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Acute Movement Disorders in Children: Emergency Department Presentation and Evaluation

Abnormal movements, outside of seizure and ataxia, are an uncommon chief complaint among children presenting to the emergency department. A working knowledge of movement disorder phenomenology, etiology, differential diagnosis, and associated potentially life-threatening conditions is essential for emergency physicians. Each movement disorder and its clinical considerations will be discussed in detail in the article.

— Ann M. Dietrich, MD, Editor

Epidemiology

As a group, acute movement disorders (*see Table 1*) in previously healthy children are rare. Some epidemiologic data exist for the more common disorders such as tic disorders, dystonia, and tremor; however, the remaining disorders have not been studied in this context. Even in movement disorder specialty centers, many disorders (such as myoclonus and chorea) account for less than 10% of the patient population.[1] While one is more likely to encounter abnormal movements due to seizure or ataxia in the emergency department, it is nonetheless important to know how to distinguish a true movement disorder.

Etiology

The potential etiologies of an acute movement disorder in a previously healthy and developmentally normal child are numerous, but can be divided into a few major categories: infectious, inflammatory, autoimmune, structural, toxic/metabolic/drug-induced (also known as extrapyramidal adverse effects), and psychogenic (as a diagnosis of exclusion). (*See Table 2.*)

Drug-induced movements are important to consider in the workup of a new movement disorder. Movement disorders are a known potential adverse effect to antipsychotic medications in particular, and the use of these medications among children has increased in recent years.[2,3] The prevalence of youth using more than one psychotropic medication has also increased.[4]

Definition and Pathophysiology

Abnormal movements can result from an insult to components of the cerebral cortex, basal ganglia, cerebellum, or spinal cord.

The planning and execution of movements start in the motor cortex. In children, damage to the motor cortex may result in paresis (as in adults with stroke), but may also manifest as seizure, or abnormal paroxysmal activity resulting from excessive synchronous discharges from cortical neurons. Seizure is the most common cause for life-threatening abnormal movements in children, with an estimated 1% of children experiencing an afebrile seizure before the age of 14.[1] The basal ganglia are the deep nuclei of the brain that control

Executive Summary

- Tics are repeated, individually recognizable (stereotyped), intermittent movements or movement fragments that are almost always briefly suppressible and are usually associated with awareness of an urge to perform the movement.
- While much debate exists about PANDAS, it may be beneficial to test a child with a new-onset OCD and/or tic disorder for an underlying group A streptococcal infection, which, if identified, requires treatment.
- One ominous condition that can present with stereotypies is Rett syndrome, a genetically based developmental regression that most commonly affects girls and is classically associated with a "hand-wringing" stereotypy.
- Drug-induced dystonic reactions (DIDR) are well-documented in the literature, and most commonly result after starting a therapeutic dose of a neuroleptic (e.g., haloperidol) or antiemetic medication (e.g., ondansetron, metoclopramide), although many other drug types have been associated. The majority of DIDRs occur within 5 days of starting a new drug.

the direction, speed, and amplitude of movement. In general, neurons of the basal ganglia use GABA as a neurotransmitter, leading to a system of inhibition and disinhibition to control movement. The facilitation of movement is largely dominated by dopamine, which becomes important in conditions such as Parkinson's disease (in which a depletion of dopamine leads to bradykinesia). The coordination and precision of movements are among the many functions of the cerebellum. Children with cerebellar pathology usually present with ataxia, usually manifested as disturbed gait. The most common cause of ataxia in a child is acute cerebellar ataxia (accounting for up to 50% of cases), which is thought to result from antiviral or autoimmune antibodies reacting against cerebellar tissues after a viral infection such as varicella or Epstein-Barr virus.[5] However, it remains a diagnosis of exclusion after other life-threatening causes are ruled out. (*See Table 2.*)

Pathology of any of the motor pathways in these structures can result in abnormal movements characterized as either excessive involuntary movements that interfere with normal functioning (hyperkinetic or dyskinetic) or suppression or slowing of voluntary movements (hypokinetic or bradykinetic).

Clinical Features

The evaluation of movement disorders requires both a detailed history and neurologic examination to document the movements (if they are occurring at the time of presentation) and to identify any other abnormalities.

History. As in many pediatric histories, parents and caregivers can provide valuable information simply based on their prior observations of the child. For example, in the case of movements involving the dominant hand, one might observe the child switching to the non-dominant hand to perform routine tasks (such as eating or writing). The patient may describe an inability to control the movements or briefly suppress them, interference with normal activities, associated muscular weakness, and pain. Table 3 provides a list of important history questions that may shed light on the etiology of the abnormal movements. As with all pediatric patients, it is important to document past medical and surgical histories, family history, social history, medications, immunization status, and allergies.

