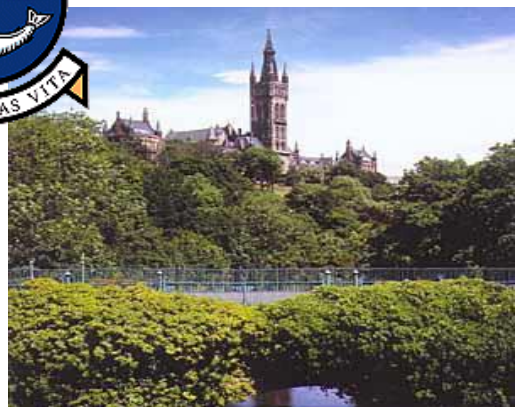
Differential Diagnosis of Hypokinetic Movement Disorders

Dr Donald Grosset
Consultant Neurologist - Honorary Professor
Institute of Neurological Sciences - Glasgow University



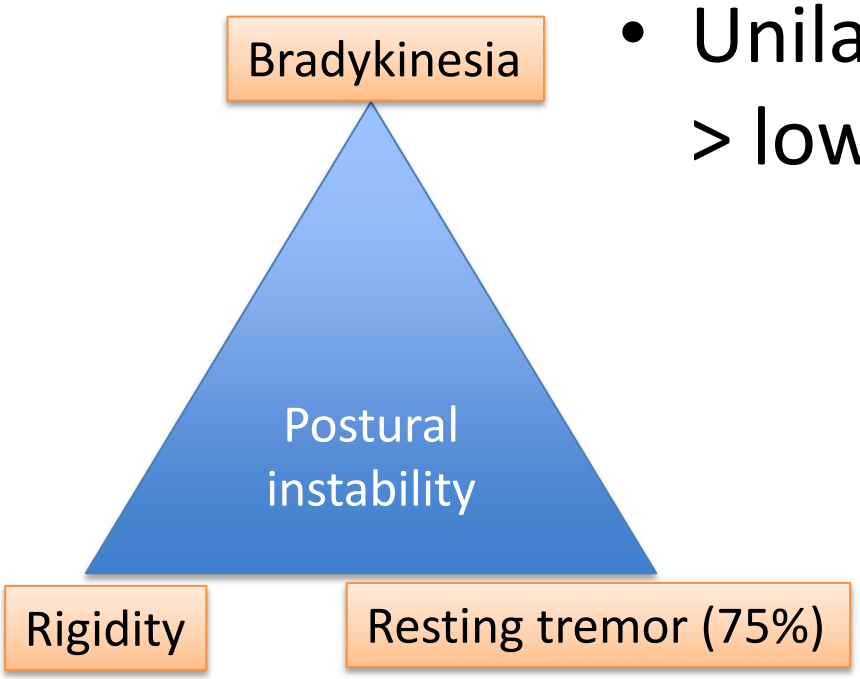
Hypokinetic

- Parkinson's Disease
- Multiple System Atrophy (MSA)
- Progressive Supranuclear Palsy (PSP)
- Corticobasal Ganglionic Degeneration (CBGD)
- Dementia with Lewy Bodies (DLB)
- Vascular parkinsonism
- Drug-induced Parkinsonism

