

PARKINSONISM: PARKINSON'S DISEASE AND PARKINSON'S PLUS SYNDROMES, AND THEIR DIFFERENTIAL COGNITIVE MANIFESTATIONS

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Definition of Parkinsonism

- An umbrella term for neurological disorders of movement: Not everyone who has parkinsonism - or parkinsonian features - has Parkinson's disease.
- Movement disorders are chronic (persist over time) and progressive (worsen), i.e. neurodegenerative.
- All movement disorders are characterized by 2-4 motor or movement abnormalities.
- Diagnosis is observational / clinical.
- Best diagnosed by a fellowship-trained neurologist who specializes in movement disorders.

Symptoms of Parkinsonism

1. Slowness (Bradykinesia)

- Gradual loss of spontaneous movement
- Decreased automatic movements
- Difficulty initiating movements
- General slowness
- Decreased facial expression

Symptoms of Parkinsonism (cont'd)

2. Tremor: Involuntary, uncontrollable muscle contraction.

3. Rigidity or Stiffness: Tightness of arms, legs, or trunk, preventing muscles from relaxing and stretching.

Can be experienced as:

- Stiffness and inflexibility of muscles
- Pain and muscle cramps
- Difficulty turning

Symptoms of Parkinsonism (cont'd)

- **Postural Instability and Gait Disturbances,**
manifested in FALLS and difficulty or inability to:
 - Rise from a chair or out of bed
 - Turn and pivot
 - Stand upright

Given these motor symptoms, plus the concurrent cognitive and behavioral impairments that we will examine, it is clear that people with any form of parkinsonism require care, from carepartners and/or specially trained home care assistants.

Parkinson's Disease (PD)

- The most common form of Parkinsonism.
- Average age of onset: 59
- 700,000 cases in the U.S.

Parkinson's Disease: A Clinical Diagnosis

- **Bradykinesia** (slowness, decreased automatic movements and initiating movement) **and** one or more of the 3 other motor symptoms:
- **Resting Tremor:** Occurs when muscles are still; improves upon moving affected limb. Present in 70-80% of PD cases.
- **Stiffness:** Inflexible muscles that do not easily relax or stretch; tightness of arms, legs, trunk; reduced arm swing; reduced facial expression; difficulty turning.
- **Postural Instability:** Stooped posture; festinating and freezing of gait.
- **Asymmetrical:** Almost always starts on one side.

Parkinson's Disease: A Clinical Diagnosis (cont'd)

- Response to levodopa (for more than 5 years). (Note: PD = loss of dopamine).
- Exclusion of other diagnoses or causes of Parkinsonism.
- Disease progression and duration (over 10 years).
- Postural instability is not the first, nor the core, feature.
 - If postural instability presents early, it suggests a possible alternate diagnosis, one of the atypical Parkinsonisms which we will discuss.

Early Cognitive Changes in PD

- Slowed mental processing
- Impaired verbal recall
- Difficulty multi-tasking or organizing complex activities
- Difficulty learning new motor skills
- Note: Just as movements are slower, so too are thought processes.

PD with Mild Cognitive Impairment (PD-MCI)

- Impaired visuospatial functions and visual memory
- Impaired executive functioning
- Difficulty ordering or sequencing new information

Reference for this and next 3 slides: Sezgin et.al. Parkinson's Disease Dementia and Lewy Body Disease, Semin Neurol 2019;39:274-282.

PD with Mild Cognitive Impairment (cont'd)

- Affects 10-20% of people with PD at the time of diagnosis.
- Cognitively normal people with PD may develop MCI during the disease.
- These people are at higher risk to progress to PD dementia.

PD Dementia (PD-D)

- Dementia within PD diagnosis: Motor symptoms precede dementia.
- Cognitive Impairment: Attention, memory, word-finding, visuospatial functions, executive function, disorientation.
 - Memory impairments include difficulty recalling recent events and the tasks involved in learning new material. Usually improve with cueing.

