Movement disorders

Movement disorders are neurological conditions that affect the speed, fluency, quality, and ease of movement. This article reviews some of the most common movement disorders seen in elderly patients.

Excessive movement

Dystonia
This is characterised by spasms or abnormal muscle contraction affecting one or more body parts.\(^1,2\) Dystonia can be primary in origin or secondary to other factors. Types and classification may be according to age of onset, distribution of affected body parts or aetiology.

Blepharospasm is dystonia affecting the eyelid muscles. It is more common in females. Spasms may produce episodes of blindness.\(^3,4\) Writer’s cramp is a common cause of forearm stiffness resulting in difficulty writing.\(^3\) To manage this condition patients usually learn to write with their non-dominant hand.

Cervical dystonia or spasmodic torticollis is the commonest dystonia, with average onset in middle age. Abnormal posturing of the head, neck and shoulders is apparent, but may spontaneously resolve. It has a significant effect on quality of life.\(^2,5\)

Secondary dystonia could arise from the use of neuroleptic medication and be associated with stroke, space occupying lesion and Wilson’s disease, to mention a few.\(^2,3\) Hereditary dystonia usually present in childhood and progress with time to become more generalised.\(^2\)

Unfortunately many types of dystonia are unresponsive to medication. Some can also be treated by botulinum toxin injection or by surgical resection of individual nerves.\(^3\) Deep brain Stimulation (DBS) is used for more generalised and severe forms of hereditary torsion dystonia, which can be quite disabling.\(^6\)

Dyskinesia
This is generalised excessive involuntary movements of body parts.\(^1\) It can be induced by drugs causing increased dopamine activity, namely levodopa, dopamine agonists or catechol-O-methyl transferase (COMT) inhibitors. It may occur in relation to the timing of medication.

Tardive dyskinesia
This is the involuntary repetitive movements of the face and tongue, grimacing, lip smacking, pursing of the lips and rapid eye blinking.\(^1\) This condition is associated with prolonged neuroleptic medication use and is more likely to affect older patients. It may also be associated with involuntary upper and lower limb movements. Treatment is centred on withdrawal of medication and substitution with another agent; however this is often difficult in clinical practice. Atypical antipsychotics are associated with reduced extrapyramidal side effects.\(^7\)

Akathisia
A subjective inner restlessness, which is evident by the inability to sit still.\(^1\) It can be caused by prolonged anti-psychotic medication, as well as cocaine and heroin withdrawal. Restless leg syndrome is common, occurring mostly at night. Dopamine agonists can be beneficial.\(^5,3,8\)

Essential tremor
Essential tremor (ET) has a frequency of 7–8Hz and is therefore finer than the tremor of Parkinson’s disease (PD). It affects 2–3% of the
population. Its aetiology is unknown. It is typically symmetrical and evident on an outstretched hand. Family history may be present. It is characteristically dampened by alcohol and exacerbated by anxiety. It is more common with increased age and can lead to spillage when drinking from a cup. In some cases differentiation between ET and Parkinsonian tremor can be challenging. The role of Dopamine Transporter (DAT) Scan as a tool to assess the basal ganglia function has proved extremely useful.

Treatment is with lipid soluble beta-blockers, eg. propranolol or primidone. Other causes of tremor include cerebellar dysfunction, which is associated with dysmetria, dysdiadochokinesia, ataxic gait, nystagmus, intention tremor, scanning dysarthria and hypotonia.

**Tics**

Tics commonly occur in children, in the form of sniffing or twitching of the face, however these are generally short-lived and typically abate or disappear by adulthood. In its severest form, Gilles de la Tourette Syndrome is characterised by multiple tics and behavioural disturbances, including attention deficit hyperactivity, shouting of sexual obscenities and obsessive-compulsive disorder. It is more common in males, presenting in the teenage years and persists. Treatment is with haloperidol and behavioural therapy.

**Myoclonus**

Myoclonus is a sudden, involuntary jerking of a group of muscles that may be provoked by sudden stimuli. Benign essential myoclonus occurs when falling asleep and is benign. Paramyoclonus multiplex is the jerking of the body, usually occurring in adolescence and is not associated with epilepsy, which disappears in adulthood. Myoclonic jerks are common and of little consequence in
neurodegenerative disorders, eg. Alzheimer’s disease and other dementia disorders.

