Parkinsons Disease

PALLIATIVE CARE AND HOSPICE REVIEW

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Goals

Review the pathophysiology of PD

Review clinical manifestations

Review treatment

Discuss palliative care and hospice in PD

Pathophysiology of PD

Dopamine depletion from the basal ganglia results in disruptions in connections to the thalamus and motor cortex

Early in the disease process, many compensatory mechanisms mask and reduce these effects but eventually the disease progresses and symptoms manifest

Patients with PD have Lewy bodies (LB) deposits. LB is NOT specific as LBs are found in 10% of ‘normal’ brains and other neurodegenerative diseases.

Epidemiology

PD is a growing source of disability and mortality among neurologic disorders.

In 1990- prevalence was 2.5 million globally. In 2016- prevalence was 6.1 million globally.

Some studies have suggested that men have a higher risk than women for developing PD.

Clinical manifestations

CARDINAL features: tremor, bradykinesia, rigidity

Postural instability is considered the 4th feature which is seen much later in the disease

Tremor- ‘pill rolling’, a rest tremor most noticed when that part of the body is being supported by gravity. The tremor in early PD is most often intermittent, and may not be noticeable to others. In fact, approximately half of patients with PD report a sensation of internal tremulousness in the limbs or body that is unrelated to the presence of observable tremor.

Bradykinesia- slow movements, major cause of disability, can be described as weakness, incoordination, tiredness. Loss of manual dexterity like button shirt, shuffling steps, eventually can be impulsive in movement.

Clinical manifestations continued…

Rigidity- increased resistance to passive movement and can affect any part of the body, can contribute to complaints of stiffness and PAIN.

Postural instability- loss of centrally mediated postural reflexes that cause imbalance, usually later in the disease.

Motor symptoms

- Craniofacial
  - Hypomimia (masked facial expression)
  - Decreased spontaneous eye blink rate
  - Speech impairment, including hypokinetic dysarthria, hypophonia, and palilalia (repetition of a phrase or word with increasing rapidity)
  - Dysphagia
  - Sialorrhea
- Visual
  - Blurred vision
  - Impaired contrast sensitivity
  - Impaired upward gaze and convergence
  - Eyelid-opening apraxia (difficult to do even when ability and desire are present).
- Musculoskeletal
  - Micrographia
  - Dystonia
  - Myoclonus
  - Stooped posture
  - Pisa syndrome (subacute axial dystonia with lateral flexion of the trunk, head, and neck)
  - Kyphosis
  - Scoliosis
  - Difficulty turning in bed
- Gait issues
  - Shuffling, short-stepped gait
  - Freezing
  - Festination

Non-motor symptoms

- Cognitive dysfunction and dementia
- Psychosis and hallucinations
- Mood disorders including depression, anxiety, and apathy/abulia
- Sleep disturbances
- Fatigue
- Autonomic dysfunction
- Olfactory dysfunction
- Gastrointestinal dysfunction
- Pain and sensory disturbances
- Dermatologic findings

Nonmotor symptoms in the psychiatric domain occurred most frequently. Psychiatric symptoms such as psychosis or dementia may cause more disability than the motor features and may be more difficult to treat.

In a single-center survey of 265 patients with PD, pain, mood disorders, and sleep problems were the most troublesome nonmotor symptoms occurring in both early- and late-stage PD

Cognitive dysfunction and dementia

Cognitive dysfunction and dementia are common in PD, and the presence of dementia appears to be an independent predictor of mortality in PD

The dementia of PD is classically considered a subcortical dementia, with psychomotor retardation, memory difficulty, and altered personality

Patients with PD typically report problems with executive function (decision-making or multitasking), memory retrieval, and visuospatial misperception. Deficits in executive functioning are often the earliest indicators of disturbed cognition. This is in contrast to Alzheimer disease (AD), the prototypical cortical dementia, with features of aphasia and apraxia.

Dementia usually occurs late in the course of PD.