The Neurologic Examination in Children. The approach to the neurologic examination in children is similar to that in adults, but may be limited depending on the child's age and cooperation. In the case of a new movement disorder, observing the child (ideally, prior to entering the room and being seen by the child) is essential and can provide a wealth of additional information, including motor skills, gait, and the ability to perform voluntary movements. It is also useful to have the child perform a series of specific tasks to see if the movements are altered (either suppressed or enhanced); this may be helpful when evaluating a tremor, for example. While movement disorders can usually be distinguished from one another based on one or more movement components, including timing/frequency, speed, duration, amplitude, characteristics, and associated findings observed, note that a patient may present with more than one movement type. Refer to Table 1 for distinguishing features of each movement disorder.

Hyperkinetic Movement Disorders

While few pediatric movement disorder epidemiologic studies exist, available data suggest that the hyperkinetic disorders are by far more common than hypokinetic, possibly accounting for greater than 90% of cases.[6]

Tics. Tics are repeated, individually recognizable (stereotyped), intermittent movements or movement fragments that are almost always briefly suppressible and are usually associated with awareness of an urge to perform the movement.[4]Tics are the most common of the hyperkinetic movement disorders, with a male predominance of 3:1 and a lifetime prevalence of 20% or greater in boys.[7] They most frequently manifest in the first decade of life, with a median age of onset of 6 to 7 years.[8] Motor tics can manifest as simple (one muscle

Table 1. Acute Movement Disorders in Children

Hyperkinetic/ dyskinetic	Definition[4] and key features	Clinical considerations
Tics	Repeated, individually recognizable (stereotyped), intermittent movements or movement fragments that are almost always briefly suppressible and are usually associated with awareness of an urge to perform the movement	Screen for signs of malignant Tourette syndrome[7]
Stereotypies	Repetitive, simple movements that can be voluntarily suppressed	If concern for autism disorder or Rett Syndrome ("hand-wringing," developmental regression), consultation/follow up with a pediatric neurologist[9]
Dystonia	Involuntary sustained or intermittent muscle contractions causing twisting and repetitive movements, abnormal postures, or both	Close monitoring/intervention for upper airway dystonia resulting in laryngospasm[10,11] In severe dystonia or status dystonicus, evaluate for rhabdomyolysis and acute renal failure[12]
Tremor	Rhythmic oscillation of a body part, produced by either alternating synchronous contractions of reciprocally innervated antagonistic muscles	Evaluate for parkinsonism
Chorea/ hemichorea	An ongoing random-appearing sequence of one or more discrete involuntary movements or movement fragments	Check blood glucose level Consider testing for acute rheumatic fever, including evaluation for carditis[13,14]
Ballism	Chorea that affects proximal joints such as shoulder or hip leading to large amplitude movements of the limbs, sometimes with a flinging or flailing quality	Check blood glucose level Consider workup for cerebrovascular cause (e.g. subthalamic nucleus CVA)
Athetosis	Slow, continuous, involuntary writhing movement that prevents maintenance of a stable posture	In neonates, consider birth complications (trauma, asphyxia) and kernicterus[15]
Myoclonus	A sequence of repeated, often nonrhythmic, brief shocklike jerks due to sudden involuntary contraction or relaxation of one or more muscles	Initiate seizure precautions if concern for associated epilepsy
Hypokinetic/ bradykinetic Parkinsonism (bradykinesia, rigidity, tremor)	Syndrome of slowness of movement (bradykinesia), tremor in the hands or legs, rigidity of muscles, shuffling gait, and postural instability	Consider a trial of levodopa
Special Cases Neuroleptic malignant syndrome (NMS)	Syndrome of fever, rigidity, mental status change, autonomic dysfunction, and movement disorder resulting from exposure to dopamine receptor-blocking drugs[16] Acute onset (contrast to serotonin syndrome) Elevated creatine kinase Transaminitis Leukocytosis	Treatment aimed at preventing life-threatening complications (dehydration, electrolyte imbalance, acute renal failure, pulmonary embolism, cardiac arrest, seizure, sepsis)[16] Supportive care includes lowering core temperatures (cooling or dantrolene for severe hyperthermia), hydration, and control of agitation with benzodiazepines[16]
Serotonin syndrome (SS)	Syndrome of mental status change, abnormal neuromuscular tone, and autonomic dysfunction resulting from exposure to serotonergic drugs[17] Subacute onset (contrast to neuroleptic malignant syndrome)	Treatment aimed at preventing life-threatening complications (seizure, disseminated intravascular coagulation, metabolic acidosis, cardiac arrest)[17] In contrast to NMS, dantrolene is NOT recommended for severe hyperthermia - consider paralysis and endotracheal intubation[17]