Myoclonus itself is associated with a number of types of epilepsy: static non-progressive myoclonus following cerebral anoxia, progressive myoclonic epilepsy with progressive encephalopathy and the more frequent juvenile myoclonic epilepsy.\(^3,11,12\)

Chorea

Chorea is jerky or explosive involuntary movement particularly affecting the proximal body parts, eg. head, face or limbs.\(^1\) It can occur following an acute neurovascular event causing unilateral chorea (hemiballismus), this typically coincides with contralateral basal ganglia pathology.

Acute chorea may be seen with small vessel disease in polycythaemia rubra vera, systemic lupus erythematosus (SLE), thyrotoxicosis and secondary to phenytoin or alcohol. Cerebrovascular disease, head trauma and space occupying lesions are also associated with chorea. Short courses of treatment with tetrabenazine under medical supervision may be helpful.\(^3\)

Sydenham’s chorea

This occurs following infection, usually streptococcal or rheumatic fever; it is more common in young people, but tends to be self-limiting. There is gradual onset of chorea over weeks. Clinical features of rheumatic fever are usually not apparent, anti-streptococcal titres and inflammatory markers can be normal. Sedation may be necessary. A high prevalence of antibiotic use has reduced the incidence of these complications.\(^3\) Chorea can also occur following infection, eg. Legionnaire’s disease etc, the so called “dancing fevers.”\(^7,13\)

Huntington’s disease

This condition presents with progressive motor, psychiatric and cognitive disorders. Chorea is the main motor symptom, dystonia, bradykinesia, rigidity and loss of postural reflexes may be apparent. Cognitive function declines with disease progression, with associated short-term memory impairment, impulsivity and distractibility.\(^14,15,16\)

This is an autosomal dominant condition with full penetrance. There is a mutation in the distal arm of chromosome 4, resulting in a defect in the protein huntingtin. This leads to excessive trinucleotide repeat sequence with “genetic anticipation phenomenon.”\(^3\) Offspring have a 50% likelihood of developing Huntington’s disease. The average age of onset is 40 years, however there are recognised juvenile and older onset varieties of the disease.\(^14,15,16\)

Anxiety, depression and obsessive-compulsive disorders are common in patients with Huntington’s disease. Communication difficulties and dysphagia may occur. Despite the motor manifestations of Huntington’s disease being most apparent, non-motor symptoms often cause significant distress and functional disability.\(^3,16\)

Neuroimaging demonstrates progressive cerebral atrophy. Referral to specialist service is recommended for diagnosis and a multidisciplinary approach is important. There is no treatment to prevent disease progression. Death occurs approximately 10–20 years from time of diagnosis.\(^3,16\)

Drug induced Parkinsonism with phenothiazines offers symptomatic relief. Genetic analysis of family members can be offered. There are nationally agreed counselling protocols. Implications of positive test will have long-term consequences on employment, emotional and reproductive choices, given the lack of curative treatment.\(^3\)

Wilson’s disease

Wilson’s disease is an autosomal recessive condition where copper is deposited throughout the body due to defective copper metabolism; in particular, the liver, basal ganglia and cornea, in the form of Kayser Fleisher rings. Children typically present with liver dysfunction; older patients present with neurological disturbance: tremor, dysarthria, dementia and involuntary choreoathetoid movements.\(^11\)

Copper and caeruloplasmin may be normal or reduced, urinary copper increased, quantification of copper in the liver at biopsy confirms diagnosis. Life-long treatment with penicillamine is effective, however neurological damage is permanent.\(^3\)

Spino-cerebellar ataxia

Disorders of the cerebellum typically produce scanning dysarthria, ataxic gait, hypotonia, nystagmus, intention tremor with dysmetria and dysdiadokinesis. Gait disturbance is a common presenting complaint. A full history incorporating alcohol intake, medication history and use of recreational drugs should be taken. Family history may suggest an inherited condition, eg. Fredreich’s ataxia. Signs and symptoms of
raised intracranial pressure should be excluded. Causes of primary cerebellar pathology include stroke, intracranial haemorrhage, neoplasm and abscess formation. Other infective associations include chickenpox and Legionnaire’s disease. Hypothyroidism is also associated with cerebellar signs. 3,11,17