The anti-dementia drugs used in Alzheimer's disease can also be used in PD dementia

- In line with AD treatment, memantine can also be used either as monotherapy or in
- Randomized controlled trials showed that the donepezil and rivastigmine moderately improved cognition in PD. After 24 weeks of follow-up, some cognitive and behavioral functional improvements were observed in patients who suffered from mild to moderate PD treated with rivastigmine, which now is approved for PD dementia treatment

Clinical manifestations of Parkisnon's Disease. Uptodate. Kelvin Chou MD. Last updated March 3, 2022

Psychosis and hallucinations

Psychosis occurs in 20 to 40 percent of drug-treated patients with PD, and visual hallucinations are the most common psychotic symptom.

When psychosis occurs, it is attributable to the underlying Lewy body disease, to anti-parkinson drug therapy, or to a combination of the two.

Delusions are a prominent feature of psychosis in PD, and are usually paranoid in nature. Examples of some common delusions include spousal infidelity, people stealing money, intruders living in the house.

Studies have shown that psychosis, *not motor dysfunction*, is the single greatest risk factor for nursing home placement in patients with PD.

Psychosis is also associated with increased caregiver burden and an increased risk of mortality.

Commonly reported risk factors for psychosis in PD include the use of high doses of anti-parkinson drugs, the presence of dementia, advancing age, impaired vision, depression, presence of sleep disorders, high comorbid disease burden, and longer disease duration.

Mood disorders

Depression, anxiety, and apathy are mood disorders that occur most often in patients with PD.

Depression — Depression is the most common psychiatric disturbance seen in PD. Though generally mild to moderate in severity, depressive symptoms in PD are associated with a negative impact on motor disability and decreased quality of life.

Recognizing depressive features in PD is a ***challenge***. The psychomotor slowing and blunted affect commonly seen with depression often resemble the bradykinesia and masked facial expression seen in PD. Furthermore, somatic features of depression, such as decreased appetite, difficulty with concentration, and sleep disturbances, are commonly seen in patients with PD who do not have depression.

Patients with PD who develop depression usually present with sadness, anhedonia, and decreased interest in activities.

Anxiety — Anxiety is the next most frequent psychiatric disturbance in PD and is estimated to occur in approximately 30 to 40 percent of patients. All types of anxiety disorders have been reported in PD, though generalized anxiety disorder and social phobia appear to be the most common. Depression and anxiety are often comorbid conditions in PD, they are also associated with "on-off" fluctuations, with worsened mood and anxiety during "off" periods, and with improvement when in the "on" state.

Apathy — Apathy has been defined as a primary loss of motivation, characterized by diminished speech, motor activity, and emotional expression

While apathy frequently accompanies depression, it can occur in patients with PD who do not have depression [91,92].

Clinical manifestations of Parkisnon's Disease. Uptodate. Kelvin Chou MD. Last updated March 3, 2022

Sleep

Sleep disorders include insomnia, daytime sleepiness with sleep attacks, restless legs syndrome (RLS), and REM sleep behavior disorder (RBD). Can affect between 55 and 80 percent of patients with PD

As noted above, **sleep difficulty** was ranked as one of the most troublesome symptoms

The most common sleep disturbances in PD are sleep fragmentation (frequent awakening throughout the night) and early morning awakening.

Approximately 40 percent of patients with PD take medications for sleep, significantly more than is taken in the general older adult population.

RLS is often reported in PD. RLS is a movement disorder characterized by an urge to move the limbs, associated with an unpleasant sensation that occurs mainly or exclusively

RLS in PD is difficult to assess because of overlapping clinical features between RLS and PD, such as "wearing off" symptoms related to levodopa therapy and akathisia (a state of motor restlessness characterized by the inability to sit or lie still).

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PD Treatment

Levodopa is the most effective drug and first drug of choice in mod or severe (or mild in older adults).

