A

lthough movement disorders do not usually present as neurologic emergencies, there are times when the abrupt onset of an unusual movement abnormality results in emergency department or intensive care unit consultations. Part 1 of this review discussed hypokinetic movement disorders emergencies. Part 2 provides a diagnostic approach to the recognition and treatment of hyperkinetic movement disorders emergencies by identifying phenomenology and reviewing common etiologies.

HYPERKINETIC DISORDERS

Chorea

Chorea consists of involuntary, irregular, purposeless movements that “flow” into one another in a random fashion. Though often referred to separately, the distinction between chorea, athetosis, ballismus, and dystonia is somewhat arbitrary and relates more to speed, amplitude, and duration of the movement rather than underlying pathology. A combination of these movements is often encountered in a single patient. Chorea may result from toxic/metabolic, vascular, and infectious/inflammatory disorders and may be acute in onset. Sydenham chorea is the most common cause of acquired, childhood-onset chorea.1 Sydenham chorea is the neurologic manifestation of rheumatic fever and is sufficient to make the diagnosis of rheumatic fever.2 Its incidence has declined dramatically with the widespread availability of penicillin; nevertheless, Sydenham chorea remains prevalent in areas where access to health care is limited.3,4 In addition to chorea, which may be generalized or hemichorea, additional features of Sydenham chorea include behavioral changes (obsessions, compulsions, hyperactivity, and emotional lability), weakness, hypotonia, and, rarely, vocalizations.5 Symptoms may begin abruptly, from 1 to 6 months after streptococcal pharyngitis.6 The diagnosis is made clinically, though elevated antistreptolysin O antibody titers are supportive.7 Treatment with penicillin may prevent the cardiac complications of rheumatic fever and is sometimes continued for several years as prophylaxis.7 If Sydenham chorea requires treatment, 1 prospective, nonrandomized trial suggested that valproic acid or carbamazepine may be used.1 Based on observational data, dopamine receptor blockers or dopamine depleters are also recommended.8

Chorea gravidarum usually begins in the first or early second trimester.9 Chorea may be unilateral or bilateral, often involving the face as well as the limbs. Dysarthria is common.10 Chorea usually resolves by the third
Although chorea gravidarum is rarely an emergency, it is likely that neurologists will encounter this entity in the context of an emergent inpatient consultation on a maternity ward.

The antiphospholipid syndrome may result in acute generalized chorea. Antiphospholipid syndrome may be primary or secondary to systemic lupus erythematosus. Antiphospholipid syndrome is thought to be the most common cause of chorea gravidarum in industrialized nations.¹² Rarely is chorea the sole manifestation of an emergent inpatient consultation on a maternity ward. Antiphospholipid syndrome may be the most common cause of chorea gravidarum in industrialized nations.¹² Other metabolic causes include hyperthyroidism and hyperglycemia (Table 2).

Hemichorea-Hemiballism

Hemichorea refers to large-amplitude, flinging movements of one side of the body that can be violent. Hemiballism resolves over days to weeks, the movements often become choreiform. Historically, the most common cause of hemiballism was stroke involving the subthalamic nucleus. However, this etiology is rare, with an annual incidence of less than 1 per 100,000 in a population-based study from Belgrade, Serbia.¹³ Although stroke remains the most common cause, only a minority of cases have lesions within the contralateral subthalamic nucleus.¹⁴-¹⁶ The second most commonly reported cause of hemiballism is nonketotic hyperglycemia. With this disorder, chorea, or ballism, may be unilateral or bilateral. It occurs more in women,¹⁷ and it may be the initial presentation of diabetes mellitus. Magnetic resonance T1-weighted images demonstrate hyperintensity in the putamen, caudate nucleus, and globus pallidus.¹⁷⁻¹⁸

(Figure 1). Magnetic resonance imaging findings result from ischemic injury due to hyperviscosity and regional metabolic failure. Like hemiballism secondary to stroke, the movements typically subside over a period of months. In some patients, abnormal movements reverse when the glucose level is normalized. Resolution of magnetic resonance imaging signal change correlates with clinical improvement in chorea.¹⁷ If treatment is required (eg, violent, self-injurious, exhausting, or distressing movements), dopamine receptor blockers or dopamine depleters such as tetrabenazine or reserpine are used (Table 3). Because the movements usually resolve over time,¹⁸ medication should be tapered after 3 months and the patient, reevaluated.

Myoclonus

Myoclonus consists of sudden, brief shocklike movements that may be due to muscle contraction (positive myoclonus) or loss of muscle tone (negative myoclonus or asterixis). Neurologists are often emergently consulted in the intensive care unit to see patients with myoclonus as a result of toxic/metabolic derangements or cerebral anoxia. It is also seen in serotonin syndrome and neuroleptic malignant syndrome. Medications including monoamine oxidase inhibitors, selective serotonin
reuptake inhibitors, serotonin-norepinephrine reuptake inhibitors, tricyclic antidepressants, opiates, levodopa, gabapentin, triptans, lysergic acid diethylamide (LSD), amphetamines, cocaine, and 3,4-methylenedioxyamphetamine (MDMA or ecstasy) may cause myoclonus. Hepatic and uremic encephalopathies are the most common metabolic derangements resulting in myoclonus and asterixis.19,20

Cerebral anoxia may result in 2 distinct myoclonic syndromes, myoclonus status epilepticus and postanoxic myoclonus (Figure 2). Myoclonus status epilepticus may begin in the hours immediately after a cerebral anoxic event. This occurs in approximately 30% of comatose adult survivors of cardiac arrest.21 The presence of myoclonus status epilepticus, defined as “spontaneous, repetitive, unre-

Figure 1. T1-weighted axial magnetic resonance image with signal hyperintensity in the putamen, caudate, and globus pallidus in a patient with hyperglycemic hemichorea.

Tics

Tics are either brief paroxysmal movements or vocalizations that are sometimes accompanied by a premonitory urge. They may be stereotyped and, unlike other hyperkinetic movements, may be voluntarily suppressed for a short period.

Motor tics are common in schoolchildren, with a prevalence of 3.2% to 9.6%.28 Even though it is rare for tics to present as an emergency, 2 situations may bring a patient with a tic disorder for emergency evaluation: tic exacerbation and neurologic compromise secondary to tics. Tic disorders wax and wane and some factors lead to marked exacerbation of tic severity including fatigue, stress (physical or emotional), infection, and medications. When this occurs, the dramatic increase in severity (amplitude, violence, or frequency) may be quite alarming to patients and their families. Medications used to treat comorbid conditions (eg, attention-deficit/hyperactivity disorder or obsessive-compulsive disorder), such as stimulants29 and

Table 3. Treatment of Hyperkinetic Movement Disorders

<table>
<thead>
<tr>
<th>Movement Disorder</th>
<th>Medication Class</th>
<th>Medication</th>
<th>Initial Daily Dose, mg</th>
<th>Recommended Maximum Daily Dose, mg</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chorea</td>
<td>Neuroleptic</td>
<td>Haloperidol</td>
<td>0.5</td>
<td>8</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Risperidone</td>
<td>0.5</td>
<td>6</td>
</tr>
<tr>
<td></td>
<td>Dopamine-depleting agent</td>
<td>Tetrabenazine</td>
<td>12.5</td>
<td>75</td>
</tr>
<tr>
<td></td>
<td>Benzodiazepine</td>
<td>Clonazepam</td>
<td>0.5</td>
<td>6</td>
</tr>
<tr>
<td>Myoclonus</td>
<td>Anticonvulsant</td>
<td>Valproic acid</td>
<td>750</td>
<td>Titrate to serum level</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Levetiracetam</td>
<td>500</td>
<td>3000</td>
</tr>
<tr>
<td>Tics</td>
<td>Neuroleptic</td>
<td>Clonidin</td>
<td>0.1</td>
<td>0.6</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Guanfacine hydrochloride</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td></td>
<td>Dopamine-depleting agent</td>
<td>Tetrabenazine</td>
<td>12.5</td>
<td>75</td>
</tr>
<tr>
<td></td>
<td>Antihypertensive</td>
<td>Clonidine hydrochloride</td>
<td>0.5</td>
<td>0.6</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Guanfacine hydrochloride</td>
<td>8</td>
<td>6</td>
</tr>
<tr>
<td>Acute dystonic reaction</td>
<td>Anticholinergic</td>
<td>Benztrapine mesylate</td>
<td>1</td>
<td>6</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Diphenhydramine</td>
<td>25</td>
<td>400</td>
</tr>
</tbody>
</table>

These agents are generally not helpful in short-term treatment but can be given with a neuroleptic that could eventually be discontinued.

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antidepressants,\textsuperscript{30,31} are often reported to exacerbate tics, although a controlled trial did not confirm this.\textsuperscript{32} In the emergency department, tics should be diagnosed, and potential exacerbating factors including psychiatric ones should be identified and removed. Pharmacologic treatment, if needed, can be initiated. Initial treatment for debilitating tics uses a neuroleptic or a dopamine-depleting agent (Table 3). Patients with focal tics may benefit from botulinum toxin injections, but this treatment is not helpful emergently.\textsuperscript{33} Neurologic compromise secondary to tics is uncommon; however, severe tics can cause both compressive neuropathies\textsuperscript{34} and cervical myelopathy.\textsuperscript{35}

**Status Dystonicus**

Patients with primary and secondary dystonia can rarely experience acute worsening with generalized, severe, dystonic spasms called dystonic storm or status dystonicus.\textsuperscript{36} These unremitting dystonic spasms may be life threatening. Reported precipitants for status dystonicus include infection, medication changes, and trauma. The unremitting dystonic spasms may lead to hyperpyrexia, dehydration, respiratory failure, and rhabdomyolysis with renal failure.\textsuperscript{37} Most patients require intensive care unit admission because orally administered agents are insufficient to arrest the dystonic spasms. The usual therapeutic approach is to use combinations of agents including anticholinergics, benzodiazepines, catecholamine-depleting agents, and dopamine receptor blockers.\textsuperscript{37,38} Extreme cases may require general anesthesia or paralyzing agents. Refractory cases may respond to neurosurgical intervention, either pallidotomy or deep brain stimulation of the globus pallidus interna, although this surgery is not an emergent procedure.\textsuperscript{39} Intrathecal baclofen therapy has been used successfully in some patients with status dystonicus.\textsuperscript{40}

**Acute Dystonic Reaction**

Acute dystonic reaction is most commonly seen after exposure to dopamine receptor blockers, both neuroleptics and antiemetics. Dystonia begins within 24 hours of exposure, and 90% of reactions occur within 5 days.\textsuperscript{41} Acute dystonic reactions are less common than tardive dyskinesia or drug-induced parkinsonism, affecting approximately 6% of patients exposed to “typical” neuroleptics and 1% to 2% of those exposed to “atypical” neuroleptics.\textsuperscript{42} Clinical manifestations are diverse, usually affecting the head and neck (Figure 3). Laryngeal dystonia, blepharospasm, cervical dystonia, oculogyric crisis, and focal limb dystonia have all been reported. Acute dystonic reactions are more common in young men,\textsuperscript{43} while tardive dyskinesia and drug-induced parkinsonism are more common in elderly individuals.\textsuperscript{42} Treatment with an intravenous anticholinergic agent, such as benztropine mesylate (1-2 mg) or diphenhydramine (25-50 mg), is very effective (Table 3). Because of the possibility of a recurrence, a short oral course of an anticholinergic (4-7 days) may be necessary.\textsuperscript{42} After an acute dystonic reaction,
patients are at higher risk for future dystonic reactions when exposed to other dopamine receptor blockers.44

Acute Torticollis

Acute nontraumatic torticollis occurs more commonly in children than adults and should be considered a medical emergency. While this disorder is unlikely to be a primary neurologic condition (dystonia), neurologists may be asked to evaluate the patient. Conditions that present with torticollis in children include posterior fossa tumors,45 cervical cord tumors,46 and infection. Acute infectious torticollis, or Grisel syndrome,47 may follow a number of infections including pharyngitis, tonsillitis, mastoiditis, or other infections involving the head or neck. It is secondary to atlantoaxial rotatory subluxation secondary to infection involving the soft tissue surrounding the cervical spine. The majority of cases described have been in patients younger than 13 years.48 Physical examination of the cervical spine. The majority of cases described have been in patients younger than 13 years.48 Physical examination may reveal painful, fixed torticollis that occurred following an infection or recent surgical procedure in the head and neck area.49 Sudeck sign may be present, in which the spurious process of the axis is palpable in the contralateral neck.50 Prompt recognition and treatment decrease the rate of neurologic complications, which nonetheless occur in 15% of cases.51 Initial management of the patient with acute torticollis should include treatment of infection and cervical immobilization followed by imaging of the head and neck with computed tomography or magnetic resonance imaging to look for an underlying space-occupying lesion or orthopedic abnormality.52

CONCLUSIONS

Although movement disorders are often not regarded as emergencies, we have reviewed acute onset of chorea/hemiballismus, dystonic storm, neuroleptic malignant syndrome, altered cognitive states and falling in Parkinson disease, parkinsonism hyperpyrexia syndrome, and serotonin syndrome, all of which can be seen in emergency department and intensive care unit consultation. A systematic approach to these problems that emphasizes the identification of the phenomenology as the key to making the diagnosis and beginning the correct treatment is outlined. These disorders are uncommon and few if any randomized controlled trials have been conducted, so treatment recommendations are based on clinical experience reported in the literature.

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**Correction**

**Error in Byline.** In the Observation titled “Bilateral Pallidal Stimulation for X-Linked Dystonia Parkinsonism” by Wadia et al, published in the August 2010 issue of the Archives (2010;67[8]:1012-1015), an author’s name was listed incorrectly. Dr Torres Diaz’s name should have appeared as Cristina V. Torres Diaz, MD. The article has been corrected online.