

The Parkinsonisms: Parkinson's disease and the Parkinson-Plus syndromes

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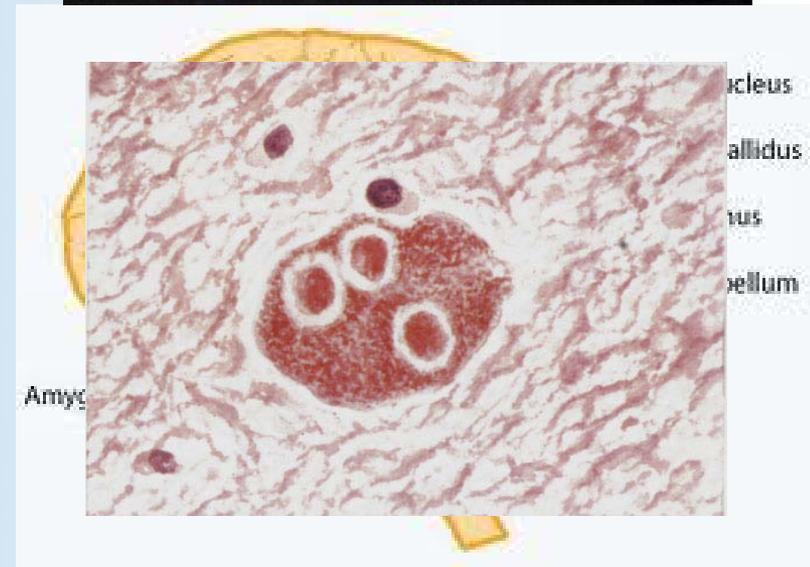