Some are treated with MAOI Selegiline, which can cause confusion in some older adults so do not use in later disease. Less than 70 yo patients with mild symptoms can be amantadine as usually well tolerated, it is known to increase dopamine release, inhibit dopamine reuptake, stimulate dopamine receptors, and possibly exert central anticholinergic effects. Anticholinergics are avoided in older adults with PD

Dopamine agonist are used typically early on in mild disease (ex-pramiprexole)

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Carbidopa-levodopa

Start with smallest doses and titrated to a clinical response.

SHOULD NOT be routinely stopped as it can cause a syndrome similar to neuroleptic malignant syndrome or akinetic crisis.

Can cause nausea, somnolence, dizziness, headache

Advanced PD and parkinsonian disorders

Monitoring progression —As an example, the Hoehn and Yahr (HY) staging scale is a simple, commonly utilized method of capturing the symptom progression in PD. The original HY scale divides the typical pattern of PD progression into five stages:

- Stage 1 – Only unilateral involvement, usually with minimal or no functional disability
- Stage 2 – Bilateral or midline involvement without impairment of balance
- Stage 3 – Bilateral disease: mild to moderate disability with impaired postural reflexes; physically independent
- Stage 4 – Severely disabling disease; still able to walk or stand unassisted
- Stage 5 – Confinement to bed or wheelchair unless aided

Progression through higher HY stages correlates with motor impairment and worsening quality of life.

However, the HY scale is heavily weighted towards postural instability as the main marker of disease severity and does not completely assess other motor impairments of PD, nor does it provide any information concerning the nonmotor symptoms of PD.

The HY scale can be used in atypical parkinsonian disorders (ie, dementia with Lewy bodies [DLB], progressive supranuclear palsy [PSP], multiple system atrophy [MSA], and corticobasal degeneration [CBD]).

Another paradigm divides progression of PD and atypical parkinsonian disorders into four arbitrary stages

- Diagnosis, beginning with the first recognition of symptoms and problems
- Maintenance, on stable medication and without postural instability
- Complex, with increasing disability, motor and nonmotor impairments, and frequent changes in medications
- Palliative, characterized by inability to tolerate adequate dopaminergic therapy, and disabling or life-threatening comorbidities

Advanced PD and parkinsoniam disorders

As symptoms become more bothersome or disabling, and motor fluctuations with dyskinesia emerge, the regimen of medications may need to be modified.

Motor fluctuations are alterations between periods of being "on," during which the patient enjoys a good response to a dose of levodopa, and being "off," during which the patient experiences worsening parkinsonian symptoms.

Dyskinesia consists of various types of abnormal involuntary movements, which are due to the effects of levodopa. A number of strategies may be useful for patients with PD who develop motor fluctuations and dyskinesia. In general, these include adjusting the levodopa regimen and adding adjunct medication such as dopamine agonists, COMT inhibitors, or MAO B inhibitors

For patients with advanced typical levodopa-responsive PD and motor fluctuations whose condition cannot be further improved by noninvasive medical therapy, *device-assisted treatment options include deep brain stimulation can be considered.*

Palliative Care

Although PD and related parkinsonian conditions are chronic diseases with a relatively slow course***, the principles of palliative care are appropriate throughout the disease progression***.

Optimally, patients with PD and associated diseases should receive palliative care according to their particular needs, whether physical, psychosocial, or spiritual. Expert consensus review suggests that palliative care should be integrated early in the disease trajectory.

Moreover, palliative care may be involved in an episodic way throughout the disease progression, with increased involvement at times of symptom or psychosocial need and reduced input at other times, but with continued monitoring to determine the need for community and medical services.

Palliative approach to Parkinson disease and parkinsonian disorders
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Palliative Care

Advanced care planning

A survey sent to 585 patients with PD there were 267 responses, and 94 percent of responders wanted to discuss prognosis and treatment information early and include their family in such discussions. Approximately one-half wanted early discussion of ACP, while over 25 percent wanted early discussions about end-of-life care planning.

