Parkinson’s Disease and other related movement disorders – a video guide to diagnosis

CAN YOU HELP?
After originally planning to use patient videos that had been uploaded to YouTube, the unforeseen complexities of gaining copyright for this material means that is unfortunately no longer an option. If you or a colleague have some footage you could contribute to the project - with the appropriate patient permissions of course - then please do contact the Academy, info@neurologyacademy.org

Parkinson’s Disease Masterclass November 2017

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Ideopathic Parkinson’s Disease

Core Motor Symptoms

Bradykinesia

Generalised slowness of movement
Usually starts with decreased manual dexterity of the fingers
In the legs get dragging of feet, shorter shuffling steps or unsteadiness
In examining assess the speed, amplitude and rhythm of finger tapping, hand gripping, pronation-supination of hand movements and heel and toe tapping
Ideopathic Parkinson’s Disease

Core Motor Symptoms

Bradykinesia

As the disease progresses get **Gait freezing and festination** (an irresistible impulse to take much quicker and shorter steps and therefore to adopt unwillingly a running pace)
Ideopathic Parkinson’s Disease

Core Motor Symptoms

Rigidity

Increased resistance to passive movement
Often begins unilaterally (on same side as the tremor (if present))
Many patients develop cogwheel rigidity (a ratchety pattern of resistance and relaxation as the limb is moved through its full range of motion)
Can affect any part of the body – striatal hand (extension of the proximal and distal interphalangeal joints and flexion of the metacarpophalangeal joints), decreased arm swing and stooped posture
Ideopathic Parkinson’s Disease

Core Motor Symptoms

Tremor

Pill rolling rest tremor most noticeable when tremulous limb is supported by gravity and not engaged in purposeful activities
Can be present with action but generally more severe at rest
Some have a re-emergent tremor when the tremor re-emerges after several seconds and has a frequency similar to the rest tremor
Ideopathic Parkinson’s Disease

Core Motor Symptoms

**Tremor**

In early PD the tremor often intermittent, starting unilaterally and spreads contralaterally after several years.

Distracting the patient by asking to perform mental calculations or voluntary repetitive movements of the contralateral limb often accentuates the tremor.

Can also involve the legs, lips, jaw and tongue but rarely the head (contrast this tremor with that of [Essential Tremor](#) and [Dystonic Tremor](#)).
Ideopathic Parkinson’s Disease

Core Motor Symptoms

Postural Instability

Usually does not appear until later in the course of PD. Tested with the ‘Pull – test’- patient with normal postural reflexes should be able to maintain balance and retropulse (step backwards) no more than one step. Patients with PD and postural instability are likely to fall or take multiple steps backwards. Patients who fall earlier most likely have another Parkinsonian syndrome such as Progressive Supranuclear Palsy or Multisystem Atrophy.
Ideopathic Parkinson’s Disease

Core Motor Symptoms

- Bradykinesia
- Rigidity
- Tremor
- Postural Instability

Pre – Motor symptoms of PD
Ideopathic Parkinson’s Disease

Other Motor Features

Craniofacial
- Hypomimia
- Decreased spontaneous eye blinking

Speech Impairment
- Hypokinetic dysarthria
- Hypophonia
- Palilalia

Dysphagia
Sialorrhoea
Ideopathic Parkinson’s Disease

Other Motor Features

**Visual**
- Hypometric saccades
- Impaired vestibulocular reflex
- Impaired upward gaze and convergence
- Eye lid opening apraxia
Ideopathic Parkinson’s Disease

Other Motor Features

Musculoskeletal

- Micrographia
- Dystonia
- Myoclonus
- Stooped Posture
- Camptocormia
- Pisa Syndrome
- Kyphosis and scoliosis
Ideopathic Parkinson’s Disease

Non - Motor Symptoms

Cognitive Dysfunction and Dementia

• Approx 80% will develop dementia during the disease
• Older age and severity of PD motor symptoms are associated with an increased risk of developing dementia
• The dementia is classically considered a subcortical dementia
• Dementia usually occurs late in the course of PD (compare this with Dementia with Lewy Bodies)
Ideopathic Parkinson’s Disease

Non - Motor Symptoms
Cognitive Dysfunction and Dementia

• Problems of **Executive function** (decision making and multi-tasking) are often the first signs of disturbed cognition.