Table 2. Emergency Etiologies of Acute Abnormal Movements in Children

Class	Etiology	Associated Movement Disorder
Structural	Tumor	Dystonia, ataxia, seizure
	Acute disseminated encephalomyelitis	Dystonia, parkinsonism, ataxia
	Post-traumatic hemorrhage	Chorea, ataxia, seizure, myoclonus
	Hydrocephalus	Ataxia
	Tonsillar herniation	Ataxia
Infectious/post-infectious	Encephalitis	Chorea, dystonia, parkinsonism, ataxia, seizure, myoclonus
	Meningitis	Ataxia, seizure
	Acute necrotizing encephalopathy	Chorea, dystonia
	Acute rheumatic fever	Chorea
	Human immunodeficiency virus infection	Chorea, myoclonus
	Epstein-Barr virus infection	Chorea
	Ramsay Hunt syndrome	Myoclonus, ataxia
	Guillain-Barré syndrome	Ataxia
	Acute cerebellar ataxia	Ataxia
	Subacute sclerosing panencephalitis	Myoclonus
	Lyme disease	Myoclonus
Toxic	Methanol	Chorea
	Carbon monoxide	Chorea
	Lead	Ataxia, myoclonus
Metabolic	Hypo/hypernatremia	Chorea, seizure, myoclonus
	Hypo/hyperglycemia	Chorea, seizure, myoclonus
	Hypocalcemia	Chorea, seizure
	Hyperthyroidism	Chorea, tremor, myoclonus
	Wilson disease	Chorea, dystonia, myoclonus
Autoimmune	NMDAR encephalitis	Chorea, dystonia, parkinsonism
	Opsoclonus-myoclonus-ataxia syndrome	Myoclonus, ataxia
	Systemic lupus erythematosus	Chorea
	Henoch-Schönlein purpura	Chorea
Neurodegenerative	Huntington disease	Dystonia, myoclonus
	Ataxia telangiectasia	Chorea, dystonia, myoclonus
	Lesch-Nyhan disease	Chorea, dystonia
	Rett syndrome	Stereotypies, myoclonus
	Parkinson disease	Parkinsonism, myoclonus
Vascular	Stroke	Chorea, dystonia, myoclonus, ataxia, seizure
	Perinatal hypoxia-ischemia	Chorea, dystonia, seizure
	Antiphospholipid antibody syndrome	Chorea, tremor, parkinsonism
	Systemic lupus erythematosus	Chorea, tremor, parkinsonism
	Moyamoya disease	Chorea
	Vertebrobasilar dissection	Ataxia
Psychogenic	Psychiatric	Any
Adapted from references 18-21		

group) or complex (multiple muscle groups), and are easily identified by their stereotyped and predictable nature (for example, intermittent shoulder shrugging). The most distinguishing feature, however, is the premonitory urge that patients describe prior to each tic. Acute onset motor tics usually do not lead to serious injuries or falls. Malignant Tourette syndrome, reported in about 5% of Tourette syndrome

Table 3. Key History Questions

Question	Examples	Associated Movement Disorder(s)
New medications (or possible ingestion of someone else's medication)?	Methylphenidate	Tremor, motor tics
	Neuroleptics (e.g., haloperidol, risperidone, prochlorperazine)	Dystonia, parkinsonism, TD, NMS
	Metoclopramide	Dystonia, parkinsonism, TD, NMS
	Antiepileptics (e.g., phenytoin, carbamazepine, valproate)	Chorea, dystonia, tremor, parkinsonism, myoclonus
	Cetirizine	Dystonia
	Levodopa	Motor tics, chorea, myoclonus
	Amiodarone	Tremor, myoclonus
	Lithium	Tremor, parkinsonism
	Beta-adrenergic agonists (e.g., albuterol)	Tremor
	Dopamine receptor antagonists	Tremor
	Selective serotonin reuptake inhibitors	Serotonin syndrome, myoclonus
Immunization status?	Subacute sclerosing panencephalitis (complication of measles)	Myoclonus (with personality changes, lethargy, progressive dementia)
Possible toxic exposure?	Lead	Tremor
	Cyanide	Parkinsonism
	Carbon monoxide	Chorea
	Methanol	Chorea
Substance use or abuse?	Cocaine, amphetamines	Chorea, tremor
	Alcohol	Tremor
Family history?	Huntington disease	Dystonia
	Wilson disease	Dystonia, chorea
	Dystonia	Dystonia
	Ataxia telangiectasia	Dystonia, chorea
	Myoclonic epilepsy	Myoclonus
	Lesch-Nyhan disease	Dystonia, chorea

patients,[22] involves a variable presentation of severe tics, suicidal ideation, and self-injurious behavior. A small number of patients may also present with psychotic symptoms, including hallucinations and paranoia.[6]