Hydrocephalus may also produce gradual onset cerebellar features with urinary and memory symptoms. Steadily progressive ataxia occurs secondary to alcohol abuse. Paraneoplastic cerebellar degeneration has been well described and associated with ovarian or lung malignancy. 3

Akinetic rigid syndromes

Parkinson’s disease

Idiopathic Parkinson’s disease (PD) is progressive and due to a deficiency of the neurotransmitter dopamine in the substantia nigra of the basal ganglia, with Lewy body deposition. This deficiency accounts for the characteristic clinical features of PD: tremor, rigidity, hypo/bradykinesia and a postural instability. Hypokinesia is a delay in the initiation of movement, whilst bradykinesia describes slowness in the execution of movement. 1,18

The prevalence of PD is 0.2%, it occurs more frequently in men with the average age being in 60s. Young and juvenile onset PD is less common. 19 Prevalence is likely to increase directly in association with the ageing population. The cause of PD is unknown; however associations with environmental toxins have been identified. Genetic mutations are associated with 5% of cases. 3,18,20

Joint pain, stiffness, tremor or difficulty initiating fine movements may occur in the early stages. Olfactory dysfunction is recognised in the pre-clinical phase. Micrographia, postural change or falls may also be the presenting complaint.

A wide spectrum of non-motor conditions are associated with PD, psychiatric problems, such as anxiety, apathy, depression, psychotic behaviour, dementia and sleep disturbance, as well as autonomic disturbance producing urinary dysfunction, constipation, sexual dysfunction, postural hypotension, weight loss, dysphagia, hyperhidrosis and salorrhoea. Joint and neuropathic pain may also occur. 3,18,24

PD tremor is typically 4-7Hz, worse at rest, initially asymmetrical, but becoming symmetrical with disease progression. It typically disappears during sleep, is described as “pill rolling” with characteristic thumb shaking. It may be exaggerated at initiation but dampens during the execution of movement. “Lead-pipe” rigidity develops and “cog wheeling” may be evident. Increased tone, brisk or normal reflexes, with flexor plantar responses are typical on neurological examination.

The posture in PD is generally a flexed one affecting all joints (neck, spine, hips, knees, elbows and wrists). Gait is shuffling (festinant) with associated loss of arm swing on the affected side. Cognitive function and sensation are normal in early onset disease. Speech and swallowing disturbances may occur with varying severity. 3,20,21

PD is a clinical diagnosis, often aided by the use of the United Kingdom PD Society’s (UKPDS) Brain Bank diagnostic criteria. It may be difficult to distinguish PD tremor from ET in the absence of typical clinical features of PD. 3,10,20 DAT scan can be used when distinction between essential tremor and PD is unclear during the early stages of the disease. A positive DAT scan finding also occurs in other PD related disorders, eg. Lewy body dementia, multisystem atrophy and progressive supranuclear palsy. 22

NICE recommend referral of a suspected PD case to specialist. Diagnosis in primary care is associated with a higher rate of error than by movement disorder specialists. 23 Treatment initiation in primary care is not recommended as this may mask symptoms. 10

Management

A multi-disciplinary approach is essential to maintain function and quality of life. 10 Treatment aims to reduce symptoms, by means of increasing dopamine level in the brain, either by reducing metabolism and or stimulating dopamine receptors.

Treatment with levodopa (dopamine precursor) with peripheral decarboxylase inhibition (to prevent systemic side-effects) is the mainstay of treatment (Madopar/Sinemet). Initial treatment with levodopa typically produces good symptom improvement. Side effects, poor or unpredictable response can occur with long-term usage. 1,21

“End of dose” akinesia and “freezing” may occur, highlighting the importance of regular medication intervals. “On and off” phenomenon occurs when patients alternate between freezing
and dyskinesia with long-term use. Confusion, visual hallucinations, hypotension and gastrointestinal upset are some of the commonly encountered side effects of levodopa.

Dopamine agonists, such as ropinirole, pramipexole and rotigotine are used to stimulate dopamine receptors and are useful in delaying treatment with levodopa or can be prescribed concurrently. Apomorphine is a subcutaneous preparation of a dopamine agonist that is used in patients with significant symptom fluctuation.