Within a separate qualitative study, patients with PD and their partners expressed a desire for a comprehensive tool for future planning and suggested a "roadmap" approach as a guide to decision-making, with opportunities to discuss planning and prognosis when certain life changes occur. Thus, discussion about the future and planning for deterioration and end of life is often acceptable to patients and families.

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Disease burden

PD patients have more physician consultations and more emergency department visits per year than did reference subjects of similar age and sex

They also have ***greater and earlier need for institutional care***

They ***have more hospital admissions compared with patients without PD***

- reasons for admission were pneumonia (13.5%), motor decline (9.4%), urinary tract infection (9.2%) and hip fractures (4.3%), and they occurred 1.5 to 2.6 times more frequently in patients than controls.
- They were almost twice as likely to be hospitalized for more than 3 months and more likely have in-hospital death

Increasing age and presence of dementia were most commonly associated with increased mortality

Symptom burden in advanced PD is high, and it has been reported to be of similar degree as in metastatic cancer

- Jeffrey, Sheun, Ching, Ng. Annals of palliative medicine, Vol 7, No 3 (July 2018). *Palliative Care for Parkinson's Disease.*

Disease progression and prognosis

Currently, there are no specific symptoms or signs in idiopathic PD that allow a practitioner to accurately predict the future course of PD for any given individual

In a recent study, 142 patients with PD followed from 2000 to 2012, with approximately 77 percent having a poor outcome (ie, death, dementia, or postural instability) at 10 years after diagnosis.

A report of 618 patients with PD found that the transition from disease impairment to disability occurred generally between three and seven years after the onset of PD. Impairment was defined by difficulty with daily activities without loss of independent function, while disability was defined by loss of independent function.

Median survival ranged from 6 to 22 years. Increasing age and presence of dementia were associated with an increased risk of mortality.

Pharmacologic issues in advanced PD

As PD progresses there are fewer dopaminergic neurons as well as a progressively lower capacity to store exogenous levodopa and convert it to dopamine for storage and release in the remaining neurons.

Additionally, as the dose requirements of levodopa increase, the patient's functioning is inhibited before his next dose of medication. This usually takes place 2–4 hours after a levodopa dose and may appear as sensory (pain, paraesthesiae), psychiatric (paranoia, anxiety, hallucinations, depression) or autonomic (sweating, belching, constipation, tachycardia, or breathlessness) symptom, or progression of motor symptoms or dystonia. This is called “end of dose wearing off”.

Wearing off has not been fully explored, but may be connected to pharmacokinetic changes

Lokk, J., Delbari, A. Clinical aspects of palliative care in advanced Parkinson’s disease. *BMC Palliat Care* **11,** 20 (2012). https://doi.org/10.1186/1472-684X-11-20

National Collaborating Centre for Chronic Conditions (UK). Parkinson's Disease: National Clinical Guideline for Diagnosis and Management in Primary and Secondary Care. London: Royal College of Physicians (UK); 2006. (NICE Clinical Guidelines, No. 35.) 11, Palliative care in Parkinson's disease. Available from: https://www.ncbi.nlm.nih.gov/books/NBK48497/

The duration of time spent in each of the stages of PD is variable. From an audit of 73 patients undertaken in Cornwall[398] the mean duration of disease was 14.6 years. The time spent in the four stages was: diagnosis 1.5 years; maintenance 6 years

In later stages of PD there may be the need to withdraw dopaminergic drugs due to lack of drug efficacy and increasing sensitivity to unwanted effects such as hallucinations.

As a general guide, medication withdrawal should be managed with help from specialists. Where possible, drug withdrawal should be gradual in order to achieve the best balance between relief of symptoms and minimal side effects. Patients and caregivers at this stage will often agree to reduce medications, exchanging greater levels of physical disability for increased mental clarity. This situation should however be reviewed on an ongoing basis as frequent adjustments may be required to maintain this balance.

Assessment tools

There are symptom assessment tools that could help better delineation of the palliative care needs in PD.