An interesting but controversial topic relating to acute onset tics/Tourette syndrome and/or obsessive compulsive disorder (OCD) symptoms is the pediatric autoimmune neuropsychiatric disorder associated with streptococcal infection (PANDAS), whose presence in the medical literature and the media is increasing. While much debate exists about this condition, it may be beneficial to test a child with a new-onset OCD and/or tic disorder for an underlying group A streptococcal infection, which, if identified, requires treatment.[23]

Stereotypies. Stereotypies are repetitive, simple movements that can be voluntarily suppressed.[4] Of the hyperkinetic movement disorders, stereotypies are the most benign. They are single, simple back-and-forth movements usually of the upper extremities, and are often precipitated by emotional excitement or stress.[4] The child is usually able to perform normal activities. As with tics, children with a stereotypy can be distracted from performing the movement, and they can be voluntarily stopped. Unlike tics, there is no premonitory urge to perform the movement.

Stereotypies are somewhat common in healthy preschool-aged children[17] and are also seen in children with autism spectrum disorders.[24] One ominous condition that can present with stereotypies is Rett syndrome, a genetically based developmental regression that most commonly affects girls and is classically associated with a "hand-wringing" stereotypy.[25] Although there are no established drug therapies, follow-up with a pediatric neurologist is recommended.

Dystonia. Dystonia involves involuntary sustained or intermittent muscle contractions causing twisting and repetitive movements, abnormal postures, or both.[4] Dystonia in children is usually a primary disorder, most frequently seen in patients with dyskinetic cerebral palsy or a genetic mutation. Dystonic movements are commonly precipitated by attempts at voluntary movement (and can be task-specific, e.g., the patient has dystonic movement while walking

forward but not backward) and usually do not persist during sleep. They can be distinguished from other movement disorders by their consistent pattern and by the sustained abnormal postures that result.

In a previously neurologically normal child, a secondary cause for dystonia must be evaluated. With a negative family history and an absence of signs of neurodegeneration (e.g., personality changes, dementia), the workup should focus on infectious and drug-induced causes. (*See Table 2.*)

Drug-induced dystonic reactions (DIDR) are well-documented in the literature, and most commonly result after starting a therapeutic dose of a neuroleptic[26] (e.g., haloperidol) or antiemetic medication (e.g., ondansetron,[27] metoclopramide[28]), although many other drug types have been associated.[11,29] The majority of DIDRs occur within 5 days of starting a new drug.[26,30]

The typical presentation involves focal posturing of the head and neck, but one retrospective study found 7% of children with a DIDR presented with laryngospasm, an airway emergency.[31] In addition to removing the offending agent, multiple sources recommend IV diphenhydramine in the acute management.[13] Benzodiazepines, anticonvulsants, and antispasmodics (e.g., baclofen) may also be tried if the movements are not adequately controlled after multiple doses. A child with a history of drug-induced dystonia is at increased risk for a second episode; thus, take extreme caution when deciding whether to administer any agent that has been implicated in the development of acute dystonic reactions.

Status dystonicus, a condition of generalized, unremitting dystonia, is usually limited to patients with preexisting dystonia and can be precipitated by infection or medication (initiation or discontinuation). The therapeutic approach is similar to that for acute dystonic reactions, but these patients require close monitoring in an intensive care unit setting while undergoing treatment.

Chorea. Chorea is an ongoing, random-appearing sequence of one or more discrete involuntary movements or movement fragments.[4] While chorea is classically taught as a "dance-like" movement disorder involving all extremities, patients can also present with hemichorea. Chorea can also occur with athetosis, which is a slower, writhing movement limited to the extremities. The movements of chorea can be brief and jerky (as seen with Sydenham chorea) or slower and flowing (as seen in Huntington disease).

The best-described and most common acquired form is Sydenham chorea (SC), a major manifestation of acute rheumatic fever (ARF) according to the Jones criteria.[14] Children with SC generally develop sudden-onset jerky movements involving the face and extremities and can also have muscular weakness (clinically demonstrated most easily as the "milkmaid's grip," or an inability to maintain a handgrip). Dysarthria, personality changes, and emotional lability are also common. SC is typically a later manifestation of ARF (with polyarthritis and carditis occurring earlier), developing within 1-6 months after a group A streptococcal infection.[32] In the appropriate clinical setting, new-onset SC should prompt an evaluation for carditis.[32,33] It has been demonstrated that children treated for ARF in the setting of SC who are free of carditis or valvulitis at diagnosis will be free of heart disease if there is no recurrence of ARF. Children with an audible murmur at diagnosis can expect resolution within the first year.[33]