Amantadine is prescribed to reduce dyskinesia. Monoamine Oxidase Type B Inhibitors (MAO-B) reduce the metabolism of dopamine. It can be used in the early stages of PD to delay the need for levodopa or in addition to levodopa. Cathecol O-Methyl T Inhibitors (COMT), eg. entacapone, slows levodopa metabolism and may increase levodopa availability and its duration of action. Patients should be warned about dopamine dysregulation syndrome (impulsiveness and excessive daytime sleepiness). Sudden withdrawal of levodopa can cause neuroleptic malignant syndrome. Regular specialist review is recommended.

Surgical intervention with bilateral subthalamic or thalamic stimulation is used in patients with motor complications demonstrating poor response to medical treatment in specialist units after careful medical and psychological evaluation. Vascular PD patients are more likely to present with gait disturbance, rather than tremor. The lower limbs are affected more severely than the upper, with small steps rather than festinant gait. They may have other features with hypo(brady)kinesia and tendency to falls. Reflexes may be brisk with other pyramidal tract signs. They are less responsive to treatment with levodopa. Presence of vascular risk factors and or neuroimaging findings of territorial stroke may prove useful in diagnosis.

Drug-induced parkinsonism

Medications such as anti-emetics (eg. metoclopramide), labyrinthine sedatives (prochlorperazines) and neuroleptics (less with atypical anti-psychotics) can induce Parkinsonian symptoms, as well as toxins including manganese and pesticides. There is limited levodopa response and symptoms improve with discontinuation/withdrawal.

Parkinson's plus syndromes

These are conditions wherein Parkinsonism exists with additional clinical features and pathology.

**Progressive Supranuclear Palsy (Steel Richardson's syndrome)**

This is a tauopathy (neurofibrillary tangles depositing) with a male preponderance. Initially there is limitation of vertical (superior and inferior), then later lateral conjugate gaze. There is loss of rapid lateral saccadic eye movements that are initially broken and later slow or absent. There is clumsiness, axial rigidity, falls, dysarthria and it is associated with dysphagia and recurrent aspiration pneumonia, as well as cognitive impairment in the later stages. Response to levodopa is poor.

**Multiple System Atrophy**

This is due to selective atrophy of a number of areas (olive, pons, cerebellum and or sympathetic chain) with oligodendroglial cytoplasmic inclusion bodies as its pathological hallmark. Patients may present with parkinsonian features and a mixture of dysarthria, sialorrhoea (often worsened by levodopa), broad based gait (unlike shuffling parkinsonism), pyramidal tract as well as cerebellar signs depending on the affected or involved areas. Tremor is often absent. Postural hypotension is common, aggravated by levodopa treatment. When cerebellar and pyramidal tract signs predominate, it is often referred to as olivopontine cerebellar degeneration.

**Shy-Drager’s Syndrome**

This is Parkinsonism with autonomic features and secondary marked postural hypotension. Response to levodopa is poor.

**Corticobasal degeneration**

Parkinsonian features accompany unilateral loss of dexterity with poor co-ordination, akinesia, rigidity and limb dystonia/apraxia which progress to become bilateral. “Alien hand phenomenon” is a classical presenting feature. Gait and speech disturbance, myoclonus, plus cognitive impairment occur commonly leading to dysphagia and total immobility, death occurs six to eight months from onset. No effective treatment is available.

**Psychogenic movement disorders**

These commonly present with tremor or dystonia as conversion or somatisation disorder, often
associated with underlying physical or mental illness or psychological stress. Onset is often sudden with rapid progression with or without periods of remission. History of previous medically unexplained symptoms or frequent primary care attendances without underlying neurological abnormality is typical. Reassurance or cognitive behavioural therapy can be helpful.1,3

Conclusion

In this article we summarise some of the most commonly encountered movement disorders in primary and secondary care. A significant proportion are due to chronic neurodegenerative disorders that may require long-term surveillance and follow up. It is highly advisable that patients with such disorders should be referred to specialists with similar interest for correct diagnosis, prognosis and effective treatment.

Conflict of interest: none declared

References