Movement Disorders
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Movement Disorder Emergencies

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Abstract: Movement disorders may present acutely, and failure to recognize and exclude important differential diagnoses can result in significant morbidity or mortality. Unfortunately, much of the literature pertaining to this topic is scattered and not easily accessible. This review aims to address this deficit. Movement disorder emergencies are discussed according to their most likely mode of presentation. Diagnostic considerations and early management principles are reviewed, along with appropriate pathophysiology where relevant. © 2004 Movement Disorder Society

Key words: movement disorder; emergency; differential diagnosis; pathophysiology

A movement disorder emergency (MDE) has been defined by Fahn and Frucht[1] as "*. . . any neurological disorder evolving acutely or subacutely, in which the clinical presentation is dominated by a primary movement disorder, and in which failure to accurately diagnose and manage the patient may result in significant morbidity or even mortality*." Much of the literature dealing with MDE is scattered and not readily accessible. In this article we review the clinical presentation, diagnosis, and management of MDE. The pathophysiology of various MDE is discussed only when directly relevant to these issues.

We have divided MDE according to the phenomenology that is likely to dominate the clinical presentation. In each section, diseases that are monophasic and that develop acutely or subacutely are mentioned first, followed by chronic diseases that can manifest acute exacerbations or relapses. For the purposes of this review, only those conditions that are likely to present to the accident and emergency or intensive care unit and for which treatment has to be instituted within hours or days will be considered in detail. Some treatable disorders that present in childhood are also discussed.

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DISORDERS PRESENTING WITH RIGIDITY OR STIFFNESS

MDE that can present with rigidity or stiffness are summarized in Table 1.

Neuroleptic Malignant Syndromes

Clinical Presentation.

Neuroleptic malignant syndrome (NMS) was first reported by Delay and colleagues in 1960.[2] NMS is characterized by the clinical triad of rigidity, dysautonomia, and alteration in mental status. By definition it is caused by neuroleptic drugs. There is generalized rigidity, often accompanied by akinesia, that can be so severe as to render the patient bedbound. Rhabdomyolysis and renal failure may ensue. Swallowing disturbance can be severe leading to aspiration pneumonia. The patient may become mute, and the conscious level may fluctuate. Dysautonomia is often prominent with fever, sweating, tachypnea, tachycardia, and labile blood pressure. With the introduction of the atypical neuroleptic agents, full-blown cases are fortunately rare, occurring in approximately 0.2% of all patients.[3] However, *forme frustes* of the syndrome are not uncommon.

TABLE 1. *Movement disorder emergencies presenting with rigidity*

Pathophysiology	Causes	Comment
Vascular	Spinal arteriovenous malformations	Can mimic stiff man syndrome
Infectious	Tetanus	Rigidity remains between spasms
	Rabies	
Drug-induced	Neuroleptic malignant syndrome	
	Parkinsonism–hyperpyrexia syndrome	
	Serotonin syndrome	
Toxic	Strychnine	Muscle tone normal between spasms
Metabolic	Hypocalcemia	
Inherited	Hyperekplexia	
	Malignant hyperthermia	
Autoimmune	Stiff man syndrome	
Psychiatric	Lethal catatonia	

All age groups can be affected, but it occurs most commonly in young and middle aged adults, particularly males. Symptoms usually develop in the first week after the introduction of a neuroleptic agent for the treatment of an acute psychotic disorder[3] but may also follow an increase in dose or change in the type of neuroleptic drug. On occasions, it may follow a single dose of a neuroleptic agent.[4] The syndrome is usually fully developed within a few days.

Haloperidol is the causative agent in almost half of the published reports of NMS.[3,5,6] Fluphenazine depot preparations and chlorpromazine are also frequent causes. Virtually all classes of D_2 receptor antagonist have been implicated, including prochlorperazine, metoclopramide, droperidol, and promethazine.[7] In most reported cases, the doses given were within the usual therapeutic range.[3] Elderly patients appear to be more susceptible to higher doses.[8] Other factors that have been implicated in the development of NMS include a rapid increase in dose, intramuscular administration, and the concomitant use of lithium.[3,7–9]

Diagnosis.

The diagnosis is made on clinical grounds, based on the presence of clinical features in the setting of a history of exposure to a neuroleptic. Certain laboratory abnormalities support the diagnosis but suffer from the limitation of being nonspecific. Serum creatine phosphokinase (CPK) is usually elevated, and if high, urinary myoglobins and renal function should also be measured to monitor for rhabdomyolysis and secondary renal failure. There may be a leucocytosis.

NMS is commonly initially misdiagnosed as sepsis, which must always be considered and excluded with appropriate investigations. The differential diagnosis also includes lethal catatonia, serotonin syndrome, malignant hyperthermia,[10] acute carbon monoxide poisoning, and salicylate, amphetamine, cocaine, and phencyclidine toxicity.[11]

Treatment.