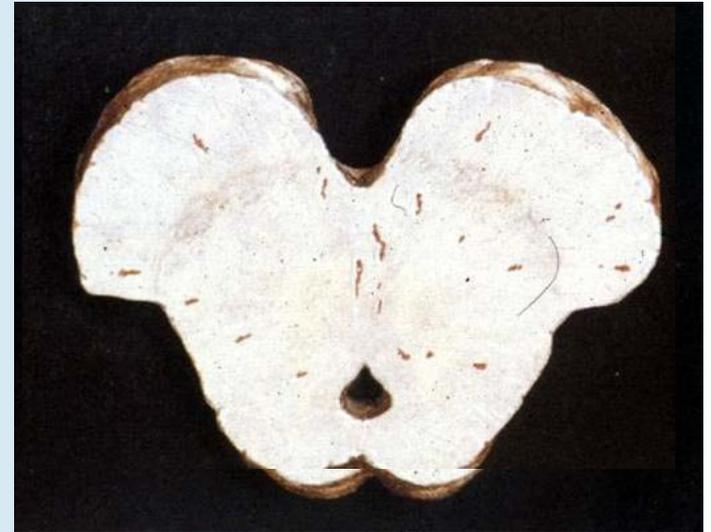
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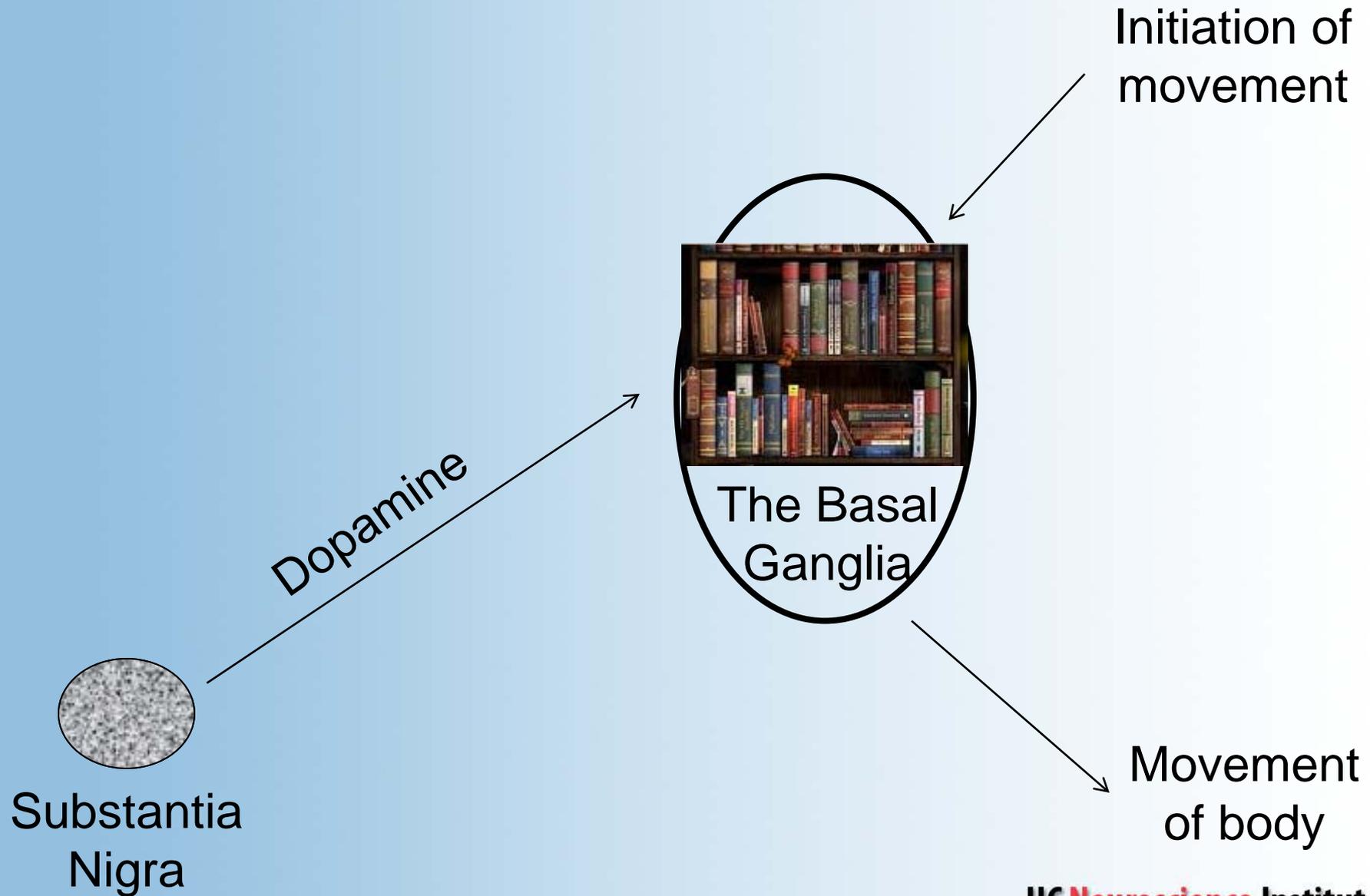
UC Neuroscience Institute