- Palliative care outcome scale (POS) is a 10-item reliable and validated core outcome measure that was designed to cover those domains considered important for palliative care, including pain control, symptom control, patient anxiety, family anxiety, information, sharing feelings, depression, self-worth, practical needs and time wasted
- POS with additional Parkinsonism Plus symptoms (POS-PP) is a 20-item validated extension of the core POS assessing symptoms (POS-S), with additional Parkinsonism Plus symptoms added
- Edmonton Symptom Assessment System (ESAS) is commonly used for symptom screening and longitudinal monitoring in patients seen by palliative care in both inpatient and outpatient settings. It has been psychometrically validated and translated into over 20 languages. It assesses nine common symptoms including pain, tiredness, nausea, depression, anxiety, drowsiness, appetite, wellbeing and shortness of breath.
- To address PD specific symptoms, there is a modified version of ESAS (ESAS-PD) with coverage of clinically relevant symptoms, including constipation, difficulty swallowing, stiffness and confusion. This tool was found to be responsive to treatment, and patients with advanced PD were able to complete ESAS-PD independently or with caregiver assistance.
- Jeffrey, Sheun, Ching, Ng. Annals of palliative medicine, Vol 7, No 3 (July 2018). *Palliative Care for Parkinson's Disease.*

Hospice

THERE MAY BE NO HERALDING SIGNS to determine terminal stage is approaching.

Based on systematic review of the literature and relevant United States Medicare guidelines for hospice eligibility in patients with dementia and other neurologic diseases, hospice referral should be considered in patients meeting any of the following three criteria: Advanced disease as manifested by any of the following:

- Rapid or accelerating motor dysfunction (including gait and balance) or nonmotor disease progression (including severe dementia, dysphagia, bladder dysfunction, and, in multiple system atrophy [MSA], stridor) and disability (restricted to bed or chair-bound status, unintelligible speech, need for pureed diet, and/or major assistance needed for activities of daily living)
- Life-threatening complications in the prior year (recurrent aspiration pneumonia, falls with fractures, pyelonephritis, sepsis, recurrent fever, or stage 3 or 4 pressure ulcers)
- Critical nutrition impairment in the prior year (inability to maintain sufficient fluid/caloric intake and dehydration, or body mass index [BMI] <18, or 10 percent weight loss over six months and refusal of artificial feeding methods)
- Motor symptoms that are poorly responsive to dopaminergic medications or that cannot be treated with dopaminergic medications due to unacceptable side effects and result in significant impairments in the ability to perform self-care

Medicare hospice benefit guidelines for determining prognosis in dementia

To be eligible for hospice, patients must meet both of the following criteria:

I. Functional Assessment Staging (FAST): Patient must be at or beyond stage 7; unable to walk, dress, and bathe without assistance; urinary and fecal incontinence (intermittent or constant); no consistently meaningful verbal communication (stereotypical phrases only or the ability to speak is limited to six or fewer intelligible words)

II. Medical conditions: Patients must have had at least one of the listed medical conditions over the prior year

I. Functional Assessment Staging (FAST)

Stage	Features
1	No objective or subjective difficulties
2	Subjective complaints of forgetting
3	Decreased job functioning evident to coworkers; difficulty traveling to new locations
4	Decreased ability performing complex tasks, eg, planning dinner for guests, handling finances
5	Requires assistance to choose proper clothes for day, season, or occasion
6a	Cannot dress without assistance; occasionally or more frequently
6b	Cannot bathe without assistance; occasionally or more frequently
6c	Cannot toilet without assistance; occasionally or more frequently
6d	Incontinent of urine; occasionally or frequently
6e	Incontinent of bowel; occasionally or frequently
7a	Speech limited to fewer than six intelligible words during an average day
7b	Speech limited to single intelligible word during an average day
7c	Unable to ambulate independently
7d	Cannot sit up independently
7e	Cannot smile
7f	Cannot hold head up independently

II. Medical conditions

1. Aspiration pneumonia
2. Pyelonephritis
3. Septicemia
4. Decubitus ulcer, multiple, stage 3 to 4
5. Recurrent fever after treatment with antibiotics
6. Inability to maintain sufficient fluid and calorie intake with 10% weight loss during the previous six months or serum albumin <2.5 g/dL

Source: Centers for Medicare & Medicaid Services. Local Coverage Determination (LCD) for Hospice Determining Terminal Status (L32015) http://www.cms.gov (Accessed on July 17, 2013).