PD Dementia (cont'd)

- Behavioral Changes: Apathy, hallucinations (mostly visual), delusions (mostly paranoid), increased depression and anxiety.
 - Apathy: Decreased spontaneity, loss of motivation and interest. Can occur any time in PD, but may be worse if it occurs along with PD dementia.
 - Depression and anxiety can occur any time in PD, but may be worse if they occur with PD dementia.

PD Dementia (cont'd)

- Increase in autonomic symptoms: e.g. constipation, urinary incontinence, or orthostatic hypotension.
- Sleep Disturbances: Excessive daytime sleepiness, insomnia, disruptions in sleep, or REM behavior disorder (RBD), i.e. acting out of dreams.

Risk Factors for PD-MCI and PD-D

- Older age at onset of disease or at diagnosis.
- Severe motor disability.
- Poor response to medication.
- Early development of confusion and hallucinations.
- Speech impairment.
- Symmetrical (as opposed to one-sided) symptoms from the outset.

Strategies for Dealing with Cognitive Difficulties of PD

- Allocate more time when learning new motor skills.
- Visualization, e.g. to relax, or to prepare for a new situation.
- Write and follow step-by-step directions for complex tasks that require attention.
- Arrange the steps of a task so as to minimize switching from one task to another; focus on one step, of one task, at a time.

Strategies (cont'd)

- Maintain predictable routines as much as possible.
- Use reminders.
- Prepare for social or professional events by reviewing pertinent information.
- Take time to explore unfamiliar surroundings.
- Minimize stress.
- Address depression.

Strategies (cont'd)

- Adequate sleep
- Good nutrition
- Exercise
- Mental exercise
- Review medications with physician.
- Family members, as well as home care providers, can implement these strategies.

The Overlap of Dementia Disorders and Parkinsonisms

- Dementia and Parkinsonism are umbrella terms encompassing types of dementias and types of Parkinsonisms respectively.
- Lewy Body Dementia (LBD) is an umbrella term that includes Dementia with Lewy Bodies (DLB) and Parkinson's Disease Dementia (PDD).
- DLB shares features with the parkinsonism called Multiple System Atrophy (MSA): REM sleep behavior disorder, and autonomic nervous system dysfunction.
- The Parkinsonian dementias comprise 5% of dementias.

Atypical Parkinsonism or Parkinson-Plus Syndromes: Parkinsonisms That Are Not PD

1. Progressive Supranuclear Palsy *
2. Multiple System Atrophy *
3. Corticobasal Degeneration *
4. Drug-Induced Parkinsonism
5. Chronic Traumatic Encephalopathy (CTE) — head trauma
6. Vascular Parkinsonism

Features that Suggest Atypical Parkinsonism

- Additional neurologic changes, along with the 4 hallmark features of slowness, tremor, stiffness, postural instability.
- Rapid disease progression.
- Poor response to dopamine medications.
- **Postural instability:** Early gait impairment and falls.
- **Tremor:** Irregular or jerky Bi-lateral symmetry at the outset.
- Early signs of autonomic dysfunction: Dizziness, bladder symptoms, constipation.

1. Progressive Supranuclear Palsy (PSP)

- **Progressive:** The early symptoms get worse, and new symptoms continue to develop over time.
- **Supranuclear:** This term describes the eye problem in PSP: Weakness or paralysis of the muscles that move the eyeballs, resulting in inability to aim the eyes properly.
- **Palsy:** Weakness or paralysis of a part of the body. PSP consists of palsy in eye movement, legs, arms, swallow mechanisms, and others.
- PSP is a "tau" disease: Abnormal accumulation of normal protein tau.
- Average age of onset: 63
- 20,000 cases in U.S.

