Approach to the patient with a movement disorder

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The characteristic feature of all movement disorders is an abnormality of the form and velocity of movements of the body. The use of the term "movement disorder" in neurology has become synonymous with basal ganglia disease and extrapyramidal features. Although it is true that many movement disorders arise from pathology within the basal ganglia, disorders such as myoclonus may also arise from other structures. Abnormalities of movement may be the only manifestation of a disease (for example, essential tremor) or may be part of a more widespread neurological disorder (for example, Creutzfeldt-Jakob disease). Basal ganglia disease is also commonly associated with neuropsychiatric symptoms and these may have a greater impact upon the patient and their family than the movement disorder itself. As in all aspects of neurology, it is important not to divorce the disorder of movement from general medical problems, since these may be directly or indirectly related (for example, chorea in systemic lupus erythematosus; myoclonic ataxia in coeliac disease; drug-induced parkinsonism caused by metoclopramide used to treat a hiatus hernia).

Classification & definitions

The key to success in diagnosing and managing these patients is to establish the phenomenology of their movement disorder. Although the broad division of patients into those who move too much (hyperkinetic disorder) or move too little (hypokinetic, or akinetic-rigid disorder) is simple enough, to the inexperienced physician, differentiating jerky dystonia from tremor, or tics from chorea, for example, may not always be straightforward. There may also be a mixed movement disorder present, such as myoclonic dystonia, for instance. Definitions of commonly encountered movement disorders are listed in Table 1. Athetosis (a writhing, sinuous distal limb movement) is a term gradually falling out of use, as these movements are more economically classified as dystonic or choreo-dystonic.

Historical features and examination (Table 2)

When approaching the patient with a movement disorder, the value of a careful history and examination can never be under-stated, even if the diagnosis may seem obvious from the moment the patient first walks in to the consultation room. A videotape recording of the movement disorder may be helpful, particularly in the case of a "compound" or "mixed" problem. It is not uncommon for the rest of the neurological examination to be normal in patients reporting a movement disorder; in other words, "what you see is what you get". If no problem is apparent, consider whether the complaint is highly action specific (for example a task-specific dystonia) and may therefore have been overlooked on the routine examination (any excuse to get a golf club out in clinic, or even a violin!). Failing this, asking the patient and their family to record a home video-segment when the problem occurs may be very revealing.

Always consider drugs, both past and present, as a potential cause for the movement disorder. Tardive dyskinesias (commonly stereotypic movements, often orofacial in distribution, although a broad spectrum of tardive drug-induced movement disorders, from tics to myoclonus, has been described) may develop after relatively short exposure to an offending dopamine receptor blocking agent (DRBA) but persist for many years. A full list of medications previously taken by the patient should be obtained from the GP, if necessary. Approximately 80% of drug-induced parkinsonism will resolve within eight weeks of discontinuing a DRBA, although recovery up to 18 months has been reported. If causality is suspected, always check with the hospital drug information service. For example, while DRBAs are well know to cause parkinsonism, a link with agents like amiodarone and cinnarizine, is less widely recognised.

Analysis of the following characteristics (adapted from Kishore and Calne 1997) may assist the diagnosis:

1. Specific distribution: for example, restless legs syndrome (RLS, although this is now known as restless limb syndrome since symptoms may also be reported in the upper limbs as well!) and painful legs and moving toes (PLMT). Parkinson's disease is typically asymmetric in onset.
2. Specific actions: for example task - specific tremor and dystonia (don't forget to ask the patient to write or pick up a cup of water if history suggests this might be helpful).
3. Speed: for example-

slow	intermediate	fast
parkinsonism, dystonia & dystonic tics	chorea, tremor	myoclonus, myoclonic tics

4. Rhythm: continuous – for example, tremor, PLMT; or intermittent, for example, asterixis ("negative myoclonus").
5. Relation to posture: for example, orthostatic tremor (presents as unsteadiness when standing still but improved when walking).
6. Relation to sleep: few movement disorders persist during sleep; examples include palatal tremor and segmental myoclonus.

Table 1 Definition of Commonly Encountered Movement Disorders

Movement	Definition
parkinsonism	a clinical syndrome with bradykinesia as the defining feature, almost always accompanied by rigidity, and often by tremor
dyskinesia	may be applied to any involuntary movement (although often used to refer to drug-induced choreas and dystonias)
tremor	a rhythmical, involuntary oscillatory movement of a body part; may be qualified by addition of a descriptive term (e.g. resting, postural)
chorea	a quick, irregular, semi-purposive and predominantly distal involuntary movement (patient may look "fidgety")
dystonia	an abnormal movement characterised by sustained muscle contractions, frequently causing twisting and repetitive movements or abnormal postures
ballism	a proximal, high amplitude movement, often violent and flinging in nature; usually unilateral in nature and often resolves through a choreic phase
tic	an abrupt, jerky non-rhythmic movement (motor tic) or sound (vocal tic) that is temporarily suppressible by will power; tics may be simple or complex
stereotypy	purposeless voluntary movements carried out in a uniform repetitive fashion at the expense of other activity (e.g. hand wringing, clapping, mouthing)