Medical guidelines for determining appropriateness of hospice referral: Non-disease-specific baseline guidelines plus comorbidities

A patient will be considered to have a life expectancy of 6 months or less if they meet the non-disease-specific **"Decline in clinical status"** guidelines described in Part I. Alternatively, the baseline non-disease-specific guidelines described in Part II plus the applicable disease-specific guidelines (listed in a separate table) will establish the necessary expectancy.

A patient will be considered to have a life expectancy of 6 months or less if there is documented evidence of decline in clinical status that is not considered to be reversible based on the guidelines below.

I. Decline in clinical status guidelines

A. Progression of disease as documented by worsening clinical status, symptoms, signs, and laboratory results

Clinical status:

a. Recurrent or intractable serious infections, such as pneumonia, sepsis, or pyelonephritis

b. Progressive inanition as documented by:

1. Weight loss of at least 10% body weight in the prior 6 months, not due to reversible causes (eg, depression or use of diuretics)
2. Decreasing anthropomorphic measurements (mid-arm circumference, abdominal girth), not due to reversible causes (eg, depression or use of diuretics)
3. Observation of ill-fitting clothes, decrease in skin turgor, increasing skin folds, or other observation of weight loss in a patient without a documented weight
4. Decreasing serum albumin or cholesterol
5. Dysphagia leading to recurrent aspiration and/or inadequate oral intake documented by decreasing food portion consumption

Symptoms:

a. Dyspnea with increasing respiratory rate

b. Cough, intractable

c. Nausea/vomiting poorly responsive to treatment

d. Diarrhea, intractable

e. Pain requiring increasing doses of major analgesics more than briefly

Signs:

a. Decline in systolic blood pressure to below 90 or progressive postural hypotension

b. Ascites

c. Venous, arterial, or lymphatic obstruction due to local progression or metastatic disease

d. Edema

e. Pleural/pericardial effusion

f. Weakness

g. Change in level of consciousness

Laboratory (when available; lab testing is not required to establish hospice eligibility):

a. Increasing pCO_2 or decreasing pO_2 or decreasing SaO_2

b. Increasing calcium, creatinine, or liver function studies

c. Increasing tumor markers (eg, CEA, PSA)

d. Progressively decreasing or increasing serum sodium or increasing serum potassium

B. Decline in KPS or PPS due to progression of disease

C. Progressive decline in FAST for dementia (from 7A on the FAST)

D. Progression to dependence on assistance with additional activities of daily living (refer to Part II, below)

E. Progressive stage 3 to 4 pressure ulcers in spite of optimal care

F. History of increasing emergency department visits, hospitalizations, or clinician visits related to the hospice primary diagnosis prior to election of the hospice benefit

II. Non-disease-specific baseline guidelines (both A and B should be met)

A. Physiologic impairment of functional status as demonstrated by KPS or PPS <70%. Note that 2 of the disease-specific guidelines (HIV and stroke/coma) establish a lower qualifying KPS or PPS.

B. Dependence on assistance for 2 or more activities of daily living:

1. Feeding
2. Ambulation
3. Continence
4. Transfer
5. Bathing
6. Dressing

Comorbidities

Although not the primary hospice diagnosis, the presence of comorbid disease that is likely to contribute to a life expectancy of 6 months or less should be considered for hospice eligibility. Comorbid diseases may include:

1. Chronic obstructive pulmonary disease
2. Congestive heart failure
3. Ischemic heart disease
4. Diabetes mellitus
5. Neurologic disease (CVA, ALS, MS, Parkinson)
6. Renal failure
7. Liver disease
8. Neoplasia
9. Acquired immunodeficiency syndrome (AIDS)/HIV
10. Dementia
11. Refractory severe autoimmune disease (eg, lupus or rheumatoid arthritis)

A patient will be considered to have a life expectancy of 6 months and be eligible for hospice services if they meet criteria for BOTH the above non-disease-specific baseline guidelines AND disease-specific guidelines (shown on a separate table).