• Clinically, patients are diagnosed with PD dementia if their illness begins with PD and they develop dementia at least one year after the onset of Parkinsons motor symptoms.
Ideopathic Parkinson’s Disease

Non - Motor Symptoms

Psychosis and Hallucinations

• Visual hallucinations are the most common psychotic symptom
• Can be attributable to PD itself or to the anti-Parkinsonian Treatment (more likely with Dopamine agonists)
• Delusions are also prominent and are usually paranoid in nature
Ideopathic Parkinson’s Disease

Non - Motor Symptoms

Mood Disorders

• Depression - very common and often predates the diagnosis
• Anxiety
• Apathy - less prevalent in those with higher levodopa equivalent dosage (mainly dopamine agonists)
Ideopathic Parkinson’s Disease

Non - Motor Symptoms

Sleep Disturbance

• Insomnia
• Daytime sleepiness with sudden onset of sleep attacks
• Restless legs syndrome
• **REM Sleep Behaviour Disorder**
Ideopathic Parkinson’s Disease

Non - Motor Symptoms

Sleep Disturbance

The most common causes for sleep awakenings in PD are nocturia, difficulty turning over in bed, cramps, vivid dreams or nightmares and pain. Tremor may also contribute as does painful dystonias in some

Fatigue
Ideopathic Parkinson’s Disease

Non - Motor Symptoms

Autonomic Dysfunction

- Postural hypotension
- Constipation
- Dysphagia
- Diaphoresis
- Urinary difficulties
- Sexual dysfunction

Also present in MSA but generally more severe than PD
Ideopathic Parkinson’s Disease

Non - Motor Symptoms

**Olfactory Dysfunction**
Common and may precede motor symptoms or occur relatively early in the course of PD
May not be noticed by patients

**Pain**
Painful sensory symptoms common
- lancinating, burning, tingling, generalised or localised
Tends to correlate with motor fluctuations
Ideopathic Parkinson’s Disease

Non - Motor Symptoms

Pain

Dystonia which is often painful can occur in early PD or when levodopa wears off
Morning dystonia affecting the foot is a common ‘off’ response to abstinence from levodopa during sleep
Progressive Supranuclear Palsy

Main Features

Progressive supranuclear ophthalmoplegia
Gait disorder, postural instability and falls
Dysarthria
Dysphagia
Rigidity
Frontal Cognitive disturbance

Mean age of onset 65yrs which is older than PD and almost no cases < 40yrs
Progressive Supranuclear Palsy

Gait

Stiff and broad based
Tendency to have their knees and trunk extended (as opposed to flexed posture of PD)
Arms slightly abducted
Impulsivity (probably from frontal lobe involvement)
Tend to pivot quickly rather than turn en block
Usually fall backwards
Gait and balance problems may worsen with Levodopa treatment
Progressive Supranuclear Palsy

Oculomotor Findings

Supranuclear opthalmoplegia may take as long as 10 years to develop
Get a slowing of vertical saccades followed by a limitation of saccadic range
Concomitant limitation of lateral gaze is often present
Pursuit movements of the eyes are slow, jerky, and hypometric with unstable fixation
The ophthalmoparesis is initially overcome by the Oculocephalic (Doll’s eyes) manoeuvre
Progressive Supranuclear Palsy

Oculomotor Findings

- Blepherospasm
- Eye lid opening apraxia

The combination of rare blinking, facial dystonia, and gaze abnormalities leads to the development of a classical facial expression of perpetual surprise or astonishment.
Progressive Supranuclear Palsy

**Motor Involvement**
Bradykinesia with marked micrographia
Axial rigidity more apparent especially neck and upper trunk
Retrocollis in some patients
One third have pyramidal signs with hyperreflexia and Babinski sign
Spastic dysarthria, dysphonia and dysphagia are profound in the middle to later stages
Stuttering and **Palilalia**
Progressive Supranuclear Palsy

Cognitive Abnormalities

Early and severe **frontal (executive)** deficits is a common finding.

