PAPER

J R Coll Physicians Edinb 2006; **36**:331–335
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Movement disorders: a brief practical approach to diagnosis and management

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ABSTRACT The characteristic feature of all movement disorders is an abnormality of the form and velocity of movements of the body. The term 'movement disorder' has become synonymous with basal ganglia disease and extrapyramidal features. Although many movement disorders do arise from pathology within the basal ganglia, disorders such as myoclonus may also arise from other structures. Abnormality of movement may be the only manifestation of a disease process, or may be part of a more widespread neurological disorder. 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; DIP caused by amiodarone). Basal ganglia disease is commonly associated with neuropsychiatric symptoms and these may have a greater impact upon the patient and their family than the movement disorder itself.

An essential first step in the approach to the patient with a movement disorder is to correctly determine the phenomenology of the problem (for example, is the dominant problem chorea or dystonia?). Once this first step has been made, appropriately targeted investigations may then be required to determine the diagnosis. Thereafter, consideration is given to treatment, based upon clinical and social factors, as well as patient preference. This overview will describe a practical approach to the patient with a movement disorder. It will also briefly consider some broad principles in the investigation and management of such cases.

KEYWORDS Chorea, dystonia, examination, movement disorder, parkinsonism, tremor

LIST OF ABBREVIATIONS Dopamine receptor blocking agent (DRBA), drug-induced parkinsonism (DIP), magnetic resonance imaging (MRI), mini-mental state examination (MMSE), multiple system atrophy (MSA), Parkinson's disease (PD), progressive supranuclear palsy (PSP), rapid eye movement (REM), restless legs syndrome (RLS), single-photon emission computed tomography (SPECT)

DECLARATION OF INTERESTS No conflict of interests declared.

Published online **June 2006**

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CLASSIFICATION AND DEFINITIONS

The key to success in diagnosing and managing a patient presenting with a disorder of movement is to establish the phenomenology of the problem. Although the broad definition of patients into those who move too much (hyperkinetic disorder) or too little (hypokinetic or akinetic-rigid disorder) is relatively straightforward, differentiating jerky dystonia from tremor, or tics from chorea or myoclonus, for example, may not be a simple task to the inexperienced physician. To make matters more complicated, the movement disorder may sometimes be 'mixed' (for example, myoclonic dystonia or dystonic tremor). 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; such movements can be more economically classified as dystonic or choreo-dystonic. An exception is 'athetoid cerebral palsy', which remains in common use. There have been no good studies for inter-rater reliability of specific classes of movement disorders, so the final classification remains somewhat subjective.

FEATURES OF CLINICAL HISTORY AND EXAMINATION

The value of a careful history and examination can never be over stated when approaching a patient with a movement disorder, even if the diagnosis may seem obvious from the outset. Age of onset is often helpful in terms of broad classification. Thus, complex tic disorder (Tourette's syndrome) typically begins in the first decade and is seven times more common in boys. Generalised dystonia is also more likely to commence in the first two decades, while focal dystonias classically present in later life. Older age is the greatest risk factor for PD, although young onset cases (defined as less than 40 years of age) are well recognised and are more likely to have a genetic basis.

TABLE 1 Definition of commonly encountered movement disorders.

Movement disorder	Definition
Parkinsonism	A clinical syndrome with bradykinesia (slow movement) as the defining feature, almost always accompanied by rigidity, and often by tremor.
Dyskinesia	May be applied to any involuntary movement, although commonly used to refer to drug-induced chorea and dystonia.
Tremor	A rhythmical, involuntary oscillatory movement of a body part; subdivided into whether the problem occurs at rest, with posture, on action, or with intention.
Chorea	A quick, irregular, semi-purposive, and predominantly distal involuntary movement (patient may look restless or 'fidgety').
Dystonia	An abnormal movement characterised by sustained muscle contraction, frequently causing twisting and repetitive movements or abnormal postures.
Ballism	A proximal, high-amplitude movement, often violent and flinging in nature; usually unilateral and may resolve 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 fashion at the expense of other activity (e.g. hand wringing, clapping, mouthing).