Overview of Parkinson's disease

- Parkinson's is a progressive neurological condition involving degeneration of dopamine cells in the substantia nigra in the brainstem
- First described in 1817 by James Parkinson
- Relatively common – 160/100,000
- Affects up to 850,000 people in the U.S.
- Affects 2% of people older than 80 years



Simplified diagram of movement



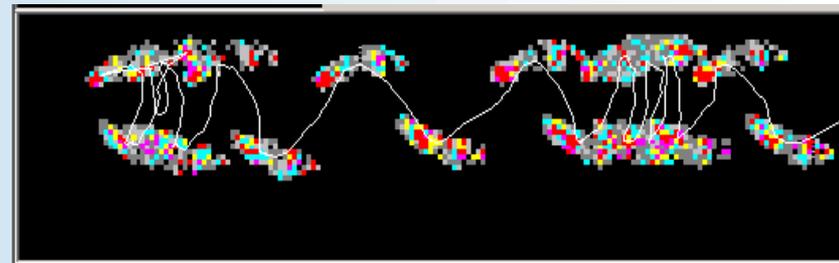
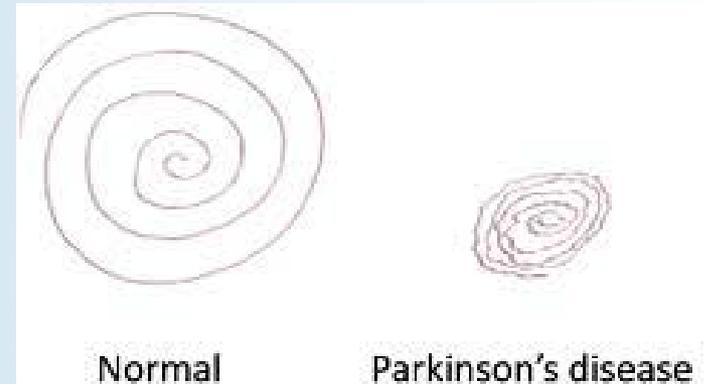
Symptoms of Parkinson's disease

- **Bradykinesia**
 - THE cardinal feature of Parkinson's disease
 - “Brady” = slow
 - “kinesia” = movement
- **Rigidity**
- **Tremor**
 - (although 25% of PD patients will not have tremor)
- **Postural instability**



Other Parkinson symptoms - motor

- Decreased arm swing
- Muscle cramps, toe curling
- Small handwriting - micrographia
- Decreased facial expression - hypomimia
- Decreased speech volume - hypophonia
- Tendency to stutter forward - festination
- Freezing of gait



Non-motor symptoms of PD

Neuropsychiatric and cognitive

- Depression
- Anxiety
- Apathy
- Cognitive difficulty/dementia
- Psychosis
- Sleep disturbances – acting out one's dreams (REM sleep behavior disorder)