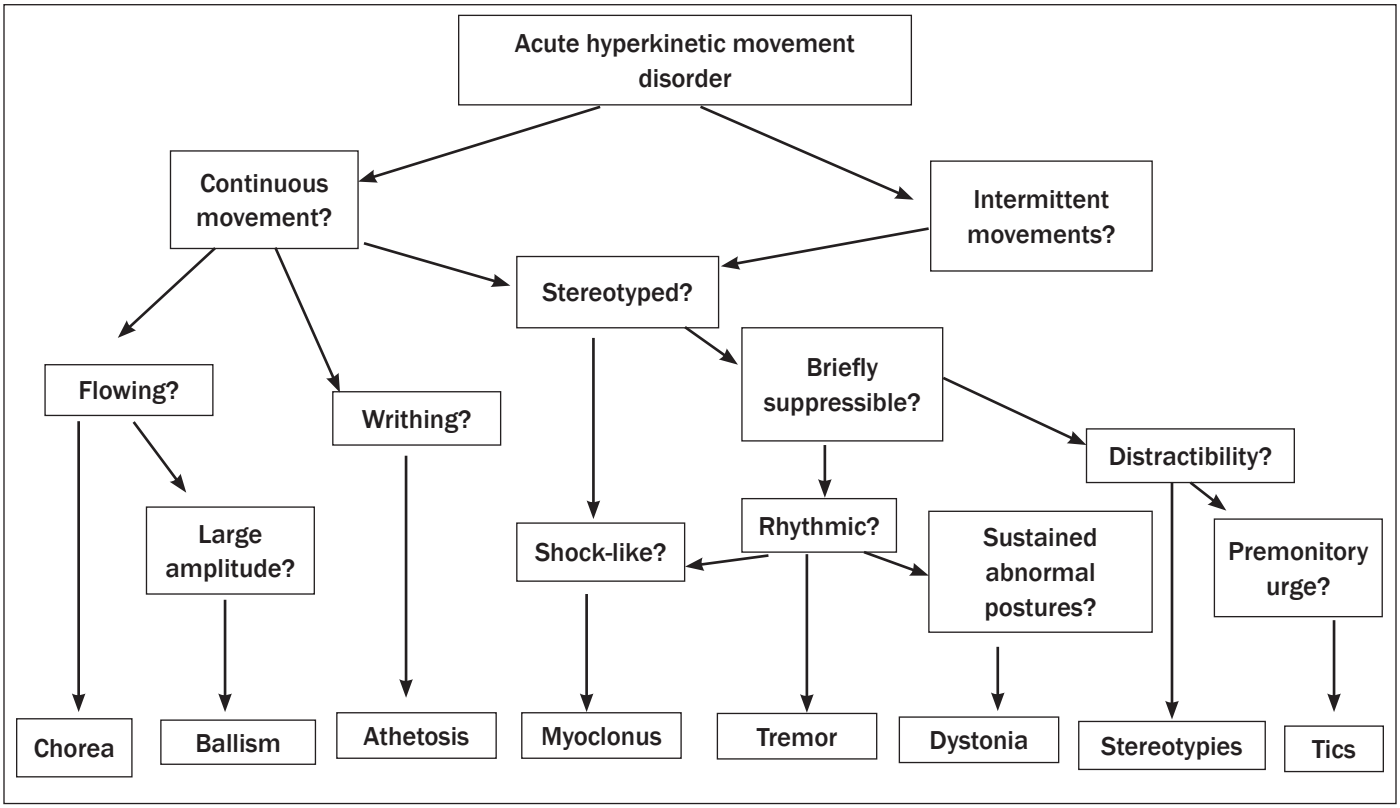
In female patients of child-bearing age, a pregnancy test should be included in the diagnostic workup of new chorea. Chorea gravidarum has become a rare phenomenon since the advent of penicillin in the treatment of ARF. It usually begins in the first or early second trimester and resolves in the third trimester or shortly after birth.[34] Today, it is most commonly seen in patients with systemic lupus erythematosus (SLE) and the antiphospholipid antibody syndrome (APS), disorders which are themselves associated with spontaneous abortion and other complications.[35]

Chorea occurs in about 10% of children with SLE. Neurologic manifestations are common in SLE, and are the first symptom in as many as one-third to two-thirds of children; the most common manifestation is seizure.[36] Chorea is somewhat more variable (i.e., can occur before or after diagnosis of SLE) and seems to be more common in girls.[37] CNS involvement conveys a worse prognosis.[21]

While cerebrovascular disease in children is very rare, it should be considered in the differential diagnosis of new chorea or hemichorea. Reported cases of chorea/hemichorea in children thought to be due to a vascular cause include moyamoya disease (a chronic, progressive cerebrovascular disease characterized by bilateral occlusion of the carotid and circle of Willis arteries, with resultant prominent collateral circulation resembling a "cloud of smoke" on angiography),[38,39] post-pump chorea (a phenomenon limited to children who have undergone recent extracorporeal bypass, as in cardiac surgery),[40] and diabetes mellitus. Cases of this movement disorder in diabetes have been reported for both diabetic ketoacidosis and nonketotic hyperglycemia.[41] Fortunately, resolution of this movement disorder can be achieved with tight glucose control, although neuroleptic medication is also usually required. This is in contrast to ischemic or hemorrhagic stroke, in which the chorea persists despite adequate treatment.

