

Motor Abnormalities in Progressive Supranuclear Palsy (PSP) & Corticobasal Syndrome (CBS)

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What is meant by “motor?”

Motor = Movement

Motor signs or abnormalities refer to the parts of a syndrome which describe movement, whether excessive and involuntary or a lack of the ability to move normally.

Motor Signs of Parkinsonism

Bradykinesia – slowness of movement AND decrement of amplitude or speed as movements are continued

Rigidity – stiffness and resistance of the muscles with passive movement of the body

Rest tremor – a slow (4-6 Hz), sometimes “rolling” tremor in a fully resting limb

Postural instability – a loss of postural reflexes that impair balance and increase the risk of falling

Gait impairment – small steps, shuffling, reduced arm swing, freezing of gait

Progressive Supranuclear Palsy

Parkinsonism tends to have more axial (midline body) rigidity

Early and frequent falls

- The first fall is an average of 16-17 months after onset (vs 108 months in Parkinson's disease)

Eye movement abnormalities

- Especially looking up or down

PSP Posture and Gait Instability

https://www.youtube.com/shorts/oj9oAl1N_8o

Posture may be more extended at the neck and trunk, and stance has more knee extension.



Falls are more spontaneous, impulsive, and tend to occur in the backwards direction.

PSP Eye Movement Abnormalities

Vertical gaze is affected prior to horizontal

- Impaired downgaze is the most sensitive sign
- “Round the houses” sign

Speed of eye movements is affected before amplitude

- Slowing of vertical movements is often the earliest sign

<https://www.youtube.com/watch?v=LU7TC0wufhg>



<https://www.youtube.com/watch?v=gL-g7sMzwY>



Dystonia In PSP

Dystonia is characterized by muscles spasms that leads to abnormal posturing of the body

Axial Dystonia

- Cervical (neck) dystonia, often retrocollis
- Trunk involvement, w/ back extension

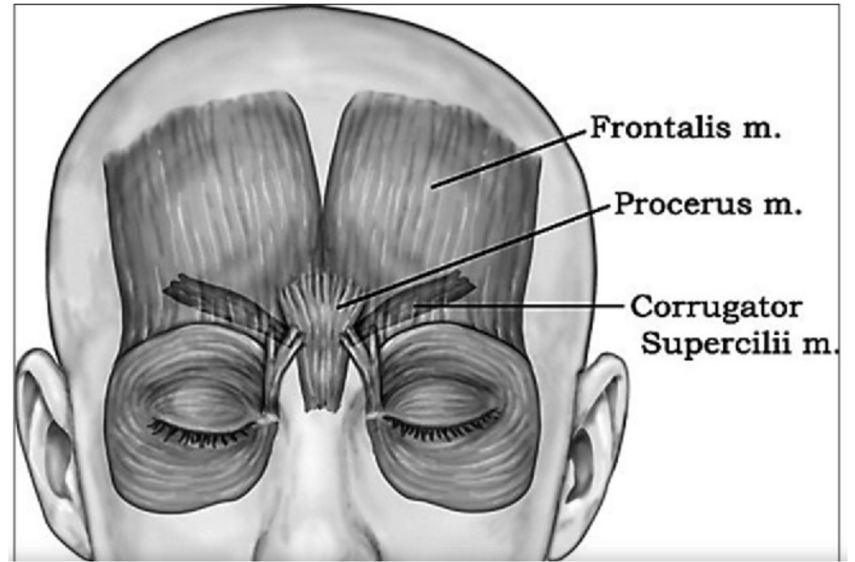
Facial Dystonia

- Blepharospasm –spasms of the eyelids
- Frontalis muscle activation can lead to a surprised appearance
- Procerus muscle activation can lead to worried appearance

