls related to these symptoms are common.[10]

Autonomic dysfunction of the genitourinary tract is also a common symptom of MSA. Patients may experience nocturia or urinary urgency and frequency, sometimes complicated by an enlarged prostate or uterine prolapse.[7]

Hyperreflexia and irregular posture may also be observed in MSA. As the disease progresses, patients often experience dysphagia, sialorrhea, dysphonia, and dysarthria.[7]

Parkinsonian signs associated with MSA include bradykinesia and rigidity. Tremors, if present, are commonly asymmetric and usually manifest as action tremors with spasticity. Despite the similarity of signs, levodopa has little effect on patients with MSA, which can help differentiate it from PD.[7] Diagnosis is based on clinical exam and patient history.

Sympathomimetics such as midodrine or droxidopa can be prescribed to alleviate orthostatic hypotension.[7] If carbidopa-levodopa is used, it should not be abruptly discontinued. Doing so can worsen MSA clinical manifestations, an effect that may be irreversible.[7]

Incidence of parkinsonism

Men and women are affected equally by parkinsonisms; onset is usually after age 50.[1-3,6] One factor complicating diagnosis and treatment is their rarity.

- LBD accounts for 4% of dementia diagnoses in meta-analysis but may represent as much as 20% of dementia diagnoses.[12]
- MSA occurs as 3.4 to 4.9 cases per 100,000 people. This increases to 7.8 per 100,000 people older than 40.[8]
- PSP is seen in 5 to 17 cases per 100,000 people.[22]
- CBD occurs in 5 per 100,000 people. This may be higher because CBD is often undiagnosed.[23]

Progressive supranuclear palsy (PSP)

Also called Steele-Richardson-Olszewski syndrome, PSP involves a degeneration of nerve cells in the brain. It can present in a variety of ways, which not only makes PSP difficult to diagnose, but can also lead to an erroneous diagnosis of PD. Parkinsonism signs in PSP includes bradykinesia, rigidity, and impaired gait.[9,12]

Vertical supranuclear gaze palsy, in which upward and downward eye movement is restricted, is diagnostic for PSP. However, the presentation of a gaze palsy in the course of PSP is variable.[9,12]

Distinguishing PSP from other parkinsonisms is challenging, but MRI may be of some use. The midbrain and superior cerebellar peduncles atrophy in PSP, a change visible on MRI and not seen in other parkinsonisms.[9]

PSP cannot be cured, but certain medications can improve signs and symptoms. Studies indicate that carbidopa-levodopa can have some effect on the gait imbalance, rigidity, and bradykinesia of PSP, but the improvement is much less dramatic than that seen in treatment for PD. Ophthalmologic support, such a botulinum toxin to improve eyelid mobility, bifocals or prisms to aid in vision, and eye drops to provide dry eye relief, can minimize the effects of PSP on vision.[8,12]

Corticobasal degeneration (CBD)

Signs and symptoms of CBD include dementia, dysarthria, myoclonus dystonia, and aphasia. Parkinsonian signs include rigidity and tremor. Some patients develop myoclonus of the arms or face. Dystonia can be observed, as well as apraxia and aphasia.[10,12] Patients with CBD can experience alien limb phenomenon, in which they fail to identify their extremity as their own.[10,14,15]

The rigidity of CBD is usually extreme. Other signs and symptoms that may be observed are impaired executive function, visuospatial dysfunction,

Select medications used to treat parkinsonisms[6,7,9,10,12]

Drug	Uses	Considerations	Applicable diseases
carbidopa/levodopa	parkinsonian symptoms	Avoid abrupt discontinuation.	LBD, MSA, CBD, PSP
cholinesterase inhibitors • donepezil • rivastigmine • galantamine	cognitive impairment	Monitor for bradycardia.	LBD, CBD
onabotulinumtoxinA	dystonia, spasticity, dysphonia	Monitor for dysphagia if used for sialorrhea.	PSP, MSA, CBD

and dyscalculia. For a diagnosis of CBD, patients must fail to respond to levodopa. Secondary criteria for a CBD diagnosis are rigidity, apraxia, and dysarthria; a minimum of two of these should be present for diagnosis.[10]

As with other parkinsonisms, treatment of CBD is directed at alleviating symptoms. For example, myoclonus may respond to the antiepileptic drug levetiracetam or a benzodiazepine.[10] Levodopa can improve symptoms for some CBD patients. Cholinesterase inhibitors can improve cognitive function. Physical, occupational, and speech therapy can improve some symptoms.[10,12]

