Dyskinesia (Abnormal involuntary movements)

What is dyskinesia?

Abnormal involuntary movements (AIMs) are known as 'dyskinesias'. There are several varieties of dyskinesia which have different clinical appearances, underlying causes and treatments. Tremor, chorea, dystonia and myoclonus are examples of types of dyskinesia which have different mechanisms and modalities of treatment.

Tics and stereotypies may also be considered to be related but some experts call these 'unvoluntary' because there is an element of voluntary control.

Cerebrovascular diseases are a common cause of secondary movement disorders. Post-stroke movement disorders include Parkinsonism and a wide range of hyperkinetic movement disorders, including chorea, ballism, athetosis, dystonia, tremor, myoclonus, stereotypies and akathisia.

Classification

Athetosis

Sinuous, slow, involuntary writhing movements affecting the fingers, hands, toes and feet. The arms, legs, neck and tongue may also be affected. Causes include asphyxia, neonatal jaundice, Huntington's chorea, cerebrovascular disease and trauma. Management can be difficult but treatment options include medications (eg, diazepam, haloperidol, tetrabenazine), surgery and retraining techniques.

Chorea [1]

Continuous jerky movements in which each movement is sudden and the resulting posture is held for a few seconds. This usually affects the head, face or limbs. The focus may move from one part of the body to another at random.

Chorea may be caused by adverse effects of drug treatments, especially medications for Parkinson's disease, epilepsy and schizophrenia. Other causes of chorea include the following:

Inherited

- Huntington's chorea - autosomal dominant inheritance, usually presents in middle age with chorea and dementia. Insidious onset with motor, cognitive and psychiatric abnormalities. There is no treatment.
- Benign hereditary chorea.
- Wilson's disease.

Acquired

- Sydenham's chorea - also known as St Vitus' dance. Mainly associated with acute rheumatic fever. It is now rare. It usually presents in children aged 7-12 years, initially with psychological symptoms of behavioural disturbance followed by generalised chorea; it usually recovers in 1-3 months. Penicillin for rheumatic fever, and diazepam, haloperidol or tetrabenazine for chorea.
- Other immune-mediated choreas - eg, in association with systemic lupus erythematosus (SLE), antiphospholipid antibody syndrome and vasculitis (eg, polyarteritis nodosa, Behçet's disease, multiple sclerosis, CNS angiitis).
- Infectious chorea - eg, meningitis, encephalitis, Lyme disease, HIV-AIDS new variant Creutzfeldt-Jakob disease and subacute bacterial endocarditis.
- Vascular chorea - eg, stroke, polycythaemia and Moyamoya disease.
- Hormonal disorders - eg, hyperthyroidism, hypoparathyroidism with hypocalcaemia.

- Pregnancy (chorea gravidarum), oral contraceptives and hormone replacement therapy.
- Metabolic: electrolyte and biochemical disturbance - eg, hypernatraemia and hyponatraemia, hyperglycaemia and hypoglycaemia, hypomagnesaemia, hypocalcaemia, hepatic and renal failure.
- Vitamin deficiency: B1 and B12.
- Paraneoplastic syndromes - eg, small cell carcinoma of the lung, renal cell carcinoma, ovarian cancer and lymphoma.
- Postoperative - following childhood cardiac surgery ('post-pump choreoathetosis').
- Other CNS conditions: trauma (including cerebral palsy), intracranial tumours.
- Senile chorea.
- Ventriculoperitoneal shunts.
- Toxins - eg, carbon monoxide, cyanide, alcohol, methanol, solvents, thallium, mercury and manganese.

Chorea may occur with athetosis and is then called choreoathetosis.

Dystonias

A dystonia is a sustained muscle contraction, frequently causing repetitive twisting movements or abnormal postures [2] . It is a dynamic condition that often changes in severity, depending on the posture assumed and on voluntary activity of the area of the body involved. The diagnosis is clinical and there are no specific tests available; therefore, expert opinion should be sought. Dystonias may be primary or secondary [3] .

Treatments available for dystonia include oral medications, botulinum toxin and surgical procedures. Oral medications are generally used for generalised and segmental dystonia. Botulinum toxin is the mainstay of treatment for focal dystonia. Surgical procedures are available for medication-refractory dystonia, markedly affecting quality of life [4] .