Retrocollis



Facial Dystonia



Blepharospasm



<https://www.youtube.com/watch?v=fEAIMHSQJWo>

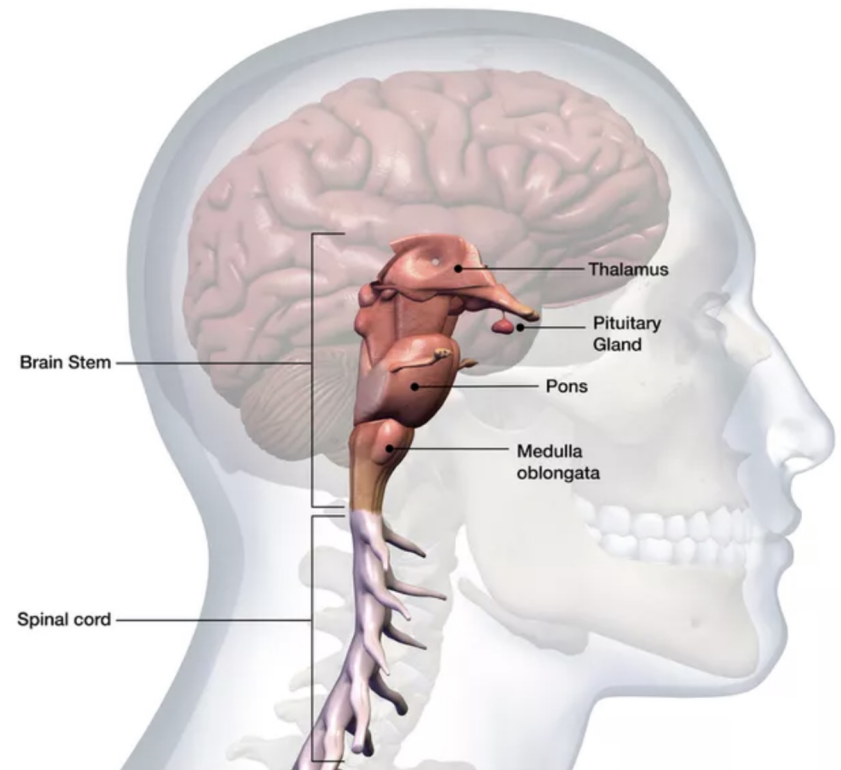
Bulbar Symptoms in PSP

Bulbar refers to the brainstem, which is affected in PSP and contains neurons that control swallowing and speech functions

Dysphagia = trouble w/ coordinated swallowing

Dysarthria = trouble with articulation of speech

- Slow, slurred, nasal, monotonous
- Stuttering, repetition, involuntary vocalizations



Hank Grebe / Getty Images

Corticobasal Syndrome (CBS)

Parkinsonism is present but onset is very asymmetric

- Akinetic, rigidity, dystonia, myoclonus

Cortex involvement

- Apraxia = “without action” – inability to perform previously familiar or learned tasks
- Cortical sensory signs – inability to identify letters or objects by touch

Alien limb phenomenon

- The limb seems to move purposefully of its own accord

Akinetic-Rigid Syndrome

Limb rigidity is very common in CBS and often present at onset

Usually asymmetric and severe

Nature of rigidity is uncertain:

