Today's Treatment

Diseases of the central nervous system

Involuntary movements

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Some involuntary movements are normal, as is blinking. Some, usually normal, may occasionally be associated with various diseases—for example, hiccup. Abnormal involuntary movements of many types may accompany diseases of the nervous system. Similar movements may be apparent in differing diseases. More than one type of movement may be displayed concurrently or successively during the course of one disease. Precise correlation between patterns of abnormal movements and the nature and sites of neurological lesions is rarely possible. An approach to the classification and treatment of movement disorders based on their pathogenesis is impracticable. They are most usefully considered descriptively, taking account of their frequency.

Tics

Tics are usually rapid, repetitive, co-ordinated, and stereotyped movements. Most can be mimicked, though in the more complex varieties only with difficulty. The distribution and extent of these movements varies widely in different people, but an individual's tic tends to be reproduced faithfully again and again. In most instances tics are not due to any organic disease of the nervous system. They are common; indeed, it has been asserted that every one has a ritualised movement which comforts him during stress. They are more often observed in social circumstances than in the surgery.

Often the movements are localised to small groups of muscles around the eyes or mouth. Sometimes they affect the shoulder girdles and limb muscles. They may initially be a conscious mannerism and later can be suppressed by conscious effort, but in time they become established as involuntary movements.

Common mild tics, including blepharospasm, sniffing, and shrugging movements, may be irritating but rarely need treatment. When well established in an adult they are virtually untreatable.

Many children develop tics, often elaborate facial movements, which alarm parents. These almost always disappear and no treatment is required other than reassurance and occasionally small doses of diazepam.

In adults tics of recent development may be induced by drugs. Amphetamine addiction leads to repeated, rapid chewing movements of the jaws. Complex tongue, mouth, and cheek movements may result from taking phenothiazines or levodopa. Withdrawal of the causal drug usually leads to rapid cessation of movements though occasionally phenothiazines may induce permanent dyskinesias, which may require treatment with agents such as tetrabenazine or thiopropazate as set out below.

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SPASMODIC TORTICOLLIS

Spasmodic torticollis is a form of tic. It may be part of a generalised and severe movement disorder, but it often occurs in isolation. The clinical disorder chiefly comprises involuntary turning of the head to one side. Sometimes backward or forward head movements occur concurrently and may occasionally predominate. This disorder, affecting men and women equally, may start at any age from 15 to 60, but most commonly in the 30s. Initially rapid and correctable rotating movements may give place to long-sustained and later to fixed deviation of the neck.

No causal neurological lesion has been found in those patients in whom the torticollis remains the sole abnormality. Spasmodic torticollis is thus sometimes regarded as a psychogenic disorder but it may well be due to as yet undefined organic changes within the extra pyramidal system. Certainly the condition often causes considerable social disability and pain.

The severity of the condition varies widely. In mild cases alleviation of anxiety (which aggravates the movements) by diazepam, 2 or 5 mg three times daily, is helpful. Most patients require more potent treatment. Tetrabenazine is probably the most effective drug available. An initial dose of 25 mg twice daily is built up by 25-mg increments at intervals of three or four days until a therapeutic response is obtained or side effects supervene. The usual maximal dose is 200 mg daily. The drug often reduces the frequency and amplitude of the torticollis but, unfortunately, few patients can tolerate the drug. Drowsiness and severe depression are frequent side effects. Overdosage leads to Parkinsonian hypokinesis. We have found that only 10% of patients who benefit from it can continue to take tetrabenazine for long. Alternatives such as haloperidol (1 mg twice or thrice daily) and thiopropazate (5 mg thrice daily) are worth trying.

Surgery should be considered for patients who are severely distressed by fixed, painful neck rotation. Many procedures have been used. Myotomy of the sternomastoid and other affected muscles alone is rarely effective for long. Intradural, bilateral section of the first three anterior cervical roots and the spinal accessory fibres is the most widely used procedure and often furnishes lasting relief. Stereotactic thalamotomy, alone or combined with section of the nerve roots, is also effective but even after combined surgery relief is sometimes incomplete. Such extensive surgery should be contemplated only when the patient is severely disabled and after careful explanation to him of the risks.