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A video recording, made by the patient's family, may be helpful in the case of a paroxysmal movement disorder. If this is not volunteered, it is a good idea to ask for one, particularly when the examination is negative. If no problem is apparent after a 'routine' neurological examination, consider whether the complaint may be task specific (e.g. certain forms of dystonia, primary writing tremor). This could be the perfect excuse to get the golf clubs out in clinic (the 'yips' as a focal dystonia when attempting to putt the ball) or even the darts ('dartitis' in darts players who have difficulty in releasing the dart).

Always consider drugs, both past and present, as a potential cause for the movement disorder. Tardive dyskinesias (commonly stereotypic movements, often orofacial in distribution) may develop after a relatively short exposure to a DRBA (e.g. chlorpromazine, haloperidol, prochlorperazine) but can persist for years thereafter. A complete list of medications previously taken by the patient should be obtained from the general practitioner, if necessary. Dopamine receptor blocking agents may also cause a range of other movement disorders, including parkinsonism and dystonia. Approximately 80% of DIP will resolve within eight weeks of discontinuing the offending agent, although recovery times of up to 18 months have been reported. Drug-induced parkinsonism may be impossible to differentiate from PD, although it tends to be more symmetric, and is more common in older women. While DRBAs are well known to cause parkinsonism, a link with agents like sodium valproate, amiodarone, and cinnarizine is less well recognised; if in doubt, it is worth checking with the hospital drug information service.

Analysis of the following characteristics (adapted from reference 4) may assist in making the diagnosis:

1 *Specific distribution* For example, RLS (although this is now known as restless limb syndrome since symptoms may also be reported in the upper limbs) and 'painful legs and moving toes'. Parkinson's disease is typically asymmetric in onset. Blepharospasm (involuntary, prolonged eye closure) affects both eyes whereas hemifacial spasm only affects one side of the face.
2 *Specific actions and relationship to voluntary movement* For example, a task-specific tremor or dystonia. Asking the patient to write or pick up a glass of water may be very revealing.
3 *Speed of the movement*

Slow	Intermediate	Fast
Parkinsonism, dystonia, and dystonic tics	chorea, tremor	myoclonus, tics

4 *Rhythm* Continuous (e.g. tremor) or intermittent (e.g. asterixis ('negative myoclonus')).
5 *Relation to posture* For example, orthostatic tremor (presents as unsteadiness when standing still, but suppressed by walking).
6 *Relation to sleep* Few movement disorders persist during sleep; examples that do, include palatal tremor and segmental myoclonus.
7 *Associated sensory symptoms* Restless limb syndrome is integrally associated with pain or discomfort; tics may be associated with a vague discomfort or abnormal sensation in the prodrome before the movement.
8 *Suppressibility* Volitional in tics (although associated with increasing unease and rebound worsening upon release), by sensory 'tricks' in dystonia (such as a light touch upon the opposite side of the face to suppress a spasmodic torticollis).
9 *Aggravating or precipitating factors* Stress and anxiety have no discriminatory value as they may worsen all movement disorders. Myoclonus may be triggered by

J R Coll Physicians Edinb 2006; **36**:331–335
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TABLE 2 Points to remember in the history and examination.

History	Time course/functional disability/effect upon quality of life.
	Past medical history, including infections (e.g. rheumatic fever) and toxin exposure.
	Drug history – current, previous, and recreational (may need to contact family doctor).
	Alcohol responsiveness.
	Family history (draw out pedigree if necessary).
	Neuropsychiatric features (with carer to inform/corroborate).
	Autonomic symptoms (may be prominent and early in MSA, a degenerative form of parkinsonism).
	Sleep problems (REM sleep behaviour disorder – screaming, combative outbursts later in a night's sleep – may occur early in PD, MSA, and dementia with Lewy bodies).
Examination	Observe casually during history: • Any involuntary movements and their distribution; • Utterances and vocalisations (Tourette's syndrome?); • Blink frequency (reduced in parkinsonism, profoundly so in PSP, increased in blepharospasm); • Excessive sighing (suggestive of atypical parkinsonism like MSA and PSP).
	Cognitive assessment (subcorticofrontal vs cortical problems) – MMSE often insensitive to the former; consider supplementing with verbal fluency task, e.g. number of words beginning with letter 'C' in a minute.
	Cardiovascular – lying and standing blood pressures, cool dusky blue periphery (MSA?).
	Gait (stance width, stride length, turning, dystonic posturing of limbs, arm swing), postural reflexes (pull test, standing behind patient) and axial tone (turn patient from side to side in vertical axis using shoulders).
	Eye movements (especially speed of fast eye movements and range).
	Limb examination (including specimen of writing and observe hand posture).
	Tremors/dystonic posturing.
	Tone – use reinforcement if necessary.
	Power and co-ordination.
	Fine finger and rapid alternating movements.
	Reflexes/plantars (areflexia in neuropathic tremor).