- **Parkinsonism**
- **Dystonia**



<https://www.youtube.com/watch?v=HYq04aHdNJo>

Apraxia In CBS

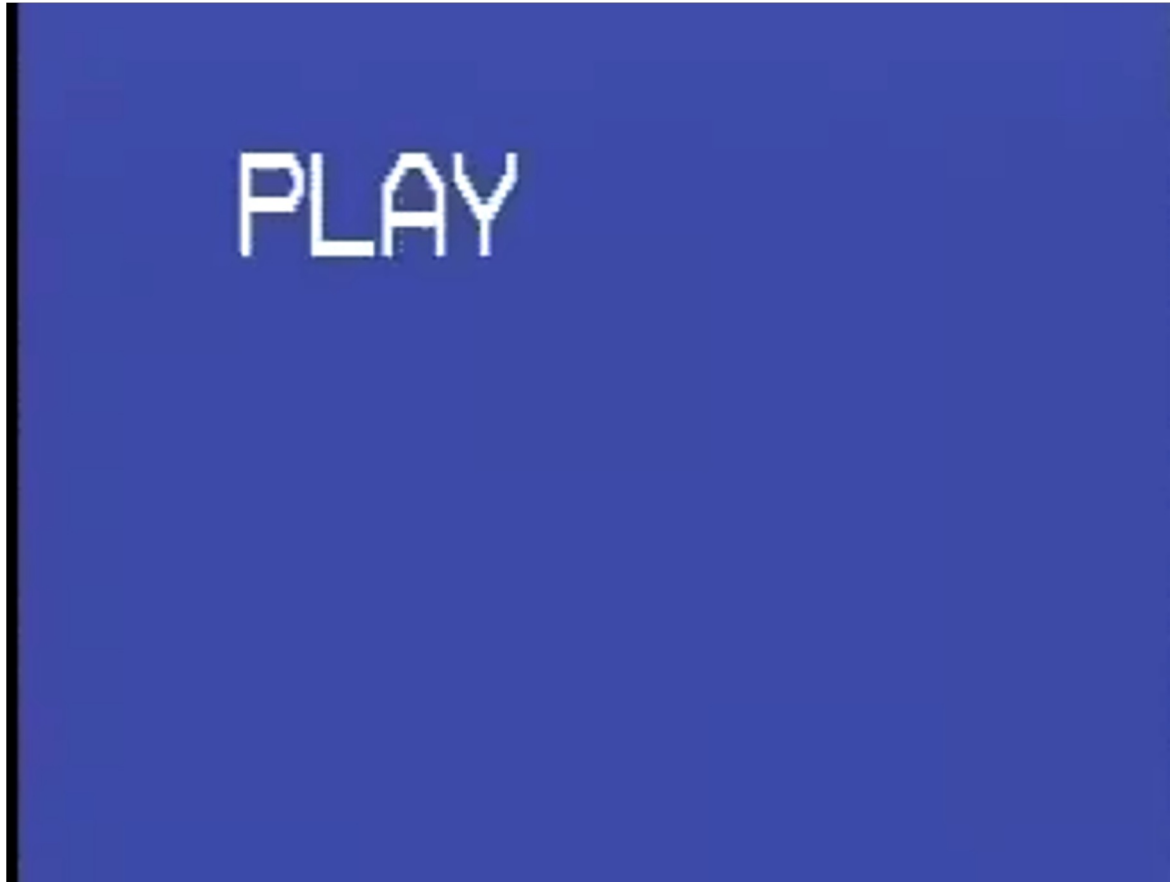
Apraxia is very common in CBS and is considered a clinical hallmark of the syndrome

Inability to perform a familiar task or execute a desired movement despite adequate strength, dexterity, and comprehension

Ideomotor = difficulty performing hand gestures (“wave goodbye”) or miming tool use (“pretend to hammer a nail”)

Limb-kinetic = inaccurate or clumsy distal limb movements

Apraxia In CBS



<https://www.youtube.com/watch?v=vTFdNk7Jl0o>

Dystonia In CBS



Dystonia is present in about 40% of people with CBS

Usually, dystonia is in an upper limb (80)%

Often associated with myoclonus

Typically occurs within the first two years of disease



Myoclonus In CBS

Myoclonus is a sudden, fast, jerk or twitch of a muscle

It is variably reported in CBS in as low as 55% and up to 93% of cases in some studies

It usually occurs in the upper extremities and occasionally the face, sometimes associated with dystonia

Often spontaneous or triggered by sensory stimulation

Myoclonus In CBS



<https://www.youtube.com/watch?v=BHeCok19jWo>

Alien Limb Syndrome

Involuntary reaching, grasping, and manipulating of a limb which seems to have purpose but is unintended

Difficulty recognizing one's limb as one's own ("alien"), especially without visual input

One manifestation is levitation of a limb

May be induced by visual or tactile stimuli

Alien Limb



<https://www.youtube.com/watch?v=ucA23SWDUXU>

https://www.youtube.com/watch?v=cN_H7uMImNY



Therapies For Motor Symptoms in PSP & CBS

Parkinsonism	Carbidopa-levodopa
Dystonia	Botox, baclofen, <i>trihexyphenidyl</i> , <i>clonazepam</i>
Gait dysfunction, falls	Physical therapy, donepezil, rivastigmine
Myoclonus	<i>Clonazepam</i> , <i>valproate</i> , <i>levetiracetam</i> , <i>zonisamide</i>
Eye movement abnormalities	Prism glasses
Swallowing problems	Speech therapy, safety precautions, diet modifications
Speech problems	Speech therapy, devices (AAC, SpeechVive, ChatterVox)