Myoclonus. Myoclonus involves a sequence of repeated, often nonrhythmic, brief, shock-like jerks due to sudden involuntary contraction or relaxation of one or more muscles.[4] Myoclonus occurs in all age groups in childhood, from benign neonatal sleep myoclonus to juvenile myoclonic epilepsy to essential myoclonus (which usually develops before age 20). While myoclonus can involve numerous body regions, the characteristic feature is the very

Figure 1. Algorithm for Distinguishing Between Hyperkinetic Movement Disorders



rapid "shock-like" duration of each movement; this generally helps to distinguish myoclonus from other hyperkinetic movement disorders. However, myoclonus may be difficult to distinguish from tics and tremor, which can also be quite rapid. Tics generally do not persist during sleep, whereas myoclonus frequently does (and some forms of myoclonus occur only during sleep). Quite possibly the best distinguishing factors, however, are the premonitory urge and distractibility that are virtually unique to tics. Tremor is best distinguished by its rhythmicity.

In addition to epileptic and nonepileptic classes of myoclonus, there are also several subtypes named for the anatomic origin (cortical, subcortical, spinal) and precipitants of movement (spontaneous, reflex, action). Cases of febrile myoclonus have also been reported and, like simple febrile seizures, resolve with temperature correction.[42] If it is an isolated and non-progressive symptom, myoclonus is usually benign. However, acute onset, progressive or symptomatic myoclonus in a previously healthy child tends to be an ominous sign and should prompt evaluation for a treatable cause.[43]

Myoclonus may occur after a cerebral anoxic event such as with drowning/near-drowning or cardiac arrest. Seizure precautions should be initiated for all patients with new symptomatic myoclonus, as it is frequently associated. Furthermore, in any patient with features of epileptic myoclonus (e.g., positive family history of seizures, myoclonus upon awakening, increasing frequency of myoclonic jerks, signs of encephalopathy), the workup should include an electroencephalogram (EEG). Benign myoclonus should have a normal EEG.

The acute management of symptomatic myoclonus should involve anticonvulsant therapy, whether or not the etiology is epileptic. Clonazepam (4-10 mg/day) and/or valproic acid (250-4200 mg/day) are effective options.[44]

A rare but important phenomenon worth mentioning is opsoclonus-myoclonus-ataxia syndrome (OMA, also known as "dancing eyes-dancing feet"). OMA is characterized by subacute limb and truncal myoclonus with involuntary and multidirectional saccades. Children with previously normal gait may develop an ataxia as well. The two most important etiologies to evaluate for children with new OMA are postviral and paraneoplastic. Fifty percent of patients with OMA will have an underlying neuroblastoma.[45] Any child presenting with OMA should, thus, have urgent imaging (CT or MRI of the chest/abdomen/pelvis) to further evaluate for neuroblastoma.[46] Excluding a brain lesion with CT/MRI is also useful, and if acute infection is suspected, CSF analysis should be included in the workup; analysis may be normal or reveal a lymphocytic pleocytosis.[47] Removal of a malignant mass alone does not result in complete resolution of the movement disorder, but most

children benefit from immunologic therapy (e.g., steroids, IVIG).

Tardive Dyskinesia (TD). This condition involves a variable mixture of orofacial dyskinesia, athetosis, dystonia, chorea, tics, and facial grimacing that appears with a delayed onset after prolonged use of dopamine receptor blocking agents.[48] Like neuroleptic malignant syndrome (discussed later), the risk of TD increases with increasing dose and duration of treatment. The incidence of TD in adults undergoing long-term second-generation antipsychotic treatment is less than 1%, and probably even lower in children and adolescents.[49] In children, TD may also occur with withdrawal of an antipsychotic agent. A recent review demonstrated an increased risk of tardive dyskinesia in patients treated with risperidone for schizophrenia; the effect may be less prevalent among young children.[50]

Classically, TD presents as stereotyped movements of the lower face, most commonly the mouth and tongue.[48] TD is rarely life-threatening, but closer observation and support may be required for patients with disabling movements (e.g., those that interfere with eating) or those with respiratory dyskinesias causing tachypnea or other signs of distress. Despite several studies evaluating pharmacologic therapy for TD, most have shown only slight to moderate benefit; the current focus, thus, is on prevention and early detection, although it may be helpful to consider a trial of benzodiazepine or anticholinergic medication to minimize the symptoms.[51]

Tremor. Tremor is the rhythmic oscillation of a body part, produced by alternating synchronous contractions of reciprocally innervated antagonistic muscles.[4] Tremor is common in adults; up to one-third of adult patients with essential tremor report onset of the movement disorder in childhood or adolescence. The defining characteristic of tremor is a rhythmic pattern that distinguishes it from other rapid movements such as myoclonus and tics. Any body part can exhibit a tremor, but the vast majority are seen in the hands.[52] Tremor is classified based on its associated movement triggers (rest, postural, action, intention). In children, the most common form is action tremor, which is further subdivided into physiologic and essential (familial).