specific stimuli such as sudden, loud noise or touch. Carbohydrate-heavy meals and fatigue may precipitate certain forms of paroxysmal dystonia, while sudden movement may induce paroxysmal kinesogenic dsytonia.

10 *Ameliorating factors* Alcohol may dramatically improve essential tremor and myoclonic dystonia. Running or walking backwards may improve a dystonic gait, leading the unwary to suspect a non-organic cause.

11 *Distractibility and inconsistency* Both are suggestive of a non-organic (functional) cause, but note the caveat in point 10 above.

Table 2 summarises a number of points to remember in the history-taking and examination of the patient with a movement disorder. The need to include brief assessments of neuropsychiatric and sleep status should be emphasised. Parkinson's disease may be accompanied in 20–30% of cases by significant depression, which may be the major determinant of quality of life, while up to 80% of people with PD may eventually develop dementia. Tourette's syndrome may be associated with attention deficit hyperactivity disorder and obsessive-compulsive disorder. Huntington's disease may be complicated by anxiety and panic attacks, depression, and schizophreniform psychosis, and is associated with an increased risk of suicide, in addition to the well-known association with dementia. Rapid eye movement sleep behaviour disorder may predate the onset of PD, multiple system atrophy, and dementia with Lewy bodies. It may lead to violent motor outbursts during sleep and self-injurious behaviour (as the person flings themselves out of bed while asleep) or injury to the bed partner, yet is readily treated by low-dose clonazepam. Sleep fragmentation is common in PD and is multifactorial in aetiology (causes include nocturia, depression, and discomfort due to under-dosing). Many of these causes are amenable to treatment.

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INVESTIGATIONS

An increasing range of blood and cerebrospinal fluid analyses, genetics tests, electrophysiological, structural, and functional imaging studies are available to supplement clinical acumen. Occasionally, tissue biopsy (for example, skin, gut, or bone marrow) may even be necessary. It goes without saying that establishing the correct phenomenology of the patient's movement disorder is an essential first step before embarking upon more complex (and often costly) investigations. Many movement disorders are diagnosed clinically and investigations may play only a supportive or exclusionary role. Thus, patients with typical PD do not require MRI brain scanning. Magnetic resonance imaging scanning may, however, be helpful for patients with 'atypical parkinsonism' or a sub-optimal response to treatment. A few general themes may be summarised:

1 Never overlook the value of 'routine' blood tests: renal, hepatic, and thyroid function tests may yield useful information as to the cause of tremor or myoclonus, for example;
2 Have a low threshold to perform a serum caeruloplasmin level in the young or middle-aged person with a movement disorder: Wilson's disease may present in protean ways and is eminently treatable. At a cut-off of 0·2 g/l, serum caeruloplasmin is a cheap and simple test (although not very sensitive, as 5–20% of homozygous carriers will have normal results);
3 Phenocopies of PD may result from diverse genetic causes, including certain spinocerebellar ataxias and juvenile-onset Huntington's disease;
4 Remember, structural imaging is of limited sensitivity in the diagnosis of most movement disorders. Few, if any, prospective studies have examined the positive predictive value of modalities such as MRI scanning in patients presenting with early 'unclassifiable parkinsonism'; and
5 Always remember the limitations of a test, to avoid over-interpretation of the result. An example is ^{123}I-ioflupane SPECT, also known as DaTSCAN. This tool is helpful in discriminating essential tremor, DIP, or psychogenic parkinsonism (when the scan is normal) from PD (when the scan is abnormal). DaTSCAN cannot, however, differentiate typical from atypical parkinsonism (that is, separating PD from conditions such as MSA and PSP). It may also occasionally be normal in early tremor-dominant PD. The same point can be made in the context of caeruloplasmin 'screening' for Wilson's disease. Given the false-negative rate, if a high clinical index of suspicion exists, further investigations, including a 24-hour urinary copper excretion and ophthalmological assessment for Kayser-Fleischer rings, should be performed.