Treatment involves the immediate cessation of all neuroleptic medications. In mild cases, this may be all that is required. For moderate to severe cases, treatment involves combating dopaminergic blockade with dopamine agonists, most commonly with bromocriptine, and of reducing muscle rigidity by blocking the release of calcium from the muscle sarcoplasmic reticulum using dantrolene. Neither form of treatment has been validated by a controlled trial. There is, nevertheless, a strong incentive to intervene, for death rates as high as 20% have been reported.[3]

Our practice is to give both bromocriptine and dantrolene for severely affected patients. For adults, a single dose of 2.5 mg of bromocriptine is given orally and if this does not cause hypotension, this dose is given three times daily and increased within a day or two to 5 mg t.d.s., with further increases based on the response. Dantrolene is given at a dose of 25 mg daily increasing to 25 mg t.d.s. or higher, depending again on the response. It is available as a powder, which can be dissolved for intravenous use. An intravenous dose of 1 mg/kg is given, and the dose is increased or repeated, according to the response, up to a maximum accumulated dose of 10 mg/kg. The solution is highly alkaline, and care should be taken to avoid extravasation. Oral bromocriptine and dantrolene are usually continued for several weeks. Many patients will benefit from admission to an intensive care unit to aid management of complications such as myoglobinuria, dehydration, aspiration pneumonia, and pulmonary embolism.[5]

Electroconvulsive therapy has also been used[12] and is the treatment of choice in patients where severe psychosis is a continuing problem.[13] It may also be used if drug

therapy fails or if lethal catatonia is suspected as an alternative diagnosis. The value of benzodiazepines such as diazepam and lorazepam is debatable.[3,13] Amantadine may be of benefit.[3,14] Apomorphine may be useful if oral administration of medication is difficult, but pretreatment with domperidone[15] is required. Domperidone can be administered rectally.[16]

One-third or more of patients relapse with reintroduction of neuroleptic therapy. At least 2 weeks should be allowed to pass after full resolution of symptoms before this reinstatement is attempted.[8] It is common practice to switch to an atypical neuroleptic such as clozapine, olanzapine, or quetiapine,[17] although NMS has also been reported in association with these drugs.[18] The concurrent use of lithium should be avoided if possible.[7]

Lethal Catatonia

Clinical Features.

Lethal catatonia was first described by Calmeil in 1832 (see Mann et al., 1986).[19] Catatonia refers to the adoption of fixed abnormal postures (catalepsy) in association with major behavioral abnormalities. Typically, the patient becomes rigid and mute, with periods of intense agitation and bizarre, repetitive movements. This condition occurs in the setting of a major psychiatric disturbance with labile mood, insomnia, anorexia, increasingly disorganized thought processes, auditory and visual hallucinations, as well as bizarre delusions. Violence is occasionally a feature, and there may be unprovoked assaults and attempts at suicide. Eventually, high fever, tachycardia, and fluctuations in blood pressure develop and can be followed after several days by exhaustion, coma, cardiac arrest, and death.

Diagnosis.

With the advent of neuroleptic drugs, lethal catatonia has become very uncommon. That it is a clinical entity distinct from NMS is supported by its occurrence in the preneuroleptic era. It differs from NMS in the severity of the behavioral abnormalities in the early stages. In its advanced stages, it becomes indistinguishable from NMS. It usually occurs in the setting of schizophrenia but may also follow infections such as meningitis and encephalitis, head trauma, drug intoxication, and metabolic disturbances such as uremia, porphyria, and Wernicke's encephalopathy.[19,20]

Treatment.

The treatment of choice is electroconvulsive therapy (ECT),[20] which is tolerated even in severely debilitated patients. Agitation may be treated with benzodiazepines. Because ECT has also been reported to be of benefit in NMS, if uncertainty regarding diagnosis exists, this option is the treatment of choice in the severely ill patient.

Serotonin Syndrome

Clinical Features.

Drugs that enhance serotonergic neurotransmission may also produce a syndrome of severe rigidity, dysautonomia, and alteration in mental status. The symptoms of serotonin syndrome (SS) evolve more rapidly than is the case with NMS (hours rather than days) and are more likely to include myoclonus, hyperreflexia, and seizures.[1,21,22] Like NMS, there is a wide spectrum of severity,[23] with some patients presenting with only mild myoclonus and agitation.[24] Severe cases can progress to rhabdomyolysis, myoglobinuria, and renal failure, as well as severe metabolic acidosis, disseminated intravascular coagulation, and adult respiratory distress syndrome.

Diagnosis.

SS is diagnosed on the basis of the clinical findings described in a patient with a history of exposure to single serotonergic drugs or, more commonly, exposure to two or more drugs with serotonergic properties.[21,22] (see Table 2). The distinction between SS and NMS can be difficult if there is a history of recent exposure to both classes of drugs, but as already alluded to, the presence

TABLE 2. *Drugs causing the serotonin syndrome*

Pharmacologic categories	Drugs
Inhibitors of serotonin reuptake	SSRI, tricyclic antidepressants, dextromethorphan, dexamphetamine, cocaine, meperidine, opiates (except morphine)
Inhibitors of serotonin metabolism	MAO-B inhibitors (selegiline), MAO inhibitor antidepressants
Agents increasing serotonin synthesis	L-tryptophan
Enhancers of serotonin release	MDMA (ecstasy), amphetamines, cocaine, fenfluramine
Serotonin agonists	Sumatriptan, ergotamines, buspirone
Nonspecific enhancers of serotonin activity	Lithium, ECT

See Lane and Baldwin.[22]
SSRI, selective serotonin reuptake inhibitor; MAO-B, monoamine oxidase-B; ECT, electroconvulsive therapy.

of myoclonus, seizures, or hyperreflexia favors the former.