HEMIFACIAL SPASM

Hemifacial spasm may legitimately be considered as a tic since it consists of repeated twitching of facial muscles. Arising

in the middle-aged or elderly it usually, first affects the upper facial muscles, particularly the orbicularis oculi, causing a spasm of variable amplitude. The movements may remain confined to the upper face or may spread, affecting lower facial muscles as well. Spasms usually occur in bouts at varying intervals. The condition is thought to be due to degenerative changes in the nucleus or peripheral part of the facial nerve but its precise cause is unknown. There is invariably slight weakness of the muscles involved in the spasms.

The condition is harmless but may lead to embarrassment if the involuntary "wink" is interpreted as meaningful by an observer. An explanation to the patient that the movements are benign is often all that is required of the doctor. If the spasms are frequent and the patient perturbed about them an injection of 0·2 to 0·3 ml of 1% phenol in glycerine around the upper fibres of the facial nerve sometimes helps. This does not abolish the spasms but will reduce them sufficiently to prevent eye closure.

Tremors

A tremor is an oscillation of part of the body. It comprises regular, rhythmic to-and-fro movements. Tremors may be characterised by their rate and amplitude and by their direction, or directions, of movement. Their significance varies with the circumstances in which they occur. Some tremors occur at rest; others are seen during the maintenance of a posture, and intention tremors occur during active movements.

REST TREMORS

The commonest cause of a tremor observed in a limb relaxed and supported is Parkinsonism. The tremor, usually described as slow, varies in rate between 4 and 8 Hz. It usually first affects the fingers and spreads proximally up the arm and later to the leg. Characteristically, rhythmical movements are seen in two planes; flexion and extension of the fingers is accompanied by abduction and adduction of the thumb. As the movement of the thumb across the flexing fingers increases there is often concurrent pronation and supination of the wrist, resulting in a "pill-rolling" movement. The tremor is often the reason why the patient seeks advice and the symptom about which he remains most concerned. The approach to treatment of the tremor should be guided by the severity of the other Parkinsonian features—rigidity and, particularly, hypokinesis.

If the tremor is mild or moderate, and rigidity and hypokinesis slight in their effects on voluntary movement, no treatment is required, though the patient should be seen at intervals so that treatment may be started if the disease progresses. Tremor of embarrassing amplitude in the absence of appreciable hypokinesis may be treated with anticholinergic drugs. Initially 2 mg benzhexol should be given twice daily and the dose gradually increased to a maximum of 5 mg three times a day or until dryness of the mouth or blurring of vision indicate that the limit of tolerance has been reached. Alternative preparations such as benztropine, caramiphen, or benapryzine may be used equally effectively in a similar way. All of these anticholinergic drugs may produce serious side effects—such as psychosis, hallucinations, constipation, urinary retention, and glaucoma.

If Parkinsonian tremor is accompanied by impairment of voluntary movements then treatment with dopaminergic drugs should be given, together with anticholinergic preparations. Treatment with levodopa and similar preparations usually helps hypokinesis more than tremor though the latter may be improved. Levodopa is most conveniently given together with a dopadecarboxylase inhibitor. Either Sinemet or Madopar may be used. One tablet of Sinemet is roughly equivalent to 1 g of levodopa given alone. A suitable initial dose is half a tablet twice daily. This should be gradually increased by half a tablet every four or five days. Two to four tablets daily are usually effective. Madopar is prepared in two strengths: one contains 100 mg of levodopa as well as benserazide and the other contains twice as much of each drug. The weaker capsules are used in the introductory period and the dose built up. Average effective doses range from 400 mg to 800 mg of levodopa given in this combined form. Though the use of these combined preparations reduces the nausea and vomiting produced by levodopa alone, they are apt to produce troublesome side effects, notaby psychiatric disturbances and induced involuntary movements.

