Differential Diagnosis of Hypokinetic Movement Disorders

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

- 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

### Hyperkinetische

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

- Bradykinesia
- Rigidity
- Resting tremor (75%)
- Postural instability

- 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. graphesthesesia), 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
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 manoeuver 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
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*FTD – 20% of behavioural variant FTD have parkinsonism in later course