CHAPTER 18
MOVEMENT DISORDERS

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Unusual movements are common in children, and one must be able to distinguish an involuntary movement disorder from the fidgetiness of an otherwise normal child. Nose rubbing, scratching, or a "nasal salute" can be secondary to an allergic process or a tic, while similar recurring episodic movements such as brushing hair from the forehead or the eyebrow can be a manifestation of a photic fit. Recurring nasopharyngeal noises can be secondary to an allergy, convulsive disorder, or tic.

Disorders of movement, though not uncommon in children, are more often observed in the adult population. Anxiety and stress tend to increase their frequency and severity and, except for ballism and some patients with severe torsion dystonia, disorders of movement disappear during sleep. Unusual movements that occur during sleep, however, also can be a manifestation of convulsive activity.

Movement disorders can have an insidious onset and are often initially overlooked by parents and even the critical eye of teachers and physicians. Occasionally, the child is thought to be clumsy, but sooner or later it becomes apparent that the recurring movements are involuntary and not simply maladroitness. Patients can have other associated alterations of their neurologic status, including dysarthria or anarthria as well as changes of posture, muscle tone, and stretch reflexes. It is common to have associated behavioral changes such as irritability, irascibility, forgetfulness, or apathy.

NEUROANATOMIC CONSIDERATIONS

Clinicopathologic studies have associated movement disorders with the extrapyramidal system. Although not specifically neuroanatomically defined, it is traditionally accepted that the extrapyramidal system includes the basal ganglia, the subthalamic nuclei (Luysi bodies) found in the basilar portion of the diencephalon, and the substantia nigra, the largest nuclear mass in the mesencephalon. The caudate nucleus, the putamen, and the pallidum comprise the basal ganglia. The claustrum is also sometimes included. The corpus striatum is comprised of the caudate nucleus, putamen, and globus pallidus. The caudate nucleus and putamen are considered as the neostriatum and commonly referred to as the striatum, whereas the globus pallidus is defined as the paleostriatum.

The basal ganglia have many internuclear connections, with the preponderance of striatal outflow coursing to the globus pallidus. There are reciprocal connections between the striatum and the substantia nigra as well as between the pallidum and

subthalamic nuclei. There are multiple intranuclear interneuronal connections. Prominent afferent pathways to the basal ganglia come from all cortical areas, with fewer afferent connections arriving from the thalamic intralaminar nuclei and brain stem centers such as the locus ceruleus and the raphe nuclei. Afferent projections to the subthalamic nucleus (Luysi) are derived from the precentral motor cortex and other regions of the frontal lobe.

The most notable efferent connections arise in the pallidum and substantia nigra and project to the thalamus. Pallidothalamic fibers course by way of the ansa lenticularis and lenticular fasciculus to the ventral anterior, ventrolateral, and centromedial thalamic nuclei. Efferent fibers from the thalamus and substantia nigra course to the ventral anterior, ventromedial, and dorsomedial nuclei, with some collateral fibers directed to the superior colliculus. Fibers from the anterior and lateral ventral nuclei project to the motor and premotor cortex and efferent striatal fibers to the premotor and supplementary motor region. The thalamus has the important function of integrating information from the basal ganglia and cerebellum en route to the cerebral motor cortex.

Clinical Syndromes with Associated Movement Disorders

Athetosis is an irregular writhing movement of the extremities, which primarily affects the distal muscles. It can be intermittent or continuous, and there are varying degrees of alternating pronation and supination, flexion and extension, or combinations thereof. The movements appear to be increased when the patient is under increased stress or tension.

It can be present in patients who have sustained some perinatal insult. These patients are commonly hypotonic during infancy though they usually have brisk stretch reflexes, and athetosis may not be apparent until after the age of 12 to 18 months. The differential diagnosis of athetosis is similar to that of chorea and the two types of movement disorders are often associated, appearing as choreoathetosis. Other causes of athetosis include prenatal injury, developmental anomalies, kernicterus, abnormalities of uric acid metabolism, Leigh disease, juvenile Niemann-Pick disease, ataxia-telangiectasia, Wilson disease, Hallervorden Spatz syndrome, Pelizaeus-Merzbacher disease, the adverse effects of neuroleptic drugs (levodopa and phenothiazines), and postencephalitic or posttraumatic states.

There is no one pathologic lesion associated with this movement disorder. If secondary to perinatal hypoxia, a neuronal loss is most prominent in the caudate nucleus, putamen, and globus pallidus but can also be demonstrated in the thalamus, amygdala, and portions of the hippocampus. Status marmoratus consists of whitish streaks resembling a "marbled appearance" of the lateral part of the corpus striatum and, less often, in the head of the caudate nucleus and thalamus. These changes are usually bilateral, and at microscopic examination show bundles of myelinated fibers, a focal loss of neurons, and glial scarring. More recently it has been shown that there is an abnormal orientation of myelinated fibers coursing through the scarred tissues without abnormality of myelin formation.

Although there is no specific treatment for athetosis at this time, physical therapy may be of some benefit in maintaining joint mobility and gait training. Administration of levodopa with a decarboxylase inhibitor has resulted in some symptomatic improvement, especially in lessening muscular rigidity. Surgical procedures, including stereotactic thalamotomy and cervical spinal cord electrode emplacement and stimulation, have been reported in the past to be of some benefit in the occasional patient, partic-

ularly those who also have corticospinal tract dysfunction. Stereotactic thalamotomy may be of some limited benefit, but there is no substantive evidence that dorsal spinal cord stimulators have lessened the symptoms.

Ballism, uncommon at any age, is particularly rare in children and is characterized by a violent flailing of one limb (monoballismus), involvement of the ipsilateral arm and leg (hemiballismus) or bilateral limbs (biballismus). The proximal muscles are primarily affected, and the flailing of the limbs can be so disabling that the patient can be thrown from the bed to the floor. This florid movement disorder can be associated with lesions usually found in the contralateral subthalamic nucleus and include ischemia or infarction, hemorrhage, tumor, and a variety of other focal pathologic abnormalities. It is rarely observed as a transient stage in the progressive deterioration of patients with subacute sclerosing panencephalitis or other viral encephalitides.

Patients can die from exhaustion, and meticulous supportive medical care is essential. Current treatment to lessen the severity of this movement disorder consists of administration of barbiturates or phenothiazines. There is usually a gradual reduction of the ballismic movements, but some patients have residual athetosis and/or chorea. In those patients who do not respond to medication, surgical lesions have been made in the globus pallidus or thalamus with varying results.

Chorea is usually thought to be secondary to perinatal trauma, manifestations of Sydenham or Huntington chorea, systemic lupus erythematosis, and a variety of other conditions (see Table 18.1). Choreiform movements are quick jerks of skeletal muscles

Table 18.1 Medical Conditions in Which Chorea Has Been Reported

Degenerative/Genetic	Canavan disease; ataxia-telangiectasia; Bassen-Kornzweig disease; benign familial chorea; Fabry disease; familial microcephaly with chorea; familial paroxysmal choreoathetosis; Friedreich ataxia; glutaric aciduria; Hallervorden-Spatz syndrome; Huntington disease; incontinentia pigmenti; Lesch-Nyhan syndrome; cerebral lipidoses; phenylketonuria; porphyria; presenile dementia; tardive dyskinesia; torsion dystonia; Wilson disease
Infectious Diseases	Diphtheria; encephalitis (St. Louis, inclusion body; mumps; pertussis; rubeola; scarlet fever; typhoid; varicella)
Metabolic/Endocrinologic	Addison disease; beriberi; burns; hypocalcemia; hypomagnesemia; hyponatremia; hypoparathyroidism; kernicterus; thyrotoxicosis; vitamin B_{12}
Neoplastic	Brain tumors
Toxic	Carbon monoxide; fenfluramine; hyoscine, isoniazide; lithium; mercury; metaclopramide; phenothiazines; phenytoin; reserpine; scopolamine
Trauma	Posttraumatic syndrome
Vascular Diseases	Cerebrovascular disease; Henoch-Schönlein purpura; lupus erythematosus; polycythemia vera

Adapted with permission from Greenhouse AH. On chorea, lupus erythematosus, and cerebral arteritis. *Arch Intern Med* 1966; 117:389–393.

that move randomly from place to place; the face, trunk, and limbs can be involved, affecting proximal as well as distal muscles. These random movements are usually bilateral but can affect only unilateral muscles (hemichorea). There are techniques to better demonstrate choreiform movements, including having the child maintain a posture of forward arm and/or leg extension for a matter of minutes. In addition to the choreic movements, the wrists are flexed and there is hyperextension ("spooning") of the metacarpophalangeal joints. If the tongue is protruded, it may be intermittently retracted, assuming a "trombone effect." When the patient grasps the examiner's finger(s), there is an intermittent squeezing of the finger(s), known as the "milkmaid's hand."

The movements are commonly not initially recognized as being abnormal but rather are ascribed to "figiditiness, nervousness, or stress." Speech can be slurred or periodically interrupted; moreover, there can be a decrease of speech output and spontaneity, as well as problems with chewing and/or swallowing.

Sydenham chorea is characterized by emotional lability, muscular hypotonia, and choreiform movements. Although the relationship of Sydenham chorea to rheumatic fever was first suggested in the 18th century, it was not until the 19th century that there was a general acceptance of this association by physicians. The majority of cases of Sydenham chorea are preceded by a streptococcal infection or rheumatic fever, usually with an interval of 2 to 7 months between the infection and the manifestations of chorea. There is a family history of rheumatic fever in about one fourth of patients, and manifestations of Sydenham chorea are found in 3.5% of parents and about 2% of siblings.

The clinical signs and symptoms have an insidious onset, usually between the ages of 3 and 13 years and are more commonly seen in females than males. Affected patients appear restless or fidgety and become emotionally labile. It is not unusual that parents do not know when the process actually began, and when the patient is first evaluated by physicians, the problem not uncommonly is considered to be on an emotional basis or ataxia of unknown origin. Facial grimacing, dysarthria, and rarely anarthria can be present. Voluntary movements and muscle power can be mildly to severely impaired, with normal stretch reflexes, and hypotonia is commonly present. Plantar responses are flexor, and the sensory system is normal.

Sydenham chorea can persist from several months to over a year before there is a gradual lessening of the abnormal movements. Unfortunately, 20% to 30% of patients can have multiple recurrences of signs and symptoms of the disease, usually within the first 2 years after symptom onset. Patients generally have complete recovery from signs and symptoms, though some may have residual clumsiness, minor restless movements, or tremor.

Because of the association with rheumatic fever, a search for streptococcal infection must be made; however, throat cultures have not been particularly reliable in identifying the organism. Anti-streptolysin-O titers, though widely used, are often of little diagnostic help, for they rise early and decline quickly. Other antibody tests include anti-desoxyribonuclease B (anti-DNase B), antihyaluronidase (AH), antistreptokinase (ASK), and anti-nicotinamide-adenine dinucleotidase (anti-NADase). A twofold rise in the titer is present if samples are obtained within 2 months of the streptococcal infection.

Valvular heart disease is found in about one third of patients with Sydenham chorea; other manifestations of rheumatic fever include polyarthritis, erythema marginatum, and subcutaneous nodules. If there is no clinical or laboratory evidence upon which to establish a rheumatic basis for patients with "pure" chorea, only later manifestations of arthritis and/or carditis will lend retrospective evidence of the disease. It is recommended, therefore, that all patients with Sydenham chorea and chorea of

unknown cause should be treated with continual antibiotic prophylaxis, consisting of either monthly injections of 1.2 million units of benzathine penicillin G, or the oral administration of 200,000 units of penicillin given two or three times daily. The prophylactic treatment should be continued at least until the end of the second decade; however, it is prudent to continue the treatment indefinitely.

Patients should remain as quiet as possible for the family setting. This may require bed rest and the administration of barbiturates, chlorpromazine, or haloperidol; valproic acid has more recently been administered with reasonably good results. Usually the choreic movements lessen within several days following the administration of medication and are often completely controlled within days to several months, although some patients require a longer treatment. If there are recurrences of chorea following drug withdrawal, the medication may have to be reinstituted. The duration of Sydenham chorea varies from about 1 month to several years; about 20% to 30% can have recurrences of chorea. There is usually complete recovery, but mild incoordination, fidgetiness, or tremor can persist.

Neuropathologic studies in the past have shown only nonspecific findings, including arteritis with some neuronal loss in the caudate nucleus, putamen, and multiple diffuse areas of neuronal loss in the cerebral cortex and cerebellum. More recent studies have shown that 46% of children with Sydenham chorea had sera containing IgG antibodies, which reacted with the neuronal cytoplasmic antigens primarily in the region of the caudate and subthalamic nuclei.

Huntington disease is a progressive degenerative disease, inherited as an autosomal dominant trait, characterized by chorea, dementia, and hypotonia. Some patients have bradykinesia or rigidity, tremors, and, rarely, convulsions. The disease usually affects adults, but about 5% of patients are less than 14 years old, and infants can be affected. Patients with the juvenile form of Huntington disease, equally represented in males and females, are four times more likely to inherit the disease process from the father; however, there is no known explanation for the higher frequency of paternal inheritance. Since there may be a higher incidence of maternal inheritance in patients with later onset of the disease, it has been suggested that there are maternal genetic factors that can influence the clinical expression of the disease. There is evidence that the gene for Huntington disease is located on the short arm of chromosome 4.

The initial manifestations of juvenile Huntington disease are generally notable for mental deterioration, rigidity, seizures, hyperkinesia, and/or choreoathetosis. The convulsive manifestations are usually generalized and manifested as tonic-clonic, myoclonic, or akinetic spells. Occasionally ataxia and tremors are present. Oculomotor abnormalities can be present and manifested by slow saccades, convergence paresis, and impersistence of gaze. Often there are concomitant head thrusts or "jerky" head movements with associated slow ocular movements, similar to but not the same as oculomotor apraxia, since vertical movements can also be affected. Moreover, patients can have an orolingual apraxia. There is progression of neurologic abnormalities with an average survival of 8 years in affected children and 25 years in adults.

The electroencephalogram (EEG) is characterized by a low-voltage "featureless" record with background activity predominantly in the beta range. Irregular slow activity can be present, and paroxysmal abnormalities are occasionally found. Computed tomography (CT) and magnetic resonance imaging (MRI) of the brain show atrophy of the caudate nuclei, but there are no other distinguishing radiographic features of the disease. Positron emission tomography (PET) is more specific in that it shows a reduction of glucose metabolism in the caudate nuclei of affected patients.

Neuropathologic findings are notable for a small brain with widened sulci and fissures. Coronal sections show atrophy of the striata, particularly notable in the cau-

date nucleus. There is a striking decrease of neurons in the striata, and the cerebral cortex, as well, shows some neuronal loss. The neurons lost in this disease are not limited to one neurotransmitter system, for a variety of neurotransmitters have been evaluated and have been found to be reduced or increased. Striking abnormalities include the reduction of gamma-aminobutyric acid (GABA), acetylcholine, and of the activities of glutamic acid decarboxylase (GAD) and choline acetyltransferase (CAT). The somatostatin-neuropeptide Y neurons of the striatum are relatively spared. These findings have suggested that the neuronal degeneration in Huntington disease is secondary to excitotoxicity by way of *N*-methyl-D-aspartate (NMDA) receptor stimulation.

There is no specific treatment for patients with this disease. Choreiform movements may benefit from the administration of tetrabenazine or reserpine, and some patients have had some benefit from haloperidol. Antidepressant drugs have been used in some cases, but there is no specific treatment for the dementia.

Benign familial chorea is a nonprogressive, though persistent chorea that begins during childhood. It is generally static, though some patients have demonstrated some improvement in later years. The disease, inherited as an autosomal dominant trait with incomplete penetrance, is commonly manifested initially by delayed acquisition of motor milestones. A generalized chorea then becomes apparent; it is usually mild but can be of notable severity in some patients. There is no associated mental deterioration or psychiatric disturbance.

Familial paroxysmal choreoathetosis (PKC) is a condition characterized by paroxysmal episodes of choreoathetosis and/or dystonia. At least two syndromes are included in this category, the more common of which is paroxysmal kinesigenic choreoathetosis, notable for recurring paroxysmal episodes of choreoathetosis that are precipitated by startle or initiation of movement and occur particularly after rest. This form of the disease usually begins in childhood or adolescence, affects males more often than females, and increases in frequency during adolescence. As the patient becomes older, the recurrent paroxysmal events seem to lessen in frequency and severity. The duration of each episode is a matter 1 to several minutes, and symptoms are commonly controlled by the administration of phenytoin. The EEG is normal. The etiology of PKC remains unknown, although there have been suggestions that it is inherited as an autosomal dominant trait.

Patients with **paroxysmal nonkinesigenic choreoathetosis (PNKC)** commonly have onset of symptoms during infancy, and males are affected more frequently than females in a ratio of 3:2. The clinical appearance of the episode is somewhat similar to that of PKC, but it persists for longer periods of time, ranging from minutes to several hours. These episodes are often precipitated by fatigue, emotional stress, alcohol, or caffeine. The frequency of these episodes is less than in PKC, sometimes occurring only several times each month. Patients with PNKC have achieved some relief from symptoms by the administration of benzodiazepines, such as clonazepam, but carbamazepine and haloperidol have also been found to control the paroxysmal events. Patients generally have responded poorly to the administration of phenobarbital or phenytoin.

Dystonia (dystonia musculorum deformans [DMD], idiopathic torsion dystonia [ITD]), is manifested by persistent muscular contractions that can involve virtually all skeletal muscles at some time and result in abnormal twisting movements and postures, sometimes so unusual it is presumed that the patient is an hysteric. There are many conditions that can result in dystonia (see Table 18.2).

The condition is generally considered in terms of the body parts involved in the abnormal posturing; viz., **focal** dystonia, which refers to posturing of a single anatomic

TABLE 18.2 Classification of Dystonia

Primary dystonia	**Generalized dystonia** (dystonia musculorum deformans, idiopathic torsion dystonia): hereditary (dominant, recessive, X-linked); sporadic **Focal, segmental, multifocal:** cranial dystonia (Meige syndrome, blepharospasm-oromandibular dystonia); spasmodic torticollis; writer's cramp and other occupational dystonias; spasmodic dysphonia; others **Idiopathic paroxysmal dystonias:** kinesigenic; non-kinesigenic
Secondary dystonia	**Diseases with known metabolic defect:** Wilson disease, G_{m1} gangliosidosis, G_{m2} gangliosidosis, metachromatic leukodystrophy, Lesch-Nyhan syndrome, glutaric aciduria, methylmalonic acidemia, homocystinuria, others **Diseases with presumed (but undefined) metabolic defects:** Hallervorden-Spatz syndrome, calcification of basal ganglia (with or without Leber optic neuropathy), dystonic lipidosis (Niemann-Pick disease), ataxia-telangectasia, neuroacanthocytosis, ceroid lipofuscinosis, Hartnup disease **Degenerative CNS disease:** Parkinson disease, dopa-responsive dystonia, progressive supranuclear palsy, Huntington disease, pallidal degeneration, olivopontocerebellar atrophy, Azorean disease **Nondegenerative CNS disorders:** perinatal anoxia/ kernicterus (can be delayed); head trauma (often delayed); cerebral infarction (often delayed); arteriovenous malformations; tumor; encephalitis (various types); toxins (especially manganese); postoperative (after thalamotomy); multiple sclerosis (often paroxysmal); drugs (especially neuroleptics, dopamine agonists) **Psychogenic dystonia**
Disorders simulating dystonia	**Orthopedic disorders:** rotational atlantoaxial subluxation **Neurologic:** seizures, posterior fossa tumor-causing torticollis, hemianopia, strabismus **Miscellaneous:** hiatal hernia in childhood (Sandifer syndrome), congenital neck muscle lesions, abnormal posture *in utero,* others

Adapted with permission from Lang AE. Movement disorder symptomatology. In: Bradley WG, Daroff RB, Fenichel GM, et al, eds. *Neurology in Clinical Practice.* Stoneham, MA: Butterworth-Heinemann, 1989; 1:315–336.

region, including the eyelids (blepharospasm), oromandibular dystonia, spastic dysphonia when the larynx is affected, torticollis when the neck muscles are involved, and writer's cramp. **Segmental** dystonia refers to abnormal posturing affecting several contiguous regions of the body, and **generalized** dystonia, when the lower limbs and other anatomic regions are involved.

ITD usually begins in childhood with inversion and plantar flexion of the ankle so that the child has a tendency to walk on the toes. It is often overlooked by the parents until the child participates in some physical activity with peers and the comparative difference in movement of the lower limbs becomes readily apparent. The symptoms and signs are initially intermittent and made worse when the patient is under inordinate stress. As time passes, the dystonia progresses and spreads to adjacent parts of the

limb(s) and, ultimately, the remainder of the torso and other limbs. One type of ITD, usually with onset of signs and symptoms between age 4 and 16 years, has been found to have a high incidence in the Ashkenazim and is notable for its rather rapid clinical course. It has been suggested that it is inherited as an autosomal dominant trait with variable penetrance. The disease has also been found in other ethnic groups, including the French-Canadians and the Swedes, in which the onset is usually later and there is a slower progression of clinical symptoms. In some families the disease has been localized to the long arm of chromosome 9q32-q34.

These abnormal movements can involve the facial, nasopharyngeal, and laryngeal muscles, resulting in abnormalities of speech, swallowing, and breathing. It is commonly believed that patients who have onset of signs and symptoms before age 11 years will progress to generalized dystonia and that those patients who have later onset can have remissions and exacerbations of symptoms. The diagnosis of ITD requires that, in addition to the dystonia, the patient has a normal perinatal and developmental history, there has been no antecedent disease or drug consumption that could possibly result in dystonia, there is no impairment of intellectual function or sensory symptoms, and there is no abnormality of copper metabolism.

The medical management of these patients is often ineffective, but at this time the best therapeutic results have been obtained from the administration of high-dose trihexyphenidyl hydrochloride (Artane). Children seem to tolerate the high doses of this medication (40 to 80 mg/day, beginning with 2 to 4 mg/day and gradually increasing the dosage) better than adults. Other drugs that have been used include baclofen, bromocriptine, carbamazepine, and the benzodiazepines; both dopamine agonists and antagonists have also been reported to be of some value. Some patients benefit from stereotactic thalamotomy, but the relapse rate, as well as the incidence of complications including dysarthria, hemiparesis, and ataxia, is significant. There is no substantive evidence that emplacement of dorsal column stimulator is of any benefit.

ITD can be a devastating disease, and physicians should enlist all support facilities available to assist the patient and family. About one third of patients become bed-bound or confined to a chair, one third remain moderately disabled, and the remaining one third remain mildly disabled yet independent.

Bobble-head doll syndrome is characterized by an intermittent "bobbing" movement of the head and neck at a frequency of 2 to 5 Hz, giving the appearance of a doll's head supported by a spring wire. Often the head movements can be voluntarily stopped on command, only to reappear when the child's attention is directed to other stimuli. Most reported patients have arachnoidal cysts or other obstructive space-occupying lesions in the region of the third ventricle or the aqueduct of Sylvius.

Hereditary chin quivering (trembling) is an uncommon clinical entity, inherited as an autosomal dominant trait, manifested by rhythmic tremor (quivering) of the mentalis muscle. It is usually intermittent, lasting minutes to hours, and has a frequency of 2 to 11 Hz; it has been reported to be persistent in some cases. Chin quivering can appear at birth or any time thereafter and persists throughout life, though it lessens with age and, on occasion, may disappear. It can occur without known provocation, but it can also be precipitated by emotional stress, after waking abruptly from sleep, during periods of intense concentration, or after being pecked on the lips by a parakeet, and it has been reported to occur when playing the violin. There is no known relationship to the palmomental, reflex nor are there any known associated neurologic abnormalities.

Recommendations for treatment have included the administration of sedatives, anticonvulsant medications, and psychotropic drugs. More recently, regular injections of botulinum toxin to the mentalis muscle have been reported to be beneficial.

Spasmus nutans is, for the most part, a self-limiting benign condition of unknown origin, characterized by an intermittent head tilt, head nodding in any direction, and low-amplitude nystagmus of high frequency that can be conjugate, dysconjugate, or monocular. The nystagmus can be horizontal, vertical, or rotary. The episodes usually last less then 1 to several minutes. The onset of spasmus nutans is commonly between the ages of 3 and 12 months and often begins during the period of winter through spring months.

This condition can persist from several months to several years, and there are no known sequelae. It must be noted, however, that spasmus nutans has been reported to occur in infant patients with gliomas of the optic nerve and/or chiasm and has been associated as well with arachnoidal or porencephalic cysts. Neuroimaging studies should always be performed when the diagnosis of spasmus nutans is first considered.

Tics

Tics are involuntary, stereotyped, quick purposeless movements that usually involve the same muscle or muscle groups. Facial, neck, and/or shoulder muscles are most often involved, as in eye blinking, widening of the palpebral fissure, dilation of the nares with facial grimacing, head movements, or shoulder shrugging. One form of tic can disappear only to have another tic affecting other muscles appear at a later time. Whether a simple movement or variations on a theme, they are repetitive and generally affect the same anatomic regions, as compared with chorea in which the muscle jerks move unpredictably from place to place. Myoclonus, a paroxysmal brief, rapid contraction of part of a muscle, a muscle, or a group of muscles, must be considered in the differential diagnosis.

As in the case of specific disorders of movement, tics usually increase in frequency and severity when the patient is under stress; they disappear during sleep. They are common in childhood, with a reported incidence ranging from 4% to 23%. Boys are affected at least three times more often than girls, and the age of symptom onset varies from 3 to 15 years with a mean of 7 years.

A simple tic, commonly called a "habit pattern," has been reported to affect up to one fourth of children at one time or another. They are rarely first noted before school age or in adolescence. Simple tics usually persist weeks to no longer than 6 months; however, other simple tics can then appear.

The pathophysiology of tics is not well understood, and there is no evidence to presume that tics have a psychogenic basis. The traditional recommendation is to quietly observe and support the child rather than to "remind" or "correct" him. There is merit, however, in understanding areas of stress for the child at home and school. This may require neuropsychologic testing, a family interview, and possibly family therapy directed by those with specific expertise in child behavior.

Occasionally, the simple tic can severely disrupt a child's activities at home and school. These patients may benefit from the daily administration of low doses (1–2 mg/day) of haloperidol; however, the administration of drugs should be reserved for only those occasional patients who are severely handicapped by the tic. Duration of therapy should be no longer than several months, for simple tics are generally a self-limiting phenomenon. Should the tic persist longer than 6 months, one must consider the possibility that the patient has a complex, chronic tic and provide careful long-term follow-up evaluations (see Table 18.3).

Complex tics, on the other hand, are coordinated movement patterns involving a

TABLE 18.3 Classification of Tics

Simple motor tics	Eye blinking, eyebrow raising, nose flaring, grimacing, mouth opening, tongue protrusion, platysma contractions, head jerking, shoulder shrugging or abduction, neck stretching, arm jerks, fist clenching, abdominal tensing, pelvic thrusting, buttock or sphincter tightening, hip flexion or abduction, kicking, knee extension, foot dorsiflexion, toe curling
Simple phonic tics	Sniffing, grunting, throat clearing, shrieking, yelping, barking, growling, squealing, snorting, coughing, clicking, hissing, humming, moaning
Complex motor tics	Head shaking, teeth gnashing, wrist shaking, finger cracking, touching, hitting, jumping, skipping, stamping, squatting, kicking, smelling hands/objects, rubbing, finger twiddling, echopraxia, copropraxia, spitting, exaggerated startle
Complex vocal tics	Coprolalia (wide variety, including shortened words), unintelligible words, whistling, "Bronx cheer" or "raspberry," panting, belching, hiccough, stuttering, stammering, echolalia, palilalia (also mental coprolalia and palilalia)

Adapted with permission from Weiner WJ, Lang AE. *Movement Disorders: A Comprehensive Survey.* Mount Kisco, NY; Futura; 1989.

number of muscles in their usual synergistic relationships. It can be difficult to separate some motor disturbances that can be associated with the tic, such as obsessive-compulsive behavior, attentional deficit disorders, and impulsivity.

Patients with **Gilles de la Tourette syndrome (Tourette Syndrome, TS)** have multiple complex, stereotyped tics, including hopping, jumping, truncal and pelvic gyrations, thigh tapping, repetitive twisting, or turning around. There can be compulsive touching or hitting of other persons, biting, or sniffing of objects within the immediate environment. Vocalizations can also be present and include grunting, clucking sounds, repetitive sounds of swallowing or as if clearing the throat, screams, shrieks, and occasional animal sounds such as barking or growling. The tic should be present for at least 1 year before the diagnosis of TS is established.

Some patients with TS will explosively utter the usually well recognized obscenities (copralalia). Others will repeat a word or phrase over and over that has been said to them (echolalia), and some patients will repeat a word or phrase with increasing rapidity (palilalia).

The tics can be voluntarily suppressed for limited periods of time only to reappear; moreover, there is a waxing and waning of the frequency and location of the symptoms. The tics, both motor and phonic, can be simple and complex, with an onset ranging from 2 to 15 years and an average age of onset of 7 years. Males are reported to be affected more often than females in a ratio ranging from 3:1 to 5:1. The clinical course of the disease is lifelong. A familial pattern is well documented, and in recent computer modeling studies of TS family pedigrees, the condition appears to be inherited as an autosomal dominant trait. Moreover, there is evidence that the gene can be mapped to the long arm of chromosome 18.

The common clinical course of the disease is initially that of an attentional deficit

and hyperactivity from age 4 to 6 years, followed by the onset of simple motor tics. From 7 to 11 years, there commonly is a rostrocaudal progression of complex motor and phonic tics, and during the second decade, obsessive-compulsive symptoms occur in up to one half of patients.

The neurologic examination is generally normal in these patients except for the presence of "soft neurologic signs" that are commonly observed in patients with specific learning disabilities. Other laboratory studies, including neuroimaging, are normal.

Haloperidol appears to be the most consistently beneficial drug, providing relief from tics in 50% to 80% of patients. It should be started at a low dose of 0.5 mg daily to be increased by another 0.5 mg each week until the patient receives a total daily dose of 2 to 3 mg. Generally patients who have had a good response require no higher drug dosage. Pimozide as well as fluphenazine have also been effective in controlling the signs and symptoms of TS.

Extrapyramidal reactions can occur following administration of these drugs and include dystonia, akathesia, hyperreflexia, opisthotonous, or oculogyric crises. The treatment of patients with complex, chronic tics is difficult and frustrating not only for the patient and family but also for the physician. Family therapy and patient support groups can be important and should be considered a part of the overall treatment plan.

Tremors

Tremors are rhythmic oscillations of variable frequency that usually affect only the hands, although the head and torso are occasionally involved. The tremor is usually apparent early in the second decade but can appear as early as 5 to 6 years of age. Some tremors are present when the patient is not otherwise moving (resting tremor), as seen in parkinsonism in the adult, whereas others occur only when the limb is moved or while maintaining a posture (action tremor). The frequency of an idiopathic tremor is usually 3 to 7 Hz; tremors of greater frequency are found in some toxic states.

A familial tremor (essential tremor), inherited as an autosomal dominant trait, can appear in early childhood. It is usually bilateral, initially affecting both hands at a frequency of about 4 to 7 Hz. It can be slowly progressive until early adult life, when it tends to stabilize, and then with aging tends to decrease in severity. However, some older patients have progression of symptoms affecting the head, neck, mouth, and tongue. Commonly, rapid tremors do not interfere with fine motor coordination, but coarse tremors may preclude any coordinated movement. Adult patients can have lessening of symptoms from the daily administration of propranolol or for a short period after a drink of spirits. Children with familial tremor rarely need any treatment.

In rare instances, tremor is a manifestation of hysteria and is observed primarily in adolescents. In these cases, the tremor is usually coarse, of variable frequency, and often has a bizarre quality. Commonly, when attempts are made to restrain the affected limb, the tremor can move to other limbs, the head, or torso.

Iatrogenic tremor, usually of rapid frequency, can occur as an adverse effect of certain drugs, including the central nervous system (CNS) stimulants, some antihistamines, and psychotropic drugs such as the phenothiazines, tricyclics, or lithium. Occasionally, tremors are present in metabolic abnormalities such as thyrotoxicosis in which patients can have a rapid, fine tremor of fingers and hands, best observed when the arms are outstretched. Other metabolic disorders in which tremors may be present include hypocalcemia, hypomagnesemia, and uremia. Tremors can also be present in patients with Hallervorden-Spatz syndrome and juvenile Huntington disease.

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