POSTURAL TREMORS

A fine, rapid (about 10 Hz in young adults) tremor is present in everyone while maintaining a posture and during movements. The oscillation is so restricted that this physiological tremor is usually undetected by the naked eye. The intensity may be increased so that it becomes visible. Anxiety, overindulgence in coffee or alcohol, and thyrotoxicosis commonly exaggerate physiological tremor. The underlying cause must be treated. Some drugs, notably lithium and the tricyclic antidepressants, may also augment physiological tremor. Since these drugs are usually prescribed for serious psychiatric illnesses they need not be withdrawn since the tremor rarely causes disability. Explanation of the situation to the patient is all that is needed.

Enhanced physiological tremor may be inherited as an autosomal dominant trait. It may begin at any time from youth to old age. Often called benign essential tremor, it is associated with no other abnormalities nor with detectable neurological lesions. First seen and often confined to the arms, it occasionally involves head, jaws, and legs. Usually there is little functional disability but sometimes patients have difficulty lifting a cup to their lips and many sufferers are disturbed about their ill-formed handwriting.

Characteristically, alcohol appreciably reduces benign essential tremor but prescribed drugs are usually ineffective though diazepam and propanalol (40 mg three times daily) may sometimes help.

INTENTION TREMORS

An intention tremor is shown when a limb moves towards a target and progressively increases in amplitude as the limb nears the target. It is a distinctive sign of cerebellar dysfunction and is most often seen in patients suffering from multiple sclerosis. Intention tremors in the arms impair the patient's ability to lift food and drink to his mouth and in the legs cause ataxia of gait. Intention tremors often lead to severe disability. Drug treatment is ineffective. Surgical procedures such as stereotactic thalamotomy or dentatomy are rarely helpful.

Simple devices may help the patient to cope. Long straws obviate the need to lift glasses. Adjustment to a wheel chair existence will prevent the patient injuring himself in falls. The skills of the occupational therapist can be most usefully employed in devising easier living conditions for the sufferer.

CHOREA

Choreiform movements are irregular, jerking, ill-sustained and unpredictable movements. The fully developed picture of chorea is easily recognised. In milder forms the condition may seem to be no more than restlessness or fidgeting. Often chorea is accompanied by athetoid movements.

SYDENHAM'S CHOREA

Sydenham's chorea occurs in adolescents and is commoner in girls. It is of rheumatic origin and often follows a streptococcal

throat infection. The onset may be abrupt or insidious. Choreiform movements are usually generalised and are accompanied by emotional lability so that often the condition is thought initially to be psychogenic. Pregnancy may precipitate a recurrence of chorea.

Because of the agitation the patient, in all but the mildest cases, should be admitted to hospital and preferably nursed in isolation. An initial course of penicillin should be given. Diazepam or haloperidol should be prescribed in doses appropriate to the patient's age and size.

Most patients recover within a month though more prolonged illnesses occur and relapses are common. Since many patients later develop rheumatic heart disease their future observation and management should be similar to that of patients who have suffered from acute rheumatism.

HUNTINGTON'S CHOREA

Huntington's chorea is of autosomal dominant inheritance and usually first presents in the 30s. Choreiform movements, often particularly prominent in the face, are accompanied by progressive dementia. The movements may be well controlled by tetrabenazine which is usually tolerated in effective doses in this condition. The mental deterioration is untreatable and institutional care may be needed as the disease progresses.

Ballism

Flinging limb movements of wide amplitude are called ballismic movements. They resemble choreiform movements but are much more violent. It is sometimes an arbitrary decision whether one calls movements severe chorea or ballism. Usually these movements suddenly affect one side of the body (hemiballism, or if less dramatic, hemichorea) as a result of a vascular lesion in elderly patients. Spontaneous improvement usually occurs over a period of weeks and during this time tetrabenazine may mitigate the disability. When hemiballism persists it can effectively be treated by stereotactic thalamotomy.

Athetosis

Athetoid movements are relatively slow, confluent, writhing movements, usually most prominent at the periphery of the limbs. They are often accompanied by choreiform movements and by forced axial rotations of the trunk and neck (torsion dystonia). Such movements usually result from cerebral hypoxia or trauma at birth or during the neonatal period and from kernicterus.

Drug treatment is usually ineffective, though tetrabenazine, haloperidol, thiopropazate, diazepam, and even benzhexol occasionally produce improvement in mild cases. Stereotaxic procedures, indicated only in severe disability, sometimes help.

Myoclonus

The term myoclonus describes sudden, shock-like muscle contractions. They may be generalised or restricted to a small group of muscles. Myoclonic jerks occurring immediately before sleep or on waking are normal and almost all people experience them occasionally.

Myoclonus may be of epileptic origin and sometimes occurs as a complication of hepatic or renal failure or as a manifestation of rare encephalopathies. Treatment is geared to the underlying cause.

Miscellaneous involuntary movements

Many of the dyskinesias discussed above arise from diseases of the extrapyramidal system, but lesions elsewhere may produce involuntary movements.

Partial epilepsy consists of repeated, circumscribed muscle contractions and often affects a hand or the face. It may continue for a prolonged period and needs treating with appropriate anticonvulsants (such as phenytoin 100 mg thrice daily).

Writer's cramp is a fairly common disability of uncertain origin. It consists of an involuntary spasm of the small muscles of the hand and sometimes of the arm which is triggered by the act of writing. Tremor and muscular aches occur, and the patient has to stop writing. The affected hand performs other fine skilled movements well. Analogous but rarer forms of cramp affect violinists, typists, and cotton "twisters." Tranquillisers of the types listed above are commonly prescribed but rarely help. Training the patient to write with his other hand is the most useful management. Sometimes patients respond to electrical deconditioning.

Extensor and flexor spasms of the legs result from severe lesions of the spinal cord, and commonly occur in disseminated sclerosis. These involuntary movements, reflexly triggered, are inconvenient, uncomfortable, and often very painful. They may respond to oral treatment with baclofen 30 mg to 60 mg daily. This drug should be introduced in small amounts (10 mg daily) and the dose slowly increased. It produces drowsiness and depression in many patients. If drug treatment is ineffective an intrathecal injection of phenol in glycerine may be needed to control severe painful spasms.

Fasciculations are visible spontaneous contractions of groups of muscle fibres or whole motor units. They are manifestations of denervation and usually produced by lesions lying proximally in lower motor neurones. A similar, common involuntary movement, benign fasciculation, occurs after unaccustomed use of muscles. The thighs and thenar eminences are often affected. These movements are apt to occur in anxious patients. They are liable to produce anxiety when they affect doctors or medical students who may believe that they presage the onset of motor neurone disease. Reassurance after a careful examination usually abolishes benign fasciculation.

Conclusion

This discussion of involuntary movements is far from complete. Analysis of movement patterns assessed with other neurological signs or associated systemic illnesses will usually allow treatable involuntary movements to be identified. In any child or young adult presenting with tremors or choreiform or athetoid movements the diagnosis of Wilson's disease (which can be treated effectively with penicillamine) should be considered. Almost any involuntary movement may be the result of taking drugs and this must be remembered.

Further reading

Detailed accounts of the nature, pathogenesis, and treatment of involuntary movements are given in:

Diseases of the Basal Ganglia, ed P J Vinken and G W Bruyn. Amsterdam, North Holland Publishing Company, 1968.

Denny-Brown, D, *The Basal Ganglia and Their Relation to Disorders of Movement*. London, Oxford University Press, 1962.

Neurophysiological Basis of Normal and Abnormal Motor Activities, ed M D Yahr and D P Purpura. New York, Raven Press, 1967.

Scientific Foundations of Neurology, ed M Critchley, J L O'Leary and B Jennett. London, Heinemann, 1972.

Recent reviews of the treatment of Parkinsonism are contained in:

Advances in Neurology, Vol 3, ed D E Calne. New York, Raven Press, 1973.

Recent Advances in Clinical Neurology, ed W B Matthews. London, Churchill Livingstone, 1975.

An account of the treatment of writer's cramp is given in:

Scientific Aspects of Neurology, ed H Garland. Edinburgh, Livingstone, 1961.