Laboratory data are unhelpful in establishing the diagnosis of SS.[21] Elevated CPK or transaminases, leucocytosis, and a reduction in serum bicarbonate level can be present but are nonspecific.

Treatment.

Most cases can be managed simply by discontinuation of the offending drug(s), fluid administration, and cardiac monitoring.[21] In severe cases, cyproheptadine, an antihistamine and serotonin antagonist, may be administered. An initial dose of 4 to 8 mg orally should be given, with a repeat dose in 2 hours. If no improvement occurs, it is discontinued, but if a response occurs it should be maintained up to a dose of 32 mg daily in four divided doses until resolution of the symptoms.[25] Gillman recommends 50 to 100 mg of chlorpromazine by IM injection as a starting dose in severe cases,[26] but this strategy is not an option if NMS is part of the differential diagnosis. Benzodiazepines have also been used.[27]

Malignant Hyperthermia

Malignant hyperthermia is a rare syndrome characterized by the rapid onset of fever, fluctuations in blood pressure, hyperkalemia, and metabolic acidosis, followed by severe muscle rigidity and secondary rhabdomyolysis. The majority of cases are triggered by various anesthetic compounds, including halogenated inhalational agents and depolarizing muscle relaxants. The clinical syndrome results from uncontrolled calcium flux across skeletal muscle membrane. In over 50% of families, there is linkage of the autosomal dominant trait to a gene encoding the skeletal muscle ryanodine receptor.[28] Urgent treatment with the muscle relaxant dantrolene is highly effective and needs to be combined with discontinuation of the triggering agents and correction of acidosis and electrolyte abnormalities. Even with these measures, mortality is around 10%.[29]

Hypocalcemia

Tetanic muscle spasms can develop acutely secondary to hypocalcemia, the severity of which varies with the magnitude and rapidity of the fall in serum calcium.[30] It occurs most commonly in the setting of thyroid or parathyroid surgery, rhabdomyolysis, hypomagnesemia, malignancy, chronic renal failure, pancreatitis, and septic shock. There are also reports of hypocalcemia and tetany developing after plasma exchange.[31] Prolongation of the QT interval with attendant risk of arrhythmia should be investigated. Rarely, there is associated laryngospasm.[30] Intravenous administration of calcium is effective therapy, together with treatment of the underlying cause.

Infective and Toxic Causes of Rigidity or Stiffness

Tetanus.

In tetanus, uncontrolled disinhibited spinal cord efferent discharges lead to profound muscular rigidity and spasms, which clinically can resemble an epileptic seizure. The exotoxin tetanospasmin, produced by *Clostridium tetani*, prevents glycine and GABA release in the spinal cord.[32,33] Muscle spasms may be triggered by touch, visual, auditory, or emotional stimuli[32] and be associated with aspiration, respiratory failure, and autonomic instability[34] with hypertension and tachycardia, alternating with hypotension, bradycardia, and recurrent cardiac arrest. Localized tetanus may cause focal limb stiffness, trismus, or spasms in a single limb.[35]

In a patient with muscle spasms and a history of a recent wound, the diagnosis may be clear. Unlike strychnine poisoning (see below), in tetanus, there is sustained muscle rigidity between spasms. There is no diagnostic test, but a measurable titre of anti-tetanus antibody effectively excludes the diagnosis. It is important to establish the state of previous tetanus immunization.

Management in the intensive care unit should involve neutralization of the unbound toxin with tetanus immune globulin, removal of the source of infection, and treatment of the organism with antibiotics. Sedation with γ-aminobutyric acid (GABA) agonists such as benzodiazepines or phenobarbitone and avoidance of excessive sensory stimulation are important adjunctive therapies.[32] Intravenous lorazepam (up to 80 mg daily in 2-mg increments) or diazepam (up to 500 mg per day) have been used, as have baclofen, methocarbamol (3–4 g every 6 hours intravenously or by feeding tube), dantrolene (1–2 mg/kg every 4 hours),[33] and intrathecal baclofen.[36–38] After recovery, full tetanus immunization should be instituted, as exposure to the toxin does not confer immunity.

Strychnine toxicity.

Strychnine blocks spinal and brainstem inhibitory interneurons, resulting in marked muscle rigidity and changes in mental status. It is still found in pesticides, some traditional herbal remedies,[39] and adulterated heroin. Initially, it presents with a hyperalert state and confusion, soon followed by hyperreflexia with rigidity, severe muscle spasms, and opisthotonus, often triggered by external stimuli. Contraction of the facial muscles may cause risus sardonicus. Unlike tetanus, muscle tone normalizes between spasms. The patient remains fully

conscious. Sustained spasms cause respiratory hypoventilation and muscle necrosis, with death ensuing rapidly from asphyxia and cardiac arrest.