Early Symptoms of PSP

- **The most common first symptoms: Postural instability, slowness, symmetric rigidity; loss of balance while walking; freezing of gait; FALLS - mainly backwards.**
- Other early symptoms may include:
 - Unexplained, vague visual problems, e.g. rapid involuntary right-left movements of the eye; slowed blinking.
 - Personality changes such as increased irritability or withdrawal.
 - Apraxia or slurred speech (a motor function: difficulty getting words out) and/or aphasia (a cognitive function: difficulty comprehending or finding words).
 - **Tremor:** Mild shaking of the hands.

As PSP Progresses

- Increased imbalance, stiffness, and difficulty walking.
- Inability to aim the eyes and to move them up and down (the down movement impairment is unique to PSP): appears 3-5 years into the disease.
- Abnormal eyelid movement: too much or too little.
- Difficulty closing the eyes; very limited blinking.
 - Eye problems interfere with reading, driving, maintaining eye contact during conversations, descending stairs, eating.

As PSP Progresses (cont'd)

- Swallow difficulty due to throat muscle weakness or incoordination.
- A combination of 3 or 4 different speech problems:
 - Slurred
 - Irregular
 - Explosive
 - Rubber-band quality - Spastic
 - Softening

PSP Dementia

- Arises early in disease course, compared to PD.
- In PSP, cognitive performance is worse, and decline more rapid, than in the other 2 Parkinson Plus syndromes we will examine.
- Deterioration in all cognitive functions: Attention, memory, visuospatial functions, executive function.
- Difficulty resisting impulses; disinhibited behavior.
- Apathy: Little or no interest in surroundings and activity.
- Dementia worsens during unrelated stress, e.g. tight clothing, bedsore pain, infection, lack of sleep.

2. Multiple System Atrophy (MSA)

- **Multiple System:** A disease of the central nervous system (brain and spinal cord) and the autonomic nervous system (a subset of the CNS: "automatic").
- **Atrophy:** "Wasting" or decrease in size, strength, and function of an organ or system.
- Average age of onset: 53
- 13,000 cases in US
- **Motor and balance impairments:**
 - **Postural instability** and severe balance difficulties
 - Inability to coordinate movements and maintain balance: a "drunken-appearing walk"
 - Weak or abnormal reflexes
 - **Stiffness/Rigidity:** Dystonia - holding part or all of a limb in a fixed position

MSA (cont'd)

- **Action Tremor:** When reaching for an object
- **Myoclonus:** Sudden jerking or twitching muscle movements
- **Speaking difficulties:** Slurred, slow, low volume
- **Chewing and swallowing difficulties**
- **Laryngeal Stridor:** Airway (larynx) obstruction
- **Sleep apnea and/or REM sleep disorder**
- ***Early autonomic dysfunction**

MSA Symptoms Arising in Autonomic Nervous System

- Sudden drop in blood pressure - Lightheadedness and fainting upon standing
- Noisy or irregular breathing
- Severe constipation; urinary urgency or incontinence
- Sleep disruption and insomnia
- Reduction in perspiration, tears, and saliva
- Cold hands and feet; heat intolerance
- Irregular heartbeat

MSA Cognitive Issues

- Cognitive disturbances occur across a wide spectrum: From mild, single-domain deficits to multiple domains and to dementia.
- Cognitive decline 5-6 years after disease onset
- Absence of hallucinations
- Progression towards dementia is gradual.
- Motor impairment is a predictor for the severity of cognitive impairment, same as with PD.

MSA Cognitive Issues (cont'd)

- Executive function is "often impaired." Difficulty organizing information by categories, understanding abstractions and instructions, and creating and following a plan.
- Attention, memory, spontaneous recall, and visuospatial functions are "sometimes impaired."
- Language functions like spontaneous speech, word-finding, and syntax (well-formed sentences) are preserved, even though speech itself is slurred.

Stankovic et.al. Cognitive Impairment in Multiple System Atrophy, Mov Disord. 2014 Jun; 29(7): 857-867.

- Disinhibited behavior: Difficulty controlling impulses and emotions.