Patient assessment

To assess a patient with a parkinsonism diagnosis:

- check vital signs and test for orthostatic hypotension.
- look for tremors, abnormal posture, or myoclonus. Note which limbs are affected, and if the symptoms are unilateral or bilateral.
- observe the patient's movements and gait for rigidity and bradykinesia.[16] Have the patient walk heel-to-toe in a straight line to assess for gait disturbances.[17] Test limbs for comparable strength, noting any akinesia or sensory neglect.[16]
- ask the patient orientation questions. As the patient speaks, observe attention span and sialorrhea, and assess for dysphonia, dysphagia, and aphasia.
- ask the patient about sleep, bowel, and urinary habits, and inquire about hallucinations.[16]
- have the patient follow a finger with his or her eyes in an "H" or "star" pattern, observing for any vertical gaze limitations.[16]
- observe for abnormal or seemingly involuntary limb movements that may indicate alien limb syndrome.[15]

Managing parkinsonisms

Caring for a patient with a parkinsonism diagnosis requires a variable approach that can be adapted as signs and symptoms fluctuate during the course of disease. For many with a parkinsonism, the exact diagnosis has not yet been made, so patient education and support is essential.[2] Patients may need diagnostic testing, undergo medication changes, and experience symptom progression on their way to a diagnosis. This can be frightening and frustrating for them and their families. Refer patients and their families to local and national resources for additional information and support.

Fall prevention is especially important for patients hospitalized with a parkinsonism. Nurses should make sure the call bell and other objects are close enough to be easily reached. Bed alarms or one-to-one observation may be required if the patient is disoriented or experiencing hallucinations.[18] It is critical to teach patients with orthostatic hypotension to reposition themselves gradually from a sitting or lying position to an upright position to minimize dizziness and syncope.[19]

Nurses should adhere to the prescribed medication schedule as closely as possible. For some patients, delaying medications can have a direct impact on symptoms. Be aware that some medications become less effective as the disease progresses.[20] Teach patients never to stop parkinsonian medications suddenly.

Bowel and urinary disturbances may play a role in the patient's disease. In addition to adequate fluids, nurses should encourage a diet that includes the recommended daily fiber intake. Depending on the patient's clinical condition, encourage physical activity. Exercise not only can improve bowel function but may also improve physical symptoms and overall health.[20]

Be aware that nonmotor symptoms of parkinsonism, such as autonomic dysfunction, cognitive impairment, and hallucinations, may be more upsetting to the patient than physical symptoms.[16] Nurses should monitor for symptoms of anxiety and depression and assist in screening when indicated.[19]

Take a multidisciplinary approach

Managing parkinsonisms requires a multidisciplinary approach. Physical, occupational, and speech therapy can all provide some benefit. A dietitian may be useful if dysphagia is present. Cognitive therapy can aid patients with executive dysfunction. Patients should be followed by a neurologist and may require other specialists such as urology or ophthalmology. Palliative care may be required for end-of-life decisions.[4,7,9,10]

Bedside screening for dysphagia can alert staff to the need for further testing by a speech pathologist. Sialorrhea, dysphonia, or dysarthria are red flags that warrant further assessment of swallowing. Reinforce the recommendations of the speech pathologist to prevent the complications of impaired swallowing.[21]

Outcomes for patients with parkinsonism are difficult to predict due to the limited research into the disorder and the difficulty in distinguishing similar disorders; autopsy may be the only way to achieve a definitive diagnosis.[4,9,10] One study found that patients with LBD died 3 years earlier than those diagnosed with PD; in the same study those with MSA died 5 years earlier than patients with PD.[4] CBD has a disease course of about 8 years before death.[2,8,9]

Because all of these diseases are ultimately fatal, advanced care planning should be discussed, and palliative care should have a place in the long-term care plan.[12]

Caring for patients with a parkinsonism can be a challenge for nurses. Their diagnosis may not be certain, and their symptoms may be unpredictable and poorly controlled. Educating the patient and family, providing education, and supporting the

multidisciplinary team helps keep the patient safe and may improve the patient's quality of life. ■

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The author and planners have disclosed no potential conflicts of interest, financial or otherwise.

DOI-10.1097/01.NURSE.0000751340.92329.ae