Hyperkinetic

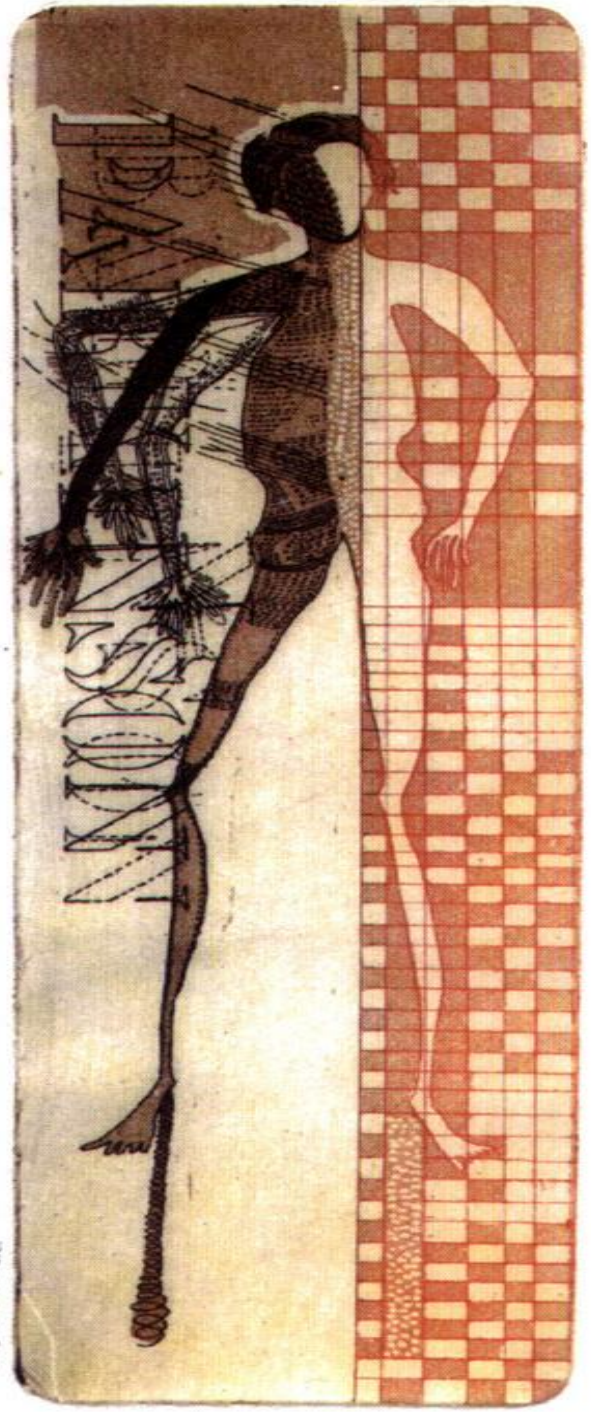
- Ataxia
- Chorea
- Dystonia
- Huntington's Disease (HD)
- Myoclonus
- Tardive Dyskinesia / Dystonia
- Tics/Tourette’s Syndrome
- Tremor
- Wilson's Disease

Diagnosis of PD



- Unilateral or asymmetrical, upper > lower limb

- Non-motor
  - Olfactory
  - RBD
  - Constipation
  - Depression



Artwork by patient illustrates:

Tremor

Jerky slow movement

Leg sticking or dragging

Unilateral onset

Spreads to other side (slowly)

Arm more involved than leg

Facial involvement

Diagnosis of PD

(Postuma et al 2015)

Absolute exclusion criteria:

- Unequivocal cerebellar abnormalities, such as cerebellar gait, limb ataxia
- Downward vertical supranuclear gaze palsy
- Diagnosis of probable behavioral variant frontotemporal dementia or primary progressive aphasia
- Parkinsonian features restricted to the lower limbs for more than 3 y
- Treatment with a dopamine receptor blocker or a dopamine-depleting agent in a dose and time-course consistent with drug-induced parkinsonism
- Absence of observable response to high-dose levodopa despite at least moderate severity of disease
- Unequivocal cortical sensory loss (eg. graphesthesia), clear limb ideomotor apraxia, or progressive aphasia

- Normal functional neuroimaging of the presynaptic dopaminergic system

- Other alternative condition known to produce parkinsonism and plausibly connected to the patient's symptoms

MSA: Case 1

- Ataxic
- Facial masking
- Initially minimal parkinsonism

- Progression quite rapid (18 months)
- Ataxic arms
- Intention tremor
- Truncal ataxia
- Gait ataxia

MSA: Case 2

- Cerebellar speech: scanning

- Autonomic systems:
  - Bladder
  - Blood pressure

MSA: Montage

- Unilateral tremor
- Rapidly progressive
- Flexed neck, anterior or rotated

MSA

Parkinsonism

MSA-P

Cerebellar and/or Autonomic

MSA-C

Compared to PD:

More symmetrical
More rapidly progressive

Cerebellar features (speech, gait)

Earlier and more severe autonomic symptoms

Tremor less marked, sometimes irregular
Neck and orofacial dyskinesia (rather than limbs)

Less cognitive impairment

PSP

- Case 1
  - Failure of up and downgaze
  - Spastic face, dysarthric
  - Oculocephalic manoever overcomes gaze paresis
- Case 2
  - Gait slow
  - Axial and neck extension
  - Stiff face and dysarthria
  - Vertical gaze palsy
- Case 3
  - Slow walking
  - Truncal movements difficult
  - Falling backwards
  - Neck extension
  - Blepharospasm
  - Gaze paresis