Pseudobulbar palsy is another characteristic feature.

Hoarse groaning voice.

REM sleep behaviour disorder is only infrequently seen in PSP, and this combined with intact olfaction can help differentiate PSP from PD and MSA.
Most often affects the hands and arms bilaterally but can be asymmetric.

Can also affect the head and voice and uncommonly the face, legs, and trunk.

Varies from a low amplitude, high frequency postural tremor of the hands to a much larger amplitude tremor that is attenuated by particular postures and actions.

Becomes immediately apparent in the arms when they are held outstretched and typically increases at the very end of goal directed movements.
Essential Tremor

Tremor of the legs unusual and PD more likely
Head tremor may be ‘yes-yes’ or horizontal ‘no-no’ and is usually associated with tremor of the hands or voice
By definition tremor should be the only neurological manifestation of ET
Can get cog-wheeling but without rigidity

Some patients develop enhanced physiological tremor due to anxiety
Essential Tremor

A positive family history (autosomal dominant) is supportive of the diagnosis.

There may be a beneficial response to alcohol.

DAT scan can reliably distinguish PD and other Parkinsonian syndromes (MSA, PSP and CBD) from Essential Tremor.
Multi-System Atrophy

Main Features

Akinetic-rigid Parkinsonism
Autonomic Failure
Cerebellar ataxia
Pyramidal signs
Rapid progression regardless of dopaminergic treatment

Two phenotypes
MSA - P (Parkinsonism) and MSA- C (Cerebellar)
Multi-System Atrophy

MSA - P

Akinesia/bradykinesia
Rigidity
Postural instability and falls (usually within 3 years of motor onset)
Irregular, jerky postural and action tremor
Can get a rest tremor in up to one third of patients
Multi-System Atrophy

**MSA - P**

Also can get:

- Stimulus-sensitive cortical myoclonus
- Hemiballism and chorea
- Dystonia unrelated to Dopaminergic treatment
- Orofacial dystonia or dyskinesia
- PISA syndrome
- Camptocormia

Speech – increased in pitch and quivering, strained element
Multi-System Atrophy

MSA - C

Gait ataxia
Limb ataxia
Ataxic dysarthria
Cerebellar dysfunction of eye movements
Cerebellar scanning dysarthria
Multi-System Atrophy

- Sleep and breathing disorder
  - At least 1/3 get RBD
  - 1/3 develop nocturnal and diurnal laryngeal stridor (and a risk for sudden death)

- Also early features (and earlier and more severe than PD) get
  - Urinary frequency and urgency
  - Incontinence
  - Incomplete bladder emptying

- Dysautonomia is a feature of both (Urinary dysfunction, Orthostatic hypotension)

- Dysphagia common in both types
Multi-System Atrophy

Cognitive Function

Relatively well preserved compared with PD and other atypical Parkinsonian syndromes

Can develop emotional incontinence (pseudobulbar affect) – crying inappropriately without sadness or laughing inappropriately without mirth
Multi-System Atrophy

Have an association with Raynaud’s phenomenon

Usually there is a poor or un-sustained response to Levodopa

Levodopa induced unilateral facial dystonic spasms are particularly suggestive of MSA
Dementia with Lewy Bodies

- Fluctuating cognition with pronounced variation in attention and alertness
- Recurrent visual hallucinations that are typically well formed and detailed
- REM sleep behaviour disorder which may precede cognitive decline
- One or more spontaneous cardinal features of Parkinsonism (Bradykinesia, rigidity or rest tremor)
Supportive Clinical Features