Corticobasal Degeneration (cont'd)

- Begins between 50 and 70 years old; life expectancy 5-10 years.
- 2,000 cases in U.S.
- Bilateral, yet one side starts worse and remains so.
- Myoclonus: Sudden jerking or twisting muscle movements that are involuntary and irregular (not rhythmic).
- **Stiffness:** Dystonia - holding part or all of a limb in a fixed posture.

3. Corticobasal Degeneration (CBD)

- **Postural instability:** Gait difficulty, clumsiness, freezing for seconds at a time; yet mild problem with balance compared to PD and PSP.
- **Apraxia:** 1) Difficulty performing complex manual movements when asked, despite having the desire or ability. 2) Difficulty using hands or feet to perform familiar movements, e.g. cutting food, buttoning, or typing.

Corticobasal Degeneration (cont'd)

- **Tremor:** Mild and less frequent compared to PD and PSP.
- **Dysarthria:** Difficulty controlling muscles of face and mouth.
- **Sensory problems, involving the inability to:**
 - Interpret spatial complexity of touch
 - Recognize objects by feel
 - Know the position of a finger or limb in space
 - **Asymmetrical Alien Limb Syndrome:** Patient has the sense that their limb does not belong to them and is controlled by some external force.

CBD Cognitive Issues

- Cognitive loss is present in half of patients at outset, and in 70% of cases eventually.
- Global loss of intellectual abilities - dementia - in late-stage disease course.

CBD Cognitive Issues (cont'd)

- Slowed thought.
- Reduced attention span.
- Memory problems: misplacing objects, repeating questions.
- Aphasia: Difficulty recalling, understanding.
- Language problems: Difficulty finding words, naming objects and people, getting the words out, adhering to correct grammar, reading. Ultimately, person loses ability to speak.
- Loss of visuospatial skills.

CBD Cognitive Issues (cont'd)

- Difficulty multitasking.
- Executive functioning: Difficulty planning and organizing; difficulty synthesizing information into a new idea.
- Difficulty resisting impulses: Inappropriate impulsive behavior, disinhibited social behavior, or obsessive-compulsive behaviors.

Psycho-social Impact of these Diseases

- Caregiving itself + fatigue, worry, decrease in self-care.
- Unpredictability and fluctuation of symptoms: Require flexibility.
- Irritability and mood shifts: These are not personal, but reflective of the disease. Need for tolerance, patience, respite.
- Apathy.
- Changes in family roles and responsibilities.
- Depression and Anxiety

Psycho-social Impact (cont'd)

- Age of onset: Workplace complications, lifestyle interference, parenting
- Social isolation due to:
 - Embarrassment and/or social stigma
 - Speech disturbances — volume, pace, clarity
 - Slowed thinking ability — impaired conversational ability
 - Eating difficulties

Not curable but...

It is our job as professionals to address concerns of safety and well-being, and to help affected individuals and families to maximize their Quality of Life and to plan for their future care needs.

"We cannot cure, but we must care and give care."