A high index of suspicion is needed to make a timely diagnosis. Biochemical analysis of gastric aspirates may confirm strychnine ingestion, but treatment with respiratory support, activated charcoal, benzodiazepines, and barbiturates needs to be instituted immediately. These drugs may increase spinal neuronal inhibition through their GABA agonist effects and reduce the spasms. Although the prognosis is poor, recovery is possible.[40]

Rabies.

Rabies is the major virus of the *Lyssavirus* genus. Although regarded as a "third world" disease, it does occur in developed countries where its rarity means the diagnosis is usually made at post mortem.[41] The typical presentation is with pain or stiffness at the site of the infecting bite. High fever, agitation, hallucinations, violent behavior, and autonomic instability follow. Diaphragmatic and laryngeal spasm, the latter particularly when drinking, cause hydrophobia. Periods of hyperexcitability are interspersed with lucid intervals. The 5- to 15-second spasms may be associated with facial grimacing, extension of the neck and back, and opisthotonus.[42] This "furious" phase is followed by loss of all central and peripheral neurological function known as "paralytic" rabies.

There may be no history of animal exposure or bite. The incubation period is said to be usually 30 to 90 days with almost all cases occurring within 1 year of the bite. However, incubation periods of 5 or more years have been described.[43] The diagnosis can be made in life by the demonstration of antigen in nerves from a nape-of-neck biopsy. Death is inevitable. The major reason for prompt diagnosis is to ensure there is provision of post-exposure prophylaxis of contacts, although human to human transmission usually only occurs after organ transplantation.

Stiff Man Syndrome.

Patients with this rare disorder may present as an emergency with severe pain and spasms of the lumbar paraspinal muscles and lower limbs. Spinal cord compression is often suspected, but there is no weakness, sensory loss, or bladder disturbance. The affected muscles are rock-hard to palpation. An exaggerated lumbar lordosis is often present, producing a characteristic horizontal crease in the small of the back. Muscle spasms can occur spontaneously or be provoked by noise or movement. The spasms may be so forceful as to produce femoral fractures, joint subluxations, and even herniation of abdominal contents.[44] Such spasms can occur in salvoes associated with life-threatening autonomic dysfunction. Sudden death occurs in approximately 10% of patients with stiff man syndrome (SMS) and is due in most cases to acute autonomic failure.[45]

The diagnosis and management of SMS has been reviewed recently in detail elsewhere.[44] In the event of an acute presentation, exclusion of spinal pathological conditions that can cause secondary SMS should be considered.

Hyperekplexia.

Sudden death is reported in infants with this autosomal dominant syndrome of exaggerated startle reflex and hypertonicity. Apnea and aspiration may result from the pharyngeal incoordination that results from being handled. Clinically, there is neck retraction and body tonic flexion that can be easily stimulated by nose tapping. Many affected families show mutations in the inhibitory glycine receptor (GLRA1) gene, with resultant increase in pontomedullary reticular excitability and abnormal spinal cord reciprocal inhibition. It is often misdiagnosed as epilepsy. Treatment with clonazepam, a GABA agonist, is highly effective, but the common nonbenzodiazepine anticonvulsants are ineffective. The hypertonicity and hyperreflexia tend to be transient, diminishing after the first year of life,[46] although rarely familial hyperekplexia and spastic paraparesis can cosegregate.[47] Forced flexion of the head toward the trunk and legs may be life-saving when prolonged rigidity impedes respiration.[48]

DISORDERS PRESENTING WITH PARKINSONISM

MDE that can present with parkinsonism are summarized in Table 3.

Parkinsonism–Hyperpyrexia Syndrome

Lowering the dose or withdrawing dopaminergic drugs in patients with Parkinson's disease can cause a syndrome indistinguishable from NMS, called the parkinsonism–hyperpyrexia syndrome.[49–51] Parkinsonism–hyperpyrexia can also develop after withdrawal of nondopaminergic drugs used to treat Parkinson's disease such as amantadine or anticholinergics and has also been reported in association with the use of tolcapone, although not entacapone. Treatment consists of reinstituting dopaminergic therapy and provision of supportive care until recovery takes place, which may take weeks. If swallowing is impaired, the drugs may be given by means of a nasogastric tube. Parenteral apomorphine has also been used.[52] Prior

TABLE 3. *Movement disorder emergencies causing parkinsonism*

Pathophysiology	Causes	Comment
Vascular and structural	Basal ganglia stroke (esp. involving globus pallidus)	
	Midbrain lesions	
	Hydrocephalus	
Infectious	Encephalitis lethargica	
	Other viral encephalitis (esp. Japanese B)	
	Mycoplasma	
Drug induced	Parkinsonism–hyperpyrexia syndrome	Caused by dopaminergic drug withdrawal
	Chemotherapy	
	Amphotericin B	
Toxic	Carbon monoxide	
	Methanol	
	Cyanide	
	Organophosphate poisoning	
	MPTP	
Metabolic	Central pontine myelinosis	Usually associated with encephalopathy
Inherited	Rapid-onset dystonia–parkinsonism	
Psychiatric	Neuroleptic-induced	

MPTP, 1-methyl-4-phenyl-1,2,3,6-tetrahydropyridine.