- Severe sensitivity to anti-psychotic agents
- Postural instability and repeated falls
- Syncope or other transient episodes of unresponsiveness
- Severe autonomic dysfunction
- Hypersomnia
- Hyposmia
- Delusions
- Hallucinations
- Apathy, anxiety and depression
Dementia with Lewy Bodies

Cognitive Dysfunction

• Often the presenting symptom
• Early impairment in attention and executive function (unlike AD when memory loss is the first feature)
• Early symptoms include driving difficult, misjudging distances, impaired job performance
• Early appearance of impaired figure copying (overlapping pentagons), clock drawing and serial 7’s (or spelling WORLD backwards)
• Fluctuation in cognition and level of alertness may occur early in the course of DLB
Dementia with Lewy Bodies

Cognitive Dysfunction

- Fluctuation in cognition and level of alertness may occur early in the course of DLB
- There may be a brief decline in ability to perform an ADL and may be dramatic enough to raise the possibility of a stroke or seizure
- Patients can appear to ‘black out’ or lose consciousness, become confused or behave in a bizarre manner
- Can become excessively somnolent
Dementia with Lewy Bodies

Cognitive Dysfunction

- Visual hallucinations are an early sign in DLB and may precede Parkinsonism

- RBD commonly associated with DLB often early in the course of the disease. Can precede the diagnosis of DLB by 20 years
Corticobasilar Degeneration

Main Features
Progressive asymmetric movement disorder
Symptoms initially affect one limb
Various combinations of:
  - akinesia, extreme rigidity, dystonia, focal monoclonus,
  - ideomotor apraxia and alien limb phenomena

Cognitive Impairment
Parkinsonian features may present later
Cognitive features include executive dysfunction, aphasia, apraxia,
behavioural change and visuospatial dysfunction with relatively
preserved episodic memory
Corticobasilar Degeneration

Main Features

Rigidity can be profound in the limb (and less so axially)
Dystonia involves abnormal posturing of the hand and foot (toe curling and foot inversion) that quickly become fixed
Along with apraxia the affected limb is rendered useless
Gait is variable and can be similar to PD or be a wide based ‘frontal lobe’ or freezing gait
Tremor less frequent than PD and more postural irregular and jerky
Dysarthria is an early feature
Corticobasilar Degeneration

Main Features

Oculomotor Dysfunction
- Abnormal eye movements in around 1/3 at presentation
- Pursuit eye movements slow and saccadic
- Vertical saccades usually normal (unlike PSP)

Cortical Dysfunction
- Characteristic of CBD
- Cognitive impairment and Behavioural changes
- Limb apraxia and alien limb
- Aphasia
- Depression
End of teaching series

Thanks for reading!
CBD – Alien limb
CBD1
Lewy Body Dementia

Lewy Body Dementia
SOME INTERESTING FACTS

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clinical, education, research

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Excerpt of "Lewy Body Dementia: What Everyone Needs to Know" with Teepa Snow, MS, OTR/L, FAOTA
Multi system atrophy 2

Approach to the Evaluation of MSA (Multiple System Atrophy)

Brad Hiner, MD

Medical College of Wisconsin

The MSA Coalition annual conference
September 7, 2013 Milwaukee, WI

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PSP 2
PSP Facial appearance and Gait
PSP Gait
Tremor and Alcohol

33 Years Old

Diagnosed by a Neurologist 4 months ago

Date Filmed 12.29.12

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Supranuclear palsy - PSP
Benign Essential Tremor and DBS
Essential Tremor
Symptoms of PSP, CBD and MSA
Dystonia
Executive Function
PD – Rigidity, Bradykinesia and tremor
PD Postural Instability
Freezing of Gait
Bradykinesia
Pisa Syndrome
Eyelid opening apraxia
Saccades

Horizontal and Vertical Saccades
Pre motor symptoms for PD
PD Rigidity
Hypommmia
Camptocormia
Palilalia

Echolalia - involuntarily repeating another's words, phrases, or sounds.