These baseline guidelines do not independently qualify a patient for hospice coverage. Refer to separate table for disease-specific guidelines to be used with these guidelines.

pCO_2: partial pressure of carbon dioxide; pO_2: partial pressure of oxygen; SaO_2: arterial oxygen saturation; CEA: carcinoembryonic antigen; PSA: prostate-specific antigen; KPS: Karnofsky Performance Status; PPS: Palliative Performance Score; FAST: Functional Assessment Staging; HIV: human immunodeficiency virus; CVA: cerebrovascular accident or stroke; ALS: amyotrophic lateral sclerosis; MS: multiple sclerosis.

Reproduced from: Centers for Medicare & Medicaid Services. Local Coverage Determination (LCD) for Hospice Determining Terminal Status (L33393) (Accessed on January 18, 2016).

erences 36,37 of 'Palliative approach to Parkinson disease and parkinsonian disorders'

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ognostic tool: a risk score to estimate survival in nursing home residents with advanced dementia.

JM, Davis RB, Shaffer ML

010 Nov;40(5):639-51.

xpectancy is challenging in advanced dementia.

k score to estimate survival in nursing home (NH) residents with advanced dementia.

ospective cohort study performed in the setting of all licensed U.S. NHs. Residents with advanced dementia living in U.S. NHs in 2002 were identified using Minimum Data Set (MDS) assessments. Mortality data from Medicare files were used to determine 12-month survival. re selected from the MDS. Cox proportional hazards regression was used to model survival. The accuracy of the final model was assessed using the area under the receiver operating characteristic curve (AUROC). To develop a risk score, points were assigned to variables in arameter estimates. Residents meeting hospice eligibility guidelines for dementia, based on MDS data, were identified. The AUROC assessed the accuracy of hospice guidelines to predict six-month survival.

40.6% of residents with advanced dementia (n=22,405) died. Twelve variables best predicted survival: length of stay, age, male, dyspnea, pressure ulcers, total functional dependence, bedfast, insufficient intake, bowel incontinence, body mass index, weight loss, and he AUROC for the final model was 0.68. The risk score ranged from 1 to 32.5 points (higher scores indicate worse survival). Only 15.9% of residents met hospice eligibility guidelines for which the AUROC predicting six-month survival was 0.53.

risk score derived from MDS data predicted six-month survival in advanced dementia with moderate accuracy. The predictive ability of hospice guidelines, simulated with MDS data, was poor.

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Clinical Intuition

“Would you be surprised if the patient were to die in the next year, months, weeks, days?”

Jeffrey Sheun Ching Ng. Annals of palliative medicine. Vol 7, No 3 (July 2018). *Palliative Care for Parkinson's Disease.*

Table 1 Guidance to identify advanced PD patients with palliative care needs

Gold Standard Framework (GSF) Proactive Identification Guidance (22)
Indicators of a limited life expectancy in advanced PD
(I) Drug treatment less effective or increasingly complex regime of drug treatments
(II) Reduced independence, needs ADL help
(III) The condition is less well controlled with increasing "off" periods
(IV) Dyskinesias, mobility problems and falls
(V) Psychiatric signs (depression, anxiety, hallucinations, psychosis)
(VI) Similar pattern to frailty listed in GSF
Hospice guidelines for neurologic disease in US (27)
(I) Critically impaired breathing including dyspnea at rest, vital capacity ,30%, O_2 need at rest, and refusal of artificial ventilation, or (II) Rapid disease progression (to bed-bound status, unintelligible speech, need for pureed diet, and/or major assistance needed for ADLs) with either
(i) Critical nutrition impairment in the prior year (inability to maintain sufficient fluid/caloric intake, continuing weight loss, dehydration, and refusal of artificial feeding methods) or
(ii) Life-threatening complications in the prior year (recurrent aspiration pneumonia, pyelonephritis, sepsis, recurrent fever, or stage 3 or 4 pressure ulcers)

PD, Parkinson's disease; ADL, activities of daily living.