Primary pure dystonia is inherited in a mainly autosomal dominant pattern [5] . Two genes have been identified: DYT1 and DYT6. It usually presents in children, with dystonic spasms of the legs on walking and occasionally of the arms, trunk or neck. It is normally progressive and spreading to the whole body, causing severe disability within about ten years. A levodopa trial should be offered to patients with early-onset dystonia without an alternative diagnosis [6] . Blepharospasm and writer's cramp are both focal dystonias. Blepharospasm involves recurrent spasms of eye closure. Writer's cramp is the inability to write or use any manual instrument, due to abnormal posture of the hand and arm [7] . Botulinum toxin injection is the first-line treatment for most types of focal dystonia [8] . Hemidystonia involves half of the body and is usually secondary to a structural lesion in the contralateral basal ganglia.

Dystonias may be secondary to a neurological condition such as a focal brain lesion, exposure to drugs or chemicals, or as part of Parkinson's disease. Infantile cerebral palsy is the most frequent cause of acquired dystonias in children. Many children also have other symptoms and signs in addition to dystonia, particularly spasticity, cognitive impairment, or epilepsy, thus qualifying to be grouped into the category of complex dystonias [9] . Intrathecal baclofen can be indicated in patients where secondary dystonia is combined with spasticity.

Deep brain stimulation (long-term electrical stimulation of the globus pallidus internus) is an effective treatment for primary generalised or segmental forms and for those patients who do not achieve sufficient relief with a more conservative approach [10] [11] . However, deep brain stimulation is less effective for secondary dystonias [12] .

Dystonia may lead to permanent contractions, by causing tendons to shorten.

Hemiballism

These are wild flinging/throwing movements of one arm or leg, usually occurring as a result of a cerebrovascular event. They can vary in intensity from mild to severe and may even cause injury. They usually subside over a period of 3-6 months but can be treated with a phenothiazine, haloperidol or tetrabenazine. They may require neurosurgery to be adequately controlled.

Myoclonus[13]

These are rapid muscle jerks that are frequently repetitive and cause significant disability. They appear as:

- Benign essential myoclonus: affects much of body, repeated as many as 50 times per minute. Presents in childhood or adolescence with mild disability. Helped by alcohol and beta-blockers.
- Progressive myoclonic encephalopathies: appear as part of a range of other neurological disorders.
- Static myoclonic encephalopathies: Lance-Adams syndrome after cerebral anoxia.
- Myoclonic epilepsies: eg, focal myoclonus - restricted to one part of the body (eg, hemifacial spasm, mainly affecting older women).

Myoclonus usually requires a combination of drugs (in large doses)[14] . Antiepileptic drugs (eg, valproate, levetiracetam and piracetam) are effective in cortical myoclonus but less so in others. Clonazepam may be helpful with all types of myoclonus. Botulinum toxin may be useful for segmental myoclonus.

Spasmodic torticollis

Torticollis is a twisting of the head and neck caused by a shortened sternocleidomastoid muscle, tipping the head toward the shortened muscle, while rotating the chin in the opposite direction. Torticollis may occur in all ages, from newborns to adults[15] . See also the separate Neck Pain (Cervicalgia) and Torticollis article.

Tardive dyskinesia

This is characterised by orofacial mouthing with lip-smacking and tongue protrusion, body rocking and distal chorea. In younger patients it may also cause axial and cranial dystonia. It usually occurs following at least six months of treatment with neuroleptics. The risk of new cases is around 5% per year of cumulative drug exposure, with age and early occurring extrapyramidal side-effects being two important risk factors[16] .

- Preventing tardive dyskinesia is the best option, and clinicians should follow best practice for prescribing antipsychotic medication, including limiting the prescription for specific indications, using the minimum effective dose, and minimising the duration of therapy[17] .

- The first-line management of tardive dyskinesia is the withdrawal of antipsychotic medication if clinically feasible. Yet, for many patients with serious mental illness, the discontinuation of antipsychotics is not possible due to disease relapse.
- Switching from a first-generation to a second-generation antipsychotic, such as clozapine or quetiapine, may be effective in reducing tardive dyskinesia symptoms.
- VMAT2 inhibitors (vesicular monoamine transporter 2) are integral membrane proteins that regulate monoamine uptake from the cytoplasm to the synaptic vesicle for storage and release. There are 3 VMAT2 inhibitors: tetrabenazine, valbenazine and deutetrabenazine. These medications have not been approved and may be unavailable for use outside the USA.