MANAGEMENT CONSIDERATIONS

Try to remember the following general principles in clinic:

1 Treat disability or poor quality of life, not recorded impairments. Thus, one patient with PD may tolerate moderate to severe tremor, rigidity and/or bradykinesia and not wish to be treated, while another case could request treatment with much less severe motor impairment;
2 Remove potentially exacerbating/causative drugs whenever possible;
3 Always consider underlying (masked) depression when there appears to be a mismatch between impairment and reported disability;
4 Patients do not always volunteer neuropsychiatric features like visual hallucinations or hypersexuality (which may be induced by dopaminergic drugs). Don't be afraid to ask;
5 Members of a multidisciplinary team generally prefer early referral;
6 Never forget the need for genetic counselling and the potential implications for other family members;
7 If a psychogenic movement disorder is suspected, the patient may best be managed by formal admission and a staged, multidisciplinary approach; and
8 Don't be frightened to admit to the patient that you're not sure about the diagnosis. Differentiating tremor-dominant PD from essential tremor, or typical from atypical parkinsonism, for example, can be very difficult. The main thing is not to 'pigeon-hole' the case too early on, at the risk of having to back-track at a later date. Invariably, time will tell.

KEYPOINTS

- Establishing the phenomenology of a movement disorder is essential in the diagnostic pathway.
- Do not forget to take a full family history, particularly in a young onset case, or one with clinically atypical features.
- Dopamine receptor blocking drugs may cause any movement disorder and their adverse effects may persist after the offending agent has been discontinued.
- Many movement disorders are diagnosed clinically and investigations may play only a supportive or exclusionary role.
- Neuropsychiatric and cognitive problems are common in movement disorders and may be dominant in determining the patient's (and carer's) quality of life.

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J R Coll Physicians Edinb 2006; **36**:331–335
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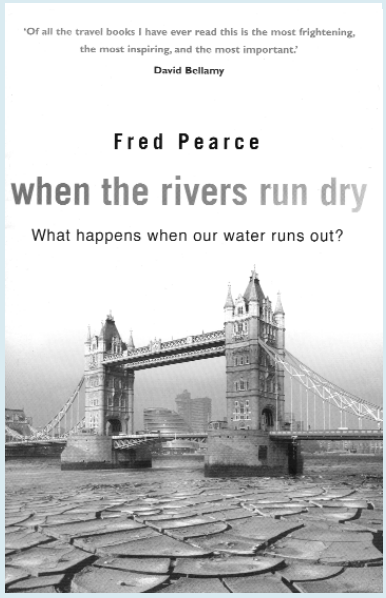
A BOOK YOU SHOULD READ

When the rivers run dry

F Pearce
ISBN 1903919576
Eden Project Books, Transworld 2006
£18.99



The threat of global warming to human life on our planet is changing our world view. Politicians vie with one another over their green credentials in relation to global warming, but much less is heard about world water problems. Fred Pearce has written about the environment for some 15 years, and in this book he explores our mistaken, profligate, and often irresponsible, use of water and its consequences, and points to some ways of avoiding future disasters.

World demand for water exceeds supply from the world's rivers; a billion people have no access to a safe and reliable water source; two-thirds of water consumed by humans goes for agriculture. Rivers have been dammed and their water removed by canals so that silt distribution has been impaired, down-river land damaged by excess salt, and outflow to the sea grossly reduced. Drainage of related wetlands has reduced the effectiveness of dams in preventing flooding. Inexpensive drilling and pumping equipment has led to serious depletion of irreplaceable underground aquifers, and desalination/reverse osmosis plants have proved expensive.

The future, however, is not hopeless, though salvation means facing human desperation, selfishness, greed, special interests, and nationalism. Engineers and hydrologists can now manage rivers better, the loss of a third to a half of water supply by leakage could be stopped, rainwater could be conserved, more sewage could be recycled, agriculture could avoid growing high-water-demand crops in water-short areas and could develop fewer water-demanding crops and systems for better water use.

Pearce tells a fascinating story well, and I believe doctors will want to know more about something so central to future human health.

Niall Finlayson,
Director of Communications, RCPE

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