Providing Support for People with Dementia

Clinical Education

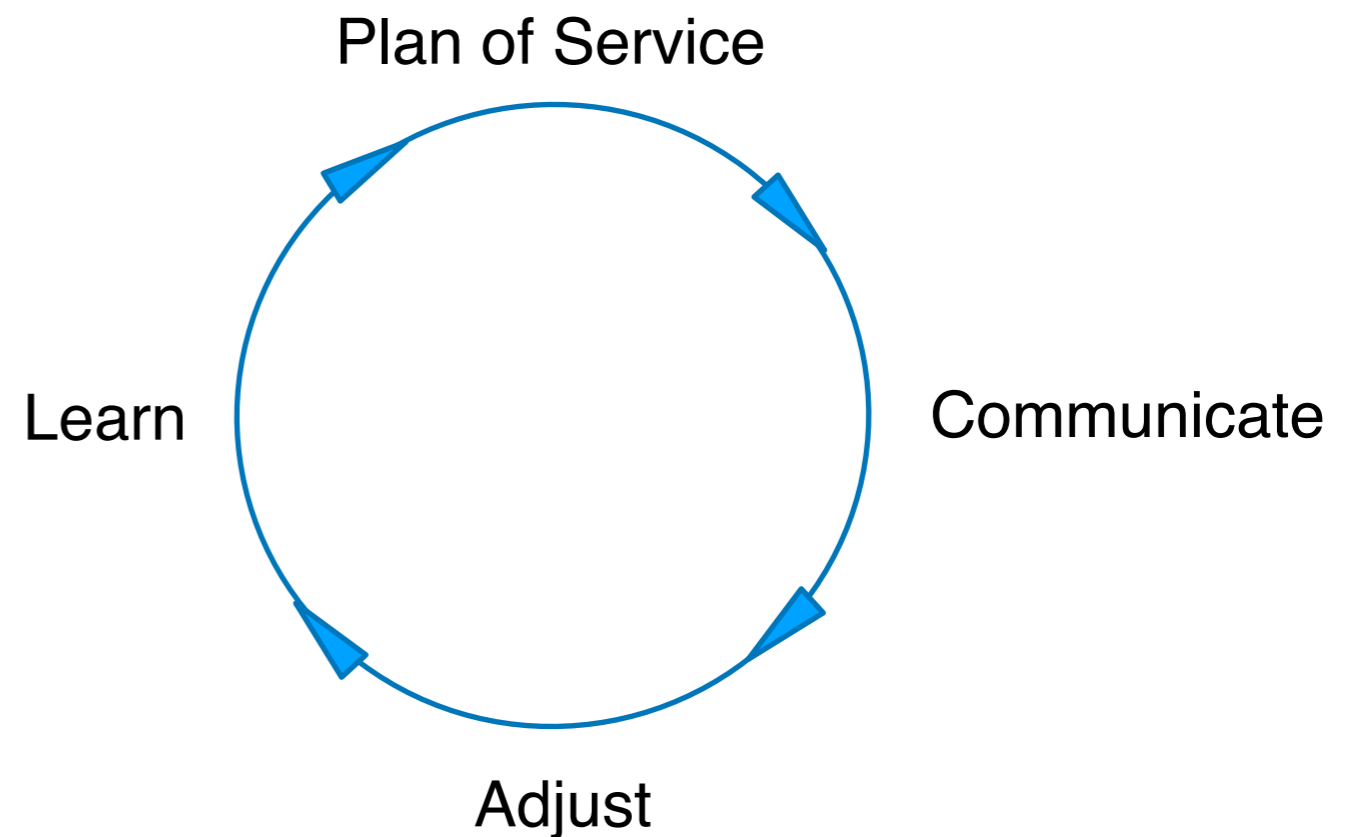
- Learning the causes and symptoms
- Patience and Understanding

Training

- Develop support techniques
- Skills Toolkit

Client-Specific Training

- Knowing the client
- Personalized support



Bridging the Gap



Broad Street



Clinical Direction

Feedback

Nurse Oversight
&
Direct Care Support



***Healthcare
Professionals***



Home

In Summary

- Parkinsonism is an umbrella term.
- Parkinson's disease is the most common Parkinsonism.
- We have looked at 3 other Parkinsonisms, the so-called Parkinson-Plus or Atypical Parkinsonisms: PSP, CBD, MSA.
- By and large, the cognitive issues of Parkinsonisms are in executive functioning, with more impulse control behavior disorders in the Atypical Parkinsonisms than in PD.
- By understanding the unique physical and cognitive issues related to Parkinsonism, and then developing techniques to help support the person and manage their symptoms, we can help individuals to receive proper care and to enjoy a better quality of life.

To learn more about Broad Street Comprehensive Parkinson's Program,
click below

<https://broadstreethomecare.com/customized-home-care/parkinsons/>

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