Epidemiologic data for pediatric tremors are sparse, but available studies suggest that childhood-onset tremor is less common than adult-onset and is usually essential.[53] Treatment with a beta adrenergic receptor antagonist, such as propranolol, may be effective for essential tremor in children (as it has been demonstrated in adults), although randomized trials are lacking.

A child with abrupt onset or rapidly progressive tremor should be evaluated for exposure to certain toxins or medications (such as antiepileptics, beta-agonists, stimulants, alcohol, or lead) and should undergo urgent evaluation with measurements of serum glucose, thyroid function tests, brain MRI (especially if focal neurologic deficits exist), and serum drug levels or urine toxicology screen (if applicable).

Summary. Figure 1 depicts a proposed manner in which one can distinguish between the hyperkinetic movement disorders, although mixed pictures may also occur and make precise identification more difficult.

Hypokinetic Movement Disorders

In contrast to the variety of hyperkinetic movement disorders, hypokinetic (or bradykinetic) disorders in children are limited to primary and secondary parkinsonism.

Parkinsonism. This condition is a syndrome of slowness of movement (bradykinesia), tremor in the hands or legs, rigidity of muscles, shuffling gait, and postural instability.[4] Parkinsonism is exceedingly rare in children. Not surprisingly, then, few epidemiologic data are available. However, as with adults, parkinsonism can be a primary (e.g., Parkinson disease) or secondary. Parkinsonism classically presents with the "TRAP" signs and symptoms: tremor at rest, muscular rigidity, akinesia, postural instability. However, available case reports of parkinsonism in children suggest that a wider variety of presentations is possible, including non-motor complaints such as paucity of speech, mutism, autonomic dysfunction, and altered mental status.[54] Patients can even present without the classic tremor in the akinetic-rigid subtype of parkinsonism.

Drugs are the most common etiology for new-onset parkinsonism in children. Reported precipitants are chemotherapeutics, antimicrobials, antipsychotics, and antiemetics (metoclopramide).[55,56] Parkinsonism is much less commonly seen with structural brain lesions, infections, and hereditary syndromes (e.g. Wilson's disease, Huntington disease), but these should be considered in the workup if the appropriate clinical context exists. Regardless of the etiology of parkinsonism, the treatment of choice is levodopa (ideally prescribed by a pediatric neurologist).

Movement Disorders: Special Cases

Neuroleptic Malignant Syndrome (NMS). NMS is a syndrome of fever, rigidity, mental status change, autonomic dysfunction, and movement disorder resulting from exposure to dopamine receptor-blocking drugs.[16] NMS is rare in adults (with incidence reports varying from 0.02-3%) and even more uncommon in children, with the literature for the latter being limited to case reports.[57] It is a potentially fatal complication of treatment with antipsychotic medications such as haloperidol (with higher doses carrying an increased risk of developing NMS), and can have a variable presentation centered around hyperthermia, rigidity, and muscle damage.[57] The pathogenesis involves central dopaminergic receptor blockage that occurs with use of these medications. Other risk factors include intramuscular administration and rapid increases in dose. The diagnostic criteria for NMS (based on the DSM-IV) include the development of severe muscle rigidity and elevated

temperature associated with the use of neuroleptic medication with two or more of the following:

- diaphoresis
- dysphagia
- tremor
- incontinence
- changes in level of consciousness ranging from confusion to coma
- mutism
- tachycardia
- elevated or labile blood pressure
- leukocytosis
- laboratory evidence of muscle injury (e.g., elevated serum creatinine kinase).

It should be noted that while rigidity must be present by definition, patients can present with hyperkinetic movements as well. It is crucial to evaluate and exclude CNS infection if there is concern for this based on history or physical exam, as it can present similarly. The differential diagnosis for NMS also includes heat stroke, malignant hyperthermia, and serotonin syndrome.

Depending on the severity of symptoms and vital sign instability, treatment will range from supportive care and observation to ICU-level monitoring. Like most presentations associated with an adverse drug effect, the offending agent should always be discontinued. The mainstays of treatment are hydration to prevent renal damage and sedation (e.g., with benzodiazepines) to control agitation. Hyperthermia can be managed with external cooling, levodopa, bromocriptine, or dantrolene. Dialysis can be utilized in severe cases of renal dysfunction, although the neuroleptic medications themselves are not usually dialyzable. Early recognition and treatment may help prevent complications and death due to cardiac arrest, pulmonary embolism, and sepsis, and while it appears the mortality rate has declined over time, there remains a high risk of morbidity.[16] If the neuroleptic medication must be continued, it should be slowly titrated to the desired dose over a period of two or more weeks to reduce the risk of recurrence of NMS.