PSP: Case 4

- Staring expression
- Dysarthric speech
- Falling backwards

PSP

Parkinsonism  Bulbar problems:

Swallowing, speech, eye movements

Cognitive decline

Compared to PD:

More symmetrical
More rapidly progressive

Bulbar features (early speech and swallowing problems)
Gaze paresis, often with functional impact

Extension in neck and trunk, upright posture

Tremor less marked

PSP versus PD: clinical appearance

- Contracted rather than flaccid facies
- Undirected rather than staring gaze
- Erect rather than stooped posture

- axial and proximal rigidity

- Dysarthria: spastic and ataxic versus hypophonic
- Absence of rest tremor

CBD

- Case 1
  - Parkinsonian mask face, with open mouth
  - Fixed adducted posture R arm
  - Flexed fingers
- Case 2
  - Parkinsonian face
  - Asymmetrical – contracture R thumb
  - Slight tremor fingers of R hand

CBD

Parkinsonism  Cortical involvement:
Cognitive decline, dysphasia, apraxia, spasticity

Compared to PD:

More symmetrical
More rapidly progressive

Earlier cognitive decline

Earlier dystonia and blepharospasm
Earlier contractures (often asymmetric)
Alien limb

Myoclonus

DLB

- Wife younger (!)

- Stooped parkinsonian posture and gait
- Reduced facial expression
- Gait stiff
- Slow progression
- ‘Moment of clarity’ – lucid periods

DLB

Parkinsonism  Earlier cognitive decline

Compared to PD:

Dementia predates PD or within 1 year

Fluctuations: Lucid periods

Earlier and more florid hallucinations

Spectrum with PDD

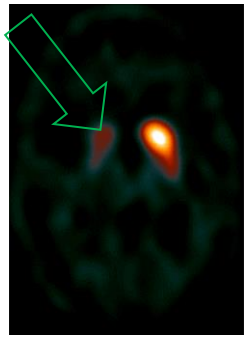
Vascular parkinsonism

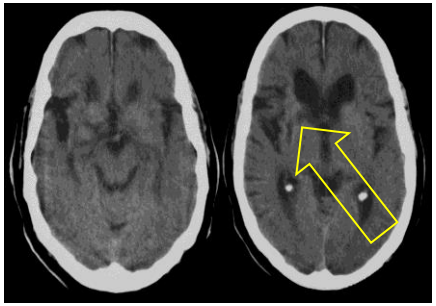
- Supporting features: subcortical vascular changes – lacunar infarcts, small vessel disease, normal FP-CIT SPECT or punched lesions

- Lower body, more symmetrical

- "Stepwise"

- Less dopa-responsive





Drug-induced parkinsonism

Dopamine-depleting drugs

- Antinauseants
- Antipsychotics
- Antiepileptic – sodium valproate (depakote)

May be asymmetrical

May unmask Parkinson's disease

Dystonia mimicking PD

- Dystonia (but may be subtle)
- Thumb extension tremor
- “Flurries” or task/position specific tremor
- Head tremor
- Dystonic voice
- No progression to develop features other than tremor and dystonia
- No clear bradykinesia

Summary – “Parkinson’s Plus”…

- MSA: + autonomic + cerebellar
- PSP: + cognitive + bulbar
- CBD: + cortical
- DLB: + (early) dementia

Caution: Co-morbidity (cerebrovascular) and Dual (or triple) pathology

Diagnosis of PD

(Postuma et al 2015)

Absolute exclusion criteria:

- Unequivocal cerebellar abnormalities, such as cerebellar gait, limb ataxia — MSA
- Downward vertical supranuclear gaze palsy — PSP
- Diagnosis of probable behavioral variant frontotemporal dementia or primary progressive aphasia — FTD*
- Parkinsonian features restricted to the lower limbs for more than 3 y — VP
- Treatment with a dopamine receptor blocker or a dopamine-depleting agent in a dose and time-course consistent with drug-induced parkinsonism — DIP
- Absence of observable response to high-dose levodopa despite at least moderate severity of disease
- Unequivocal cortical sensory loss (eg. graphesthesia), clear limb ideomotor apraxia, or progressive aphasia — CBD
- Normal functional neuroimaging of the presynaptic dopaminergic system
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*FTD – 20% of behavioural variant FTD have parkinsonism in later course