7. Relation to voluntary movement: for example – action tremor and action dystonia.
8. Associated sensory symptoms: PLMT, RLS and phantom dyskinesias; tics may be associated with an vague discomfort or unusual sensation in the prodrome before the movement.
9. Suppressibility: volitional in tics (but associated with increasing unease and rebound worsening of tics upon release), by sensory tricks in dystonia (including the "geste antagoniste") and by activity in rest tremor.
10. Aggravating or precipitating factors: stress and anxiety are of no discriminating value in that they worsen all movement disorders; myoclonus may be worsened by specific stimuli e.g. sudden, loud noise; heavy carbohydrate-containing meals and fatigue may precipitate paroxysmal *non*-kinesogenic dystonia; rapid movement triggering paroxysmal kinesogenic dyskinesia.
11. Ameliorating factors: alcohol may relieve essential tremor and myoclonic-dystonia, sometimes quite dramatically; walking backwards or running may improve a dystonic gait, leading the unwary to suspect a non-organic cause.

Table 2 Historical and Examination Features to Remember

History	Time course / functional disability / effect upon quality of life Past medical history, including infections & toxin exposure Musculoskeletal symptoms (e.g. frozen shoulder with early PD) Drug history – past & previous & recreational (need to contact GP?) Alcohol consumption & responsiveness Family history (with pedigree drawn out if necessary) Neuropsychiatric features (plus carer to inform/corroborate) Autonomic symptoms Sleep problems (REM sleep behavioural disorder suggests PD, DLB or MSA)
Examination	Observation during history of (involuntary) movements including excessive sighing (?atypical parkinsonism) Cognitive assessment (subcorticofrontal vs cortical problems? – MMSE often insensitive to former, consider verbal fluency test, Luria, go/no-go task) Cardiovascular – lying & standing blood pressure, cool periphery (MSA?) Gait, postural reflexes (pull test) & axia tone Eye movements (especially saccadic speed & latency) & blink frequency Limb examination (include specimen of writing & observe posture) • tremors/dystonic posturing (including postural & action) • tone – use reinforcement if in doubt • power & co-ordination • fine finger and rapid alternating movements Reflexes / plantars / primitive reflexes

Table 3 Ten Useful Diagnostic Pointers

Symptom or Sign	Underlying Conditions to Consider
Tremor of onset over 50	'Tremor-dominant' Parkinson's disease (or possibly dystonic tremor) > essential tremor
Able to carry two cups of tea/pints of beer but unable to do up buttons?	'Yes': Parkinson's disease > essential tremor 'No': Essential tremor > Parkinson's disease
Excessive sighing	Atypical parkinsonism (MSA or PSP)
Cold, dusky blue hands	MSA
Sudden, brief dystonic or choreic movements, often unilateral, when patient moves quickly	Paroxysmal kinesogenic dyskinesia
Male>>female with early onset motor and / or sensory tics (voluntarily suppressible); 'magic number', obsessive-compulsive tendencies	Gilles de la Tourette syndrome
Clicking sound in the ear (look in the mouth!)	Essential palatal tremor (tremor of tensor veli palatine connecting with Eustachian tube)
Jerks precipitated by tapping the "snout" area of the face	Reticular reflex myoclonus
Personality disorder, dysarthria, asymmetric tremor (tongue tremor?) in late teens-early 20's	Wilson's disease
Inconsistent or incongruous movements, with non-anatomical sensory loss & 'giving way'	Psychogenic movement disorder

> = more likely than; MSA = multiple system atrophy; PSP = progressive supranuclear palsy

Ancillary studies

An increasing range of blood and cerebrospinal fluid analyses, genetic tests, electrophysiological, structural and functional imaging studies exist to supplement clinical acumen. Occasionally, tissue biopsy (including skin, muscle, small bowel, bone marrow aspirate) may even be necessary. These will be dealt with more fully by other articles in this management series. Suffice it to say that establishing the correct phenomenology of the movement disorder is essential as to which 'line' of more complex investigations is initiated.

There should, however, be a low threshold to undertaking serum caeruloplasmin estimation, since Wilson's disease may present with tremors, dystonia or parkinsonism and is eminently treatable. At a cut-off of 0.2g/l, serum caeruloplasmin is a cheap and simple test, although not very sensitive as 5-20% of homozygous carriers will have normal results. Thus, while an abnormal result should prompt further screening (ophthalmological assessment and urinary copper excretion minimum) a normal serum caeruloplasmin level does not fully exclude Wilson's disease.

Management considerations

Some key general points I try to remember in clinic:

- Treat disability or poor quality of life, *not* recorded impairments
- Remove potentially exacerbating/causative drugs whenever possible
- Always consider underlying depression when there seems to be a marked mis-match between impairment and reported disability
- Patients don't always volunteer neuropsychiatric features like visual hallucinations; ask!
- Members of the multidisciplinary team generally prefer an early referral
- Never forget the need for genetic counselling and implications for other family members (Gasser 2003)
- If a psychogenic movement disorder is suspected, the patient will be best managed by a formal admission and a staged, multidisciplinary approach

Further Reading

Gasser T, Bressman S, Dürr A *et al. State of the art review: Molecular diagnosis of inherited movement disorders.* Mov Disord 2003;18: 3-18.

Kishore A, Calne DB. *Approach to the patient with a movement disorder and overview of movement disorders.* In: Movement Disorders: Neurologic Principles and Practice. Eds, Watts RL, Koller WC. McGraw Hill (New York). 1997: Chapter 1; 3-14.

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