Tics [18]

These are repetitive stereotyped movements. The patient can initiate them voluntarily and can also intentionally suppress them for a short time:

- Simple tic - sudden rapid twitch always occurring at the same site. Occurs in a quarter of all children and resolves within a year. May persist into adulthood; rarely treated.
- Complex multiple tics - more extensive and severe. When occurring with the patient speaking, particularly swearing, they may represent Tourette's syndrome. They may also appear as a symptom of encephalitis lethargica and of neuroacanthocytosis; they can also be drug-induced.

Tremor

See also the separate Tremor article.

This is a rhythmic movement of part of the body. Essential tremor and Parkinsonian tremor are the most common forms of tremor [19] . There are three types of pathological tremor:

- Static - occurs in a relaxed limb when fully supported at rest. Causes include Parkinson's disease, Parkinsonism, other extrapyramidal diseases, multiple sclerosis.

- Postural - occurs if a limb is static (can also remain during movement). Types include physiological tremor, exaggerated physiological tremor - eg, in thyrotoxicosis, anxiety states, alcohol abuse, drugs (eg, sympathomimetics, antidepressants, valproate, lithium), heavy metal poisoning ('hatter's shakes' from mercury). Neurological disease - eg, severe cerebellar lesions, Wilson's disease, neurosyphilis, peripheral neuropathies, benign essential (familial) tremor, task-specific tremors (eg, primary writing tremor).
- Kinetic or action tremor - occurs during voluntary active movement of an upper body part. Intention tremor is one that occurs when a tremor worsens as a goal-directed hand movement nears its intended target. Brainstem or cerebellar disease including multiple sclerosis, spinocerebellar degenerations, vascular disease, tumours.

There are also psychogenic tremors.

Tremors and dystonias that are not secondary to Parkinson's disease may be effectively treated with deep brain electrical stimulation[20] . Benign essential tremor is treated with alcohol in moderation. Beta-blockers or primidone are also used.

Levodopa-induced dyskinesia[21]

Approximately 50% of people with Parkinson's disease will experience levodopa-induced dyskinesia (LID) 4-5 years after initiation of levodopa treatment. However, the percentage of those patients experiencing troublesome LIDs requiring intervention is actually much lower than 50%. There are three main forms of LIDs:

- 'Peak-dose' dyskinesias are choreic movements related to high levodopa plasma concentrations.
- Diphasic on/off dyskinesias, which coincide with rising and decreasing plasma concentrations of levodopa and may include both chorea and dystonia.
- 'Off' dystonia, which is an often painful dystonic posture, appears early in the morning or at night and occurs when plasma levels of levodopa are very low.

Assessment of dyskinesia

Many different rating scales have been developed to assess people with dystonia [22] [23] .

The Abnormal Involuntary Movement Scale is used to assess tardive dyskinesias and other AIMs [24] [25] .

Dyskinesia treatment and management

The management of AIMs will depend on the underlying cause. Drug treatments include [26] :

- Tetrabenazine - mainly used to control movement disorders in Huntington's chorea and related disorders. Haloperidol, olanzapine, risperidone and quetiapine can also be used to suppress chorea in Huntington's disease but are all unlicensed.
- Haloperidol - can improve motor tics and symptoms of Tourette's syndrome and related choreas. Other treatments for Tourette's syndrome include pimozide and sulpiride (both unlicensed).
- Trihexyphenidyl in high dosage - can also improve some movement disorders.
- Chlorpromazine and haloperidol - are used to relieve intractable hiccup.
- Propranolol or another beta-adrenoceptor blocking drug - may be used to treat essential tremor or tremors associated with anxiety or thyrotoxicosis.
- Primidone - may provide relief from benign essential tremor.
- Piracetam - is used as an adjunctive treatment for myoclonus of cortical origin.

Further reading

- The Dystonia Society
- The Differential Diagnosis of Chorea (videos); Oxford University Press (USA)

- Cardoso F; Huntington disease and other choreas. Neurol Clin. 2009 Aug;27(3):719-36, vi.

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