loading with rectal domperidone can be used to prevent vomiting.[16]

Acute Severe de novo Parkinsonism

Acute severe parkinsonism is rare. It can be seen after carbon monoxide[53] or cyanide poisoning[54,55] and (rarely) after viral encephalitis,[56] including encephalitis lethargica. MPTP was a cause of acute parkinsonism in the past.[57] Destruction of the putamen has been reported with methanol poisoning,[58] and reduced striatal metabolism with acute reversible parkinsonism has been reported in accidental petroleum product ingestion.[59] Zeidler and colleagues drew attention to two cases of obstructive hydrocephalus and parkinsonism, in association with Parinaud's syndrome, where the parkinsonian features resolved immediately after shunting.[60] There are also several reports of acute transient parkinsonism developing after organophosphate poisoning or exposure,[61,62] which responded to amantadine and biperiden.[63] Cytotoxic therapy, including cyclophosphamide and high-dose cytosine arabinoside before bone marrow transplantation, can cause an acute encephalopathy with parkinsonian features.[64] Amphotericin B has also been implicated in acute parkinsonism.[65] Both high-dose methylprednisolone[66] and L-dopa have been reported to be effective in chemotherapy-induced parkinsonism.[67] A rare form of dominantly transmitted rapid-onset dystonia–parkinsonism linked to chromosome 19 that can develop over hours to days has been reported.[68]

Encephalitis Lethargica

Initially described by von Economo in 1917, sporadic cases of encephalitis lethargica continue to occur.[69,70] Suspected by some to be secondary to the influenza virus at the time, this suspicion is now thought to be unlikely, with autoantibodies reactive against basal ganglia antigens recently being demonstrated in cerebrospinal fluid (CSF).[71,72] It presents with sleep disturbance (somnolence, sleep inversion, insomnia), extrapyramidal disturbance (parkinsonism, rest tremor), dyskinesia (oculogyric crises, dystonia, tics, stereotypies), and/or ophthalmoplegia. Von Economo also noted that neuropsychiatric features (catatonia, obsessive–compulsive disorder, and mutism) were common in survivors. Dramatic responses to intravenous methylprednisolone treatment have been reported and lend support to an autoimmune hypothesis.[73]

DISORDERS PRESENTING WITH DYSTONIA

MDE that can present with dystonia are summarized in Table 4.

Acute Dystonic Reactions Secondary to Drugs

Drugs are the most common cause of acute focal dystonia. This may be life-threatening if the airway or breathing are compromised. Oculogyric crises, laryngeal dystonia, blepharospasm, torticollis, trismus, dysarthria, and dystonia have all been described, usually within 24 hours of commencing the drug.[74] Neuroleptics and dopamine blocking antiemetics are by far the most commonly implicated medications (see Table 5). Acute dystonic reactions are most often seen in young males.[75] As a rule, neuroleptic drugs are more likely to cause acute dystonic reactions in younger patients and tardive dyskinesia or parkinsonism in older patients.[76]

Acute dystonic reactions are usually self-limiting but can be distressing. The dystonia is usually reversed

TABLE 4. *Movement disorder emergencies causing dystonia*

Pathophysiology	Causes	Comment
Vascular and structural	Basal ganglia stroke (esp. putamen)	
	Posterior fossa and cervical cord lesions	Childhood disorder
	Atlantoaxial subluxation	
Infectious	Retropharyngeal abscess	Childhood disorder
	Encephalitis lethargica	
Drug induced	D2 antagonists (neuroleptic and antiemetics)	
	Withdrawal of intrathecal baclofen	Can be inadvertent
Toxic	Carbon monoxide	
	Methanol	
	Cyanide	
	Organophosphate poisoning	
Metabolic	Leigh's disease	Associated with encephalopathy
	Biotin-responsive basal ganglia disease	Associated with encephalopathy
	Aminoacidurias and urea cycle disorders	Associated with encephalopathy and ataxia
Inherited	Disorders of catecholamine metabolism	
Degenerative	Laryngeal dystonia in multiple system atrophy	
Psychiatric	Pseudodystonia	
Idiopathic	Adductor laryngeal breathing dystonia	
	Paroxsymal dystonia	
	Status dystonicus	Can occur in setting of any generalized dystonia

within minutes by parenteral anticholinergic therapy such as benztropine 1 to 2 mg IV or IMI, repeated in 20 minutes if not effective. It is common for the dystonia to return as the effect of the parenteral medication wears off and for this reason oral anticholinergic therapy should be continued for 4 to 7 days in a tapering dose after acute therapy.[75,77] Patients should be warned to avoid the precipitating drug and that they are at higher than average risk if exposed to other drugs that are associated with dystonic reactions.[78]

Adductor Laryngeal Breathing Dystonia (Gerhardt's Syndrome)

Adductor laryngeal breathing dystonia (ALBD) is a rare task-specific dystonia in which the vocal cords undergo adductor spasm during inspiration but not other activities such as speaking.[79,80] It is the converse of spasmodic dysphonia in which the vocal cords adduct involuntarily during speech but function normally during breathing. The clinical presentation is with severe stridor, and there is a risk of life-threatening respiratory obstruction. It can occur sporadically and in isolation, in which case a misdiagnosis of idiopathic bilateral vocal cord paralysis may be made. It may be the only manifestation of a drug-induced dystonia. In approximately half of cases, other manifestations of dystonia are present. Botulinum toxin injections into the overactive thyroarytenoid muscles is effective treatment in most patients and may avert the need for tracheostomy.