Hospice management

Pain

- Pain management remains mainstay of hospice care
- Rigidity- increased resistance to passive movement and can affect any part of the body, can contribute to complaints of stiffness and PAIN.
- NSAIDS, acetaminophen, morphine/narcotics
- Some suggest- If levodopa and exercise aren't adequately addressing pain, consider injectable botulinum toxin A (BOTOX) treatment. It can alleviate dystonia by targeting and weakening overactive musclesParkinsonism
- **Continuation** of antiparkisonian therapy is important to **reduce rigidity**.
- For patients with GI tubes, levodopa tabs can be crushed. Also available as oral disintegrating carbidopa-levodopa (Parcopa)
- Reduction of these therapies is a common strategy in patients who are no longer receiving therapeutic benefit, tapering should be over several weeks or more as sudden withdrawal can happen with hyperthermia and rigidity. ALSO can CAUSE anxiety, panic, sweating, nausea, pain, dizzy, drug cravings

Hospice management

Dyspnea and secretions

- Any patient in the final stage of life may have problems clearing secretions
- “Death rattle" -gurgling and crackling sounds that result from increased secretions can be distressing to the family
- Increased airway secretions may interfere with a patient's ability to sleep, worsen dyspnea, precipitate uncomfortable coughing spells, and predispose to infections.
- In addition to reassuring the patient's family, proper positioning and encouraging the family to cleanse the mouth with sponge sticks can be helpful.
- Discontinuing nonessential intravenous fluids or enteral feedings combined with positioning the patient on his or her side helps move the secretions out of the airway.

continued

Constipation

Nausea

- Ondansetron is commonly used
- Avoid haloperidol and metoclopramide as these can worsen parkinsonian symptoms

Hydration and nutrition

- Dysphagia and aspiration are common at end of PD
- Avoid dry mouth with oral care and sips
- The use of infusions of fluid, either intravenously or subcutaneously, is debated, but there is little evidence that the continuation of clinically assisted hydration would prolong life or extend the dying process, or whether stopping hydration will hasten death
- The presence of an infusion can be unpleasant for a patient and may lead to problems such as fluid overload, increased chest secretions, and bladder/vomiting issues

Continued

Psychosis

- The atypical antipsychotics, clozapine and quetiapine are possibly the most effective treatments, while olanzapine probably does not improve psychosis. Risperidone is not a true atypical antipsychotic and has been found to exacerbate PD motor symptoms. In general, all these drugs should be prescribed in low doses with slow upwards titration in order not to cause side effects.

Summary

PD is a progressive neurodegenerative disease with varying levels of progression for each patient.

The cardinal features of PD are tremor, bradykinesia, rigidity, and postural instability.

Cognitive dysfunction and dementia are common in PD, and the presence of dementia appears to be an independent predictor of mortality in PD

Studies have shown that psychosis, *not motor dysfunction*, is the single greatest risk factor for nursing home placement in patients with PD.

The Hoehn and Yahr (HY) staging scale is a simple, commonly utilized method of capturing the symptom progression in PD.

Early palliative care, education, and advanced care planning are advised.

Hospice care is similar to other disease processes but……**Continuation** of anti-parkisonian therapy as long as able is important to **reduce rigidity**.

Katz M, Goto Y, Kluger BM, et al. Top Ten Tips Palliative Care Clinicians Should Know About Parkinson's Disease and Related Disorders. J Palliat Med 2018; 21:1507.

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