Serotonin Syndrome. Serotonin syndrome is a condition of mental status change, abnormal neuromuscular tone, and autonomic dysfunction resulting from exposure to serotonergic drugs.[17]

Classic serotonin syndrome (SS) is a triad of fever, altered mental status, and myoclonus, which emerges in the setting of exposure to serotonergic drugs, including antidepressant medications (selective serotonin reuptake inhibitors [SSRI], serotonin-norepinephrine reuptake inhibitors [SNRI], monoamine oxidase inhibitors [MAOI], tricyclic antidepressants [TCA], etc.), although several less obvious culprits have been reported, such as analgesics (triptans, opiates except morphine) and illicit drugs (MDMA, LSD, cocaine). SS in young children has been reported through accidental ingestion or even after taking an appropriate weight-based dose.[58,59] The presentation can be quite variable in severity, from mild myoclonic jerks to a seizure-like state leading to rhabdomyolysis. (*See Table 4.*) SS is diagnosed clinically and may be difficult to distinguish from NMS. Although the onset of SS is generally over several days, with treatment it resolves within 24 hours. The creatinine kinase elevation seen with virtually every case of NMS is much less common (though not unheard of) in SS. Myoclonus and hyperreflexia are more characteristic of SS, though they are not always seen.

The mainstay of treatment of SS is discontinuation of the serotonergic agent. Further management is largely supportive and consists of close vital sign monitoring, fluid resuscitation, external cooling, and sedation with benzodiazepines.[17] In cases of severe hyperthermia (temperature greater than 41.1 degrees Celsius), control of excess muscular activity with paralysis and endotracheal intubation is necessary. However, paralysis with succinylcholine should be avoided in these patients due to possible rhabdomyolysis; a non-depolarizing paralytic agent (such as rocuronium or vecuronium) should be used instead. In contrast to NMS, dantrolene is not recommended in the management of hyperthermia in SS (as no survival benefit has been demonstrated in SS).[17] Treatment with a serotonin antagonist may also be considered in severe cases. Cyproheptadine use is supported by several case reports in the literature, although there are no randomized trials studying its efficacy and, thus, no FDA approval exists for its use in SS.[60]

Table 4. Hunter Criteria for Serotonin Syndrome

Criteria
Presence of a serotonergic agent
One or more of the following:
• spontaneous clonus
• inducible clonus and agitation or diaphoresis
• ocular clonus and agitation or diaphoresis
• tremor and hyperreflexia
• hypertonia
• temperature above 38 degrees Celsius and ocular clonus or inducible clonus

Adapted from: Dunkley EJ, et al. The Hunter Serotonin Toxicity Criteria: Simple and accurate diagnostic decision rules for serotonin toxicity. *QJM* 2003;96:635.

Psychogenic Movement Disorders

Hyperkinetic or hypokinetic movement disorders associated with an underlying psychological disorder in which an organic cause for the movements is excluded are psychogenic movement disorders.

Psychogenic movement disorders can present with any movement type. While this diagnosis is difficult

Table 5. Suggested Initial Workup for Acute Movement Disorders in Children

Movement Disorder	Differential Diagnosis	Initial Studies to Consider (immediately available results)	Other Suggested Evaluation (as inpatient or outpatient)
Tics	Simple motor tics Complex motor tics Tourette Syndrome	CT/MRI brain (if focal neurologic deficits)	
Stereotypies	Physiologic Autism disorder Rett syndrome	CT/MRI brain (if focal neurologic deficits)	Genetic testing - Autism disorder - Rett syndrome
Chorea Hemichorea Hemiballism Choreoathetosis	Wilson's disease - chorea - choreoathetosis - "wing-beating" tremor - progressive dystonia	Liver function enzymes CT/MRI brain (may see ventriculomegaly and brain atrophy in advanced cases) Slit lamp examination - Kayser-Fleischer rings (neurologic disease)	Serum/urine copper level Ceruloplasmin levels
	Systemic lupus erythematosus - chorea	CT/MRI brain (if focal neurologic deficits or seizure) CBC, reticulocyte count - hemolytic anemia, leukopenia, thrombocytopenia Urinalysis — cellular casts Electrocardiogram — pericarditis	ANA Anti-DNA antibody Anti-Sm antibody Antiphospholipid antibody Lupus anticoagulant
	Acute rheumatic fever (ARF) - Sydenham chorea	CBC with differential Throat swab (rapid streptococcal antigen test