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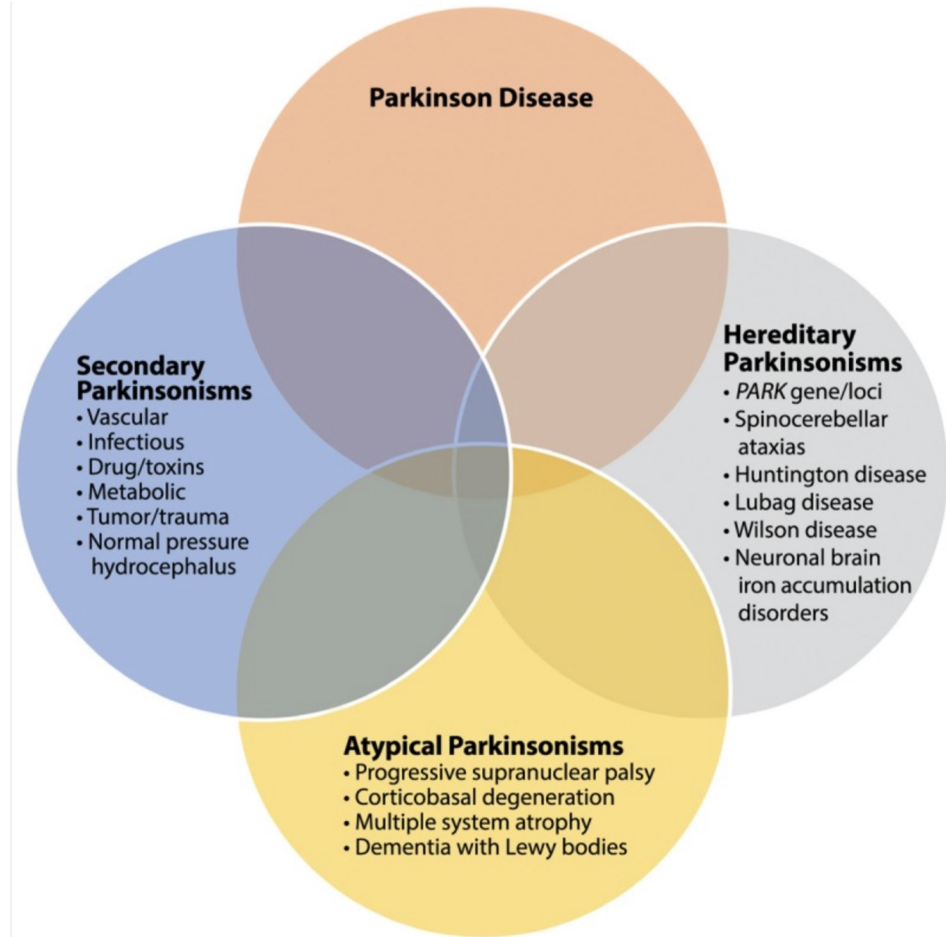
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Atypical Parkinsonism

Sometimes referred to as “**Parkinson’s Plus**”

Parkinsonism is present, plus other features that would be *atypical* if seen at the same stage in Parkinson’s disease



What About It Is Atypical?

Often more symmetric than idiopathic Parkinson's disease


Rest tremor is often not present

Response to levodopa is usually absent, minimal, or short-lived

Faster progression than Parkinson's disease

What About It Is Plus?

Types and Symptoms of Atypical Parkinson's Disease



Dementia with Lewy bodies:
Parkinson's motor symptoms, worsening dementia, hallucinations

Progressive supranuclear palsy (PSP):
Problems with balance, limited eye movements and appearing surprised in facial expression

Corticobasal ganglionic degeneration (CBS): Abnormal one-sided muscle movements, difficulty solving problems

Multisystem atrophy (MSA): Severe dizziness, impaired balance, problems with coordination, ataxia, difficulty speaking, blood pressure problems

<https://www.verywellhealth.com/atypical-parkinson-s-disease-5206945>)