TABLE 5. *Drug-induced movement disorder emergencies*

Movement	Drugs
Chorea	*Common:* L-Dopa
	Uncommon: Phenytoin, carbemazepine, tricyclic antidepressants, estrogen, cocaine, baclofen, trazadone, anticholinergics
Myoclonus	*Common:* SSRIs
	Uncommon: Tricyclic antidepressants, lithium, MAO inhibitors, carbamazepine, penicillin and cephalosporin antibiotics, cocaine, opiates, amantadine L-dopa, bromocriptine
Tremor	*Common:* Neuroleptics, valproate, alcohol, sympathomimetics
	Uncommon: Opiates, immunosuppressives, hypoglycemic agents, antibiotic and antiviral agents, anticonvulsants, antiarrhythmics, antidepressants, xanthines, corticosteroids, thyroxine, amiodarone
Dystonia	*Common:* Neuroleptics (including anti-emetics), L-dopa
	Uncommon: Dopamine agonists, phenytoin, carbemazepine, SSRI and tricyclic antidepressants, cocaine
Parkinsonism	*Common:* Neuroleptics (including anti-emetics)
	Uncommon: Flunarizine, cinnarazine, tricyclic antidepressant, tacrine, chemotherapeutic agents, carbamazepine, phenytoin, valproate, lamotrigene, MPTP
Tics	*Uncommon:* Carbemazepine, phenytoin, dexamphetamine, methylphenidate, cocaine
Akathisia	*Common:* Neuroleptics
	Uncommon: Tricyclic antidepressants, SSRIs, calcium channel antagonists

SSRIs, selective serotonin reuptake inhibitors; MAO, monoamine oxidase; MPTP, 1-methyl-4-phenyl-1,2,3,6-tetrahydropyridine.

Acute Torticollis in Childhood

Sudden onset of torticollis in a child is a sign that needs to be assessed with great care as several serious conditions may present in this way.[81] These conditions include posterior fossa and cervical cord tumors, cervical syrinx, colloid cysts of the third ventricle, and bone tumors. Torticollis may result from infections such as tonsillitis and retropharyngeal abscesses.[82] Torticollis has also been attributed to atlantoaxial rotatory subluxation, but a recent study has cast doubt on this finding.[83] Depending on the clinical situation, the child with torticollis may need neuroimaging of the brain and cervical spine, antibiotics, immobilization of the neck and surgical review.

Status Dystonicus

Patients with both primary and secondary dystonia can rarely develop a life-threatening disorder of unremitting, severe generalized dystonic spasms. This has been termed status dystonicus or dystonic storm. Status dystonicus can arise after intercurrent infection, alteration in medications, or for no obvious reason. Patients are at risk of respiratory or airway compromise, aspiration pneumonia, or renal failure from secondary rhabdomyolysis. Some succumb from exhaustion. The response to conventional drug treatment is often poor. Oral benzodiazepines, levodopa, benzhexol, tetrabenazine, pimozide, haloperidol, baclofen, propanolol, and anti-epileptic agents such as carbamazepine have been used with limited benefit.[84,85] Triple therapy with oral tetrabenazine, high-dose benzhexol, and a dopamine blocker such as haloperidol was found to be useful in a few cases by Manji and coworkers.[85] Sedation and paralysis with ventilation in the setting of an ICU is sometimes necessary to prevent the secondary complications mentioned above. Intrathecal baclofen has been used with some success,[86,87] and if medical therapy fails, functional surgery such as bilateral ventrolateral thalamotomy,[88] unilateral pallidotomy,[89] and bilateral pallidal stimulation[90] can be effective.

Metabolic Causes of Subacute Dystonia

Acute or subacute dystonia, often associated with an encephalopathy and/or striatal necrosis, can develop in the setting of an acute infective insult (e.g., viral encephalitis, including encephalitis lethargica and mycoplasma pneumoniae) or in several different metabolic disorders (e.g., Leigh's disease, mitochondrial cytopathies), most of which are not treatable. The possibility of an inherited disorder of catecholamine production, should always be considered as this may improve with levodopa therapy. The CSF in such cases may show abnormalities of catecholamine metabolites and pterins. A biotin-responsive basal ganglia disease has been described in Saudi Arabia.[91] It commences with a subacute encephalopathy and progresses without treatment to severe rigidity, dystonia, and quadriparesis. If recognized early, treatment with biotin 5 to 10 mg/kg per day can reverse the neurological abnormalities.