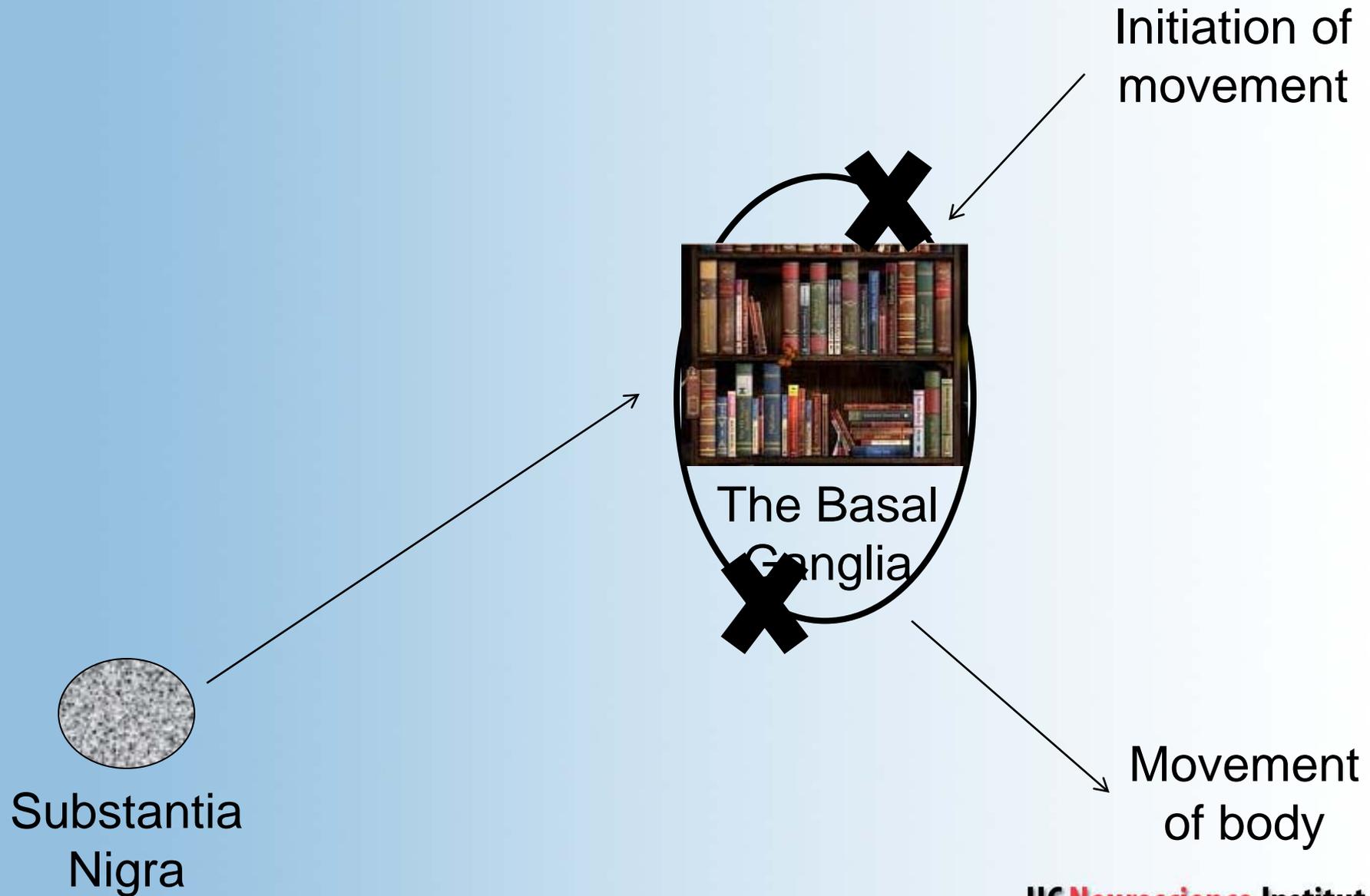
Autonomic

- Constipation
- Excessive sweating
- Orthostasis/lightheadedness
- Urinary dysfunction
- Sexual dysfunction
- Drooling

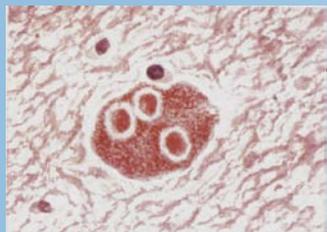
Sensory

- Decreased or absent sense of smell
- Pain
- Numbness/tingling

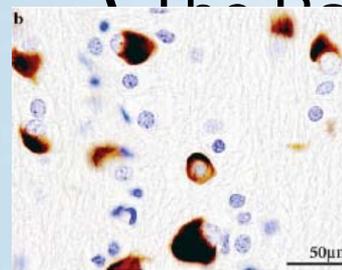
Simplified diagram of movement



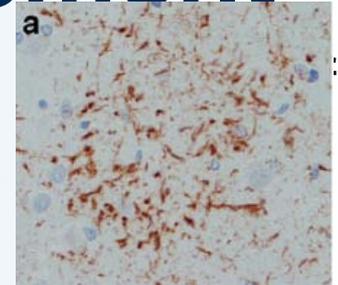
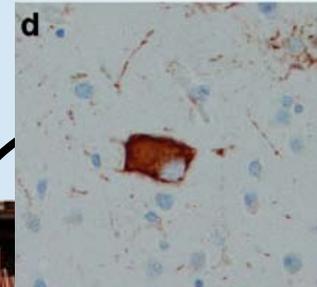
Simplified diagram of movement



Substantia Nigra



The Basal
Ganglia

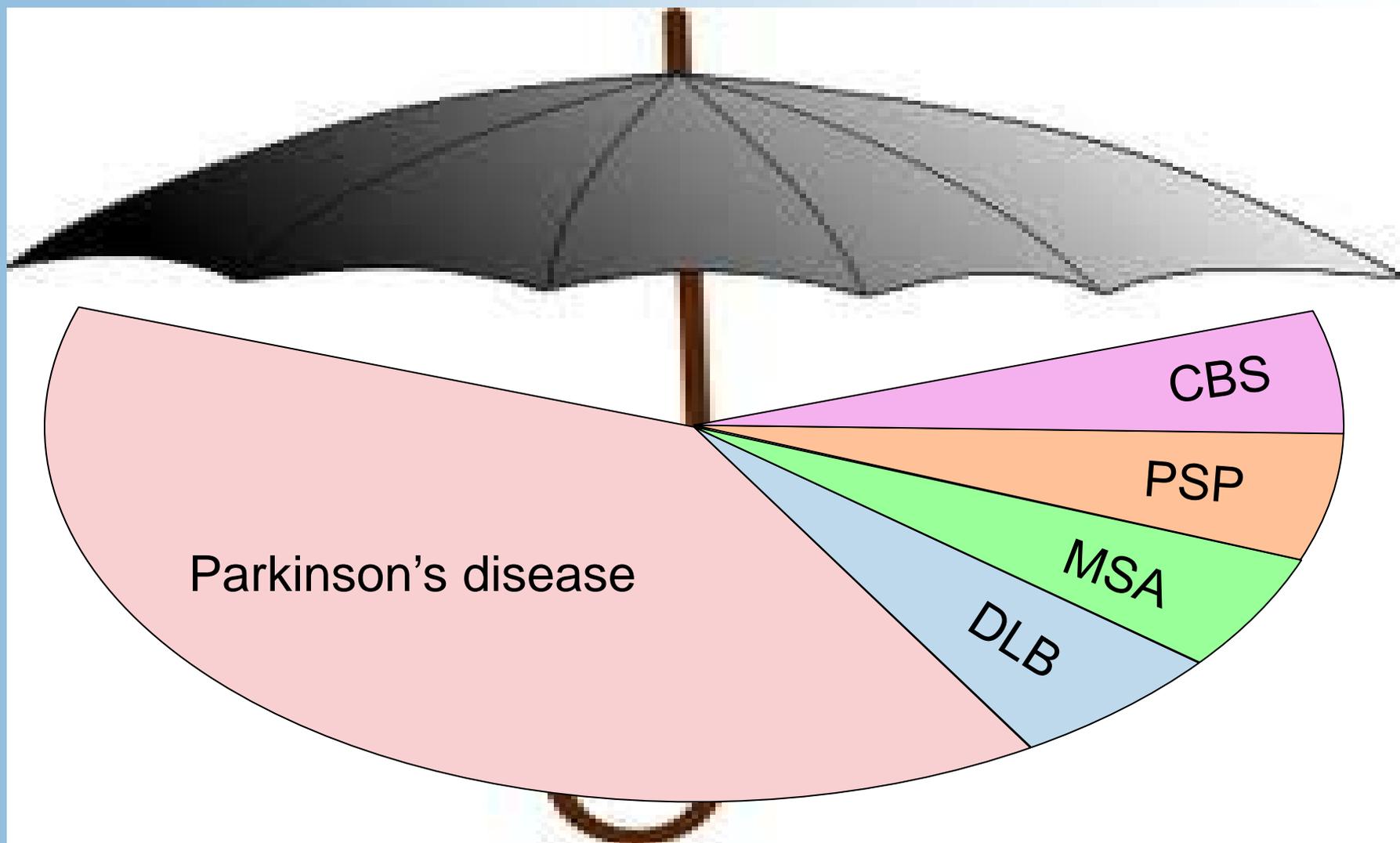


Movement
of body

Parkinsonism

- **Because other pathological brain conditions can cause similar symptoms to Parkinson's disease, we use the term "parkinsonism" to refer to the clinical symptoms of bradykinesia, rigidity, and others**

Parkinsonism



Atypical parkinsonism or Parkinson-plus syndromes

- **Dementia with Lewy bodies (DLB)**
- **Multiple System Atrophy (MSA)**
- **Progressive Supranuclear Palsy (PSP)**
- **Corticobasal Syndrome (CBD)**

Parkinson-Plus

- Taken as a whole, these conditions can progress at a different rate than Parkinson's disease (often more rapidly)
- They can have certain additional symptoms that are either
 - not usually seen in Parkinson's disease or
 - seen in PD, but they occur much earlier than expected

Parkinson-Plus

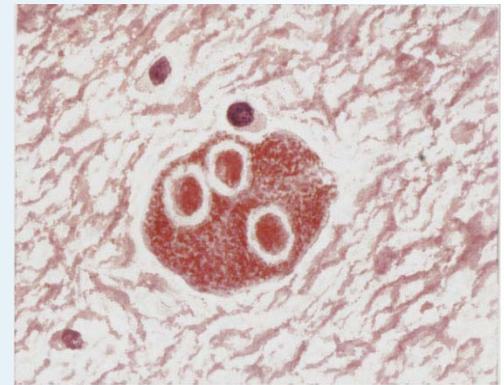
- They do not respond as well to Parkinson medications (e.g. Sinemet), and sometimes do not respond at all
- We know less about these diseases as they are all more recently recognized (over the last 40 years) and we are only just beginning to understand them

Why is the diagnosis important?

- **An accurate and early diagnosis will help to guide appropriate treatment, counsel the patient and family as to prognosis, and help us to continue to better understand these challenging conditions**
- **As the causes of these differing disorders become more clear over time, specific therapies will become available**

Dementia with Lewy bodies (DLB)

- Originally felt to be a subtype of Alzheimer's, described in 1984
- Clinically similar to Parkinson's dementia that can occur later in the course of PD
- With DLB, however, dementia usually starts at the same time as the parkinsonism
- Hallucinations are often present
- Mental status fluctuates widely during a day
- Can respond paradoxically to some anti-psychotic drugs, causing worsening of hallucinations



Multiple system atrophy (MSA)



- **Previously known as Shy-Drager syndrome first described in 1960**
- **Rare – 4/100,000**
- **Dominated by problems with the autonomic nervous system**
 - Orthostatic hypotension (low blood pressure)
 - Urinary incontinence
 - Excessive sweating
 - Erectile dysfunction
- **Some forms can have more severe coordination/balance difficulty (ataxia)**

Progressive Supranuclear Palsy (PSP)

- First described in 1964
- Rare – 5-6 people/100,000
- Early balance difficulty and falls
- Parkinsonism – slowness, stiffness, tremor
- Early cognitive impairment
- Difficulty with vertical eye movements (supranuclear palsy)
- Forced eyelid closure (blepharospasm)
- Swallowing problems
- Hypersensitivity to light
- Mood fluctuations, from laughing to crying in a matter of minutes (pseudobulbar palsy)



Corticobasal syndrome (CBS)

- Also known as corticobasal degeneration (CBD), described in 1967
- Quite rare, less common than PSP, MSA
- Similar to PSP, has early cognitive impairment
- Difficulty figuring out how to use objects or perform tasks (apraxia)
- Parkinsonism with dystonia (abnormal muscle pulling), usually very asymmetric

Treatment of Parkinson-Plus syndromes

- Levodopa (Sinemet) can be helpful in some, particularly MSA and DLB, perhaps less in PSP and CBD
- Important to identify and *prevent* complications
 - Falls
 - Swallowing/choking
- **Treat to alleviate some of the symptoms**
 - Botulinum toxin injections for dystonia, blepharospasm
 - Therapies to raise blood pressure in MSA
 - Dementia drugs helpful in DLB
 - Physical/Occupational/Speech therapies

Goals for treatment

- **The challenge is to work on improving the symptoms we can while recognizing these conditions progress and cause increasing disability**
- **Later in the disease course, home hospice is a very good resource, providing help and medical assistance in the home**

Take home points

- 1. Parkinsonism is an umbrella term encompassing Parkinson's disease, but also the less common Parkinson-plus syndromes**
- 2. The Parkinson-plus syndromes typically progress more rapidly than Parkinson's disease and have additional symptoms**
- 3. The response to medication in these syndromes is often less than in PD**
- 4. A correct diagnosis can help to understand the prognosis for a patient, and to work to *prevent* complications, and enhance patient comfort**