Sudden Withdrawal of Intrathecal Baclofen

Continuous intrathecal baclofen delivered by means of a permanent catheter is increasingly being used in the treatment of severe dystonia and spasticity. A life-threatening syndrome similar to NMS can be precipitated by the inadvertent or sudden withdrawal of this drug, such as if the catheter tip becomes dislodged. High fever, altered mental status, and profound muscular rigidity that may progress to rhabdomyolysis has been described.[92] Treatment includes giving high doses (up to 120 mg/day in divided doses) of oral or enteral baclofen. Benzodiazepines and dantrolene have also been used.[92,93]

Laryngeal Dystonia in Multiple System Atrophy

The onset of stridor, initially at night but later throughout the day, is a grave symptom in the setting of atypical parkinsonism; the mean survival was less than 1 year in those not undergoing tracheostomy in one study.[94] It results from failure of the vocal cords to abduct normally and occurs in up to 39% of patients with a diagnosis of multiple system atrophy.[95–97] There appears to be an association with dysphagia and hoarseness, and the incidence of sudden death is much greater than in the nonstridor group. Treatment markedly decreases the relative risk of death and is the only independent risk factor for survival.[96] Assessment with polysomnography, arterial blood gases, and laryngoscopy to confirm the diagnosis is recommended. Treatment options include correction of the stridor with (nasal) continuous positive airway pressure where possible, botulinum toxin injections to the laryngeal adductors, and tracheostomy when the former are ineffective.[94]

DISORDERS PRESENTING WITH CHOREA OR BALLISM

MDE that can present with chorea or ballism are summarized in Table 6.

Severe Parkinsonian Dyskinesia

Patients with Parkinson's disease can present with a sudden severe exacerbation of dopa-induced dyskinesia, causing exhaustion and, rarely, rhabdomyolysis. The addition of a long-acting dopamine agonist or an underly-

TABLE 6. *Movement disorder emergencies causing chorea or ballism*

Pathophysiology	Causes	Comment
Vascular and structural	Basal ganglia stroke (esp. subthalamic nucleus or caudate)	
Infectious	Encephalitis	Children and young adults
Drug induced	*See Table 5*	
Metabolic	Hyperglycemia	Associated with MRI abnormalities
	Hypoglycemia	Autonomic symptoms may be absent or minimal
	Leigh's disease	Associated with encephalopathy
Inherited	Disorders of catecholamine metabolism	
Degenerative	Parkinson's disease	Medication induced
Autoimmune	Sydenham's chorea	
	Antiphospholipid syndrome	
	Systemic lupus erythematosus	
	Hashimoto's encephalopathy	Associated with encephalopathy and myoclonus

MRI, magnetic-resonance imaging.

ing infection were the precipitants in a case series described by Factor and Molho.[98] A similar syndrome has been reported in patients treated with catechol-methyl-0-transferase (COMT) inhibitors.[99] Associated respiratory dyskinesia may cause dyspnea, which can be distressing to the patient. Management includes exclusion of infection, careful reduction in dopaminergic medication, and the addition of amantadine, which lessens dyskinesia in some, although not all, patients.

Acute Generalized Chorea

Chorea usually develops gradually and is rarely disabling or life-threatening, especially early in the course. Occasionally, the onset can be acute and severe. Cerebral imaging, with computed tomography and magnetic resonance imaging (MRI) are recommended to exclude cerebral hemorrhage or infarction, as well as to examine the basal ganglia for signal abnormalities. Biochemical testing is important to exclude hyper- or hypoglycemia. Serological testing is required to exclude autoimmune disease.

Postinfectious chorea following Group A streptococcal infection (Sydenham's chorea) can present acutely or subacutely and, rarely, can cause continuous incapacitating ballistic movements (chorea insatiens).[100] Chorea associated with systemic lupus erythematosus or primary antiphospholipid syndrome (APLS) rarely presents as an emergency. It may respond to immunotherapy. Anticoagulation is indicated if associated with thromboembolic manifestations, especially in the setting of the catastrophic APLS.[101]

Acute Hemichorea or Hemiballism

Acute hemichorea or hemiballism most commonly results from infarction or hemorrhage of the basal ganglia, usually in the contralateral subthalamic nucleus, caudate nucleus, or putamen.[102] Patients with this problem should undergo urgent imaging. In children, a rare cause is moyamoya disease, which may be missed on routine MRI.[103,104] The chorea may be violent and distressing, although in most cases, it subsides spontaneously over the course of a few weeks.

Treatment of Acute Chorea.

Mild chorea often does not require treatment. Where chorea and ballismus are of sufficient severity to cause functional disability and exhaustion, several different approaches have been advocated.[1,105] Combining a benzodiazepine with either haloperidol, olanzapine,[106] or tetrabenazine,[107] titrated over days to maintain a balance between control of the movement disorder and side effects such as sedation can be effective. An attempt should be made to periodically wean symptomatic treatment as chorea can often fluctuate in severity or resolve spontaneously.

DISORDERS PRESENTING WITH MYOCLONUS

MDE that can present with myoclonus are summarized in Table 7. Myoclonus and asterixis are most commonly seen in the setting of a metabolic or toxic encephalopathy such as renal or liver failure. However, there are several other causes.

Drug-Related

Myoclonus occurring as part of the serotonin syndrome has been discussed already. Drug-induced myoclonus can also occur with several other medications (see Table 5). Opiate toxicity causing myoclonus is a relatively common problem in chronic oral administration for malignancy.[108] This condition is often generalized and can be periodic or associated with rigidity. It may respond to naloxone or benzodiazepines. Opiate-withdrawal myoclonus may be

TABLE 7. *Movement disorder emergencies causing myoclonus*

Pathophysiology	Causes	Comment
Vascular and Structural	Brainstem and thalamic infarction (esp. VL, VPL nuclei)	Negative myoclonus/asterixis
Infective	Focal encephalitis	
Drug induced	Serotinergic drugs	Complex drug interactions
	Opiate Induced	May respond to naloxone or benzodiazepines
	Opiate withdrawal	Responds to benzodiazepines and *not* naloxone
	Lithium	Cortical action myoclonus
	Tricyclic antidepressants	Especially serotonin syndrome
	Imepenem, cefuroxime	
Epilepsia partialis continua	Subdural haemorrhage, cortical sinus thrombosis, anti-Hu paraneoplastic encephalitis	

VL, ventrolateral; VPL, ventroposterolateral.

stimulus-sensitive, responding to benzodiazepines, but not to naloxone. The intrathecal administration of opiates may also precipitate myoclonus. Lithium can cause cortical action myoclonus at both therapeutic and toxic doses,[109] as can tricyclic antidepressants[110] and antibiotics such as imipenem[111] and cefuroxime.[112]

Focal Cerebral Pathology

Involuntary movements, including myoclonus, are reported in patients with basilar artery occlusion and brainstem ischemia; early recognition may allow prompt treatment.[113] The acute onset of focal limb asterixis (negative myoclonus) can occur secondary to focal cerebral pathology such as cerebral infarction.[114] The thalamus is the most commonly implicated site of focal pathology where both unilateral and bilateral asterixis have been reported.[115]

Epilepsia partialis continua is defined clinically as a syndrome of continuous focal jerking of a body part, usually localized to a distal limb, occurring over hours, days, or even years.[116] Focal encephalitis, tuberculosis, anti-Hu paraneoplastic encephalitis,[117] and vascular lesions, including subdural hemorrhage and cortical sinus thrombosis are some causes.[118]

DISORDERS WITH PSYCHIATRIC PRESENTATIONS

Acute Psychosis in Parkinson's Disease

Acute psychosis in parkinsonism most commonly occurs in the setting of a dementia, which may be mild and unnoticed. The psychosis may be triggered by a change of environment (such as a hospital admission for an elective procedure, or holiday), urinary or chest infection, and, most commonly, recent introduction of drugs, particularly dopamine agonists, anticholinergic drugs, amantadine, or a COMT inhibitor. General metabolic disorders causing delirium need to be excluded and the family questioned about preexisting psychiatric disorders such as depression. Characteristically, the patient develops visual hallucinations and persecutory delusions and is agitated and irrational. Consciousness is usually well preserved.

Patients with mild symptoms and care support are best managed at home. If hospitalization is necessary, joint management by the neurology and psychiatry teams is usually required. The use of restraints and cot sides to limit their movements should be avoided because of the risk of worsening agitation and provoking violence or self-injury. Ideally, single nursing in a quiet room should be provided, with the mattress on the floor. Friedman and Factor[119] recommend the stepwise withdrawal of anticholinergics, followed by selegiline, dopamine agonists, amantadine, and COMT inhibitors. If the patient remains agitated, it may be necessary to lower the dose of levodopa to the point where the patient's mobility is curtailed. Tapering of drugs sequentially over days is recommended to reduce the risk of provoking parkinsonism–hyperpyrexia syndrome.

Most patients settle on this regimen. In an aggressive patient posing an immediate danger to him or herself or to others, a benzodiazepine such as diazepam may be given orally or parenterally.[120] Potent major tranquilizers such as haloperidol can cause rapid worsening of the parkinsonism and should only be used for a few days, and then in low dosage, in the acutely disturbed patient where benzodiazepines have proved inadequate. To gain control of the psychosis in the longer term (over the next few weeks), it is usually necessary to introduce an atypical neuroleptic agent. These agents rapidly dissociate from the dopamine D2 receptor,[121] have a shorter duration of effect,[122] and are less likely to cause extrapyramidal side effects or neuroleptic malignant syndrome than traditional neuroleptics. The doses used are approximately one-tenth that used in schizophrenia.[122] Clozapine has been shown clearly to have a reduced risk of extrapyramidal side effects but at the cost of a small but not insignificant risk of serious hematological and car-

diac toxicity. Quetiapine is a good alternative and rarely causes clinically significant deterioration of parkinsonism.[123] Olanzapine or risperidone can also be used but both occasionally do cause worsening of extrapyramidal function or even NMS.

Psychogenic Movement Disorders

Psychogenic movement disorders are often florid and of sudden onset and, therefore, can present to the emergency department. Variability in the phenomenology and distribution of involuntary movements, reduction or, conversely, entrainment of involuntary movements during mental or motor distraction are pointers toward the diagnosis.[124–126] The importance in considering this diagnosis is the potential avoidance of unnecessary investigations. However, confident diagnosis in the acute setting can be difficult, and exclusion of underlying pathology may be necessary in combination with more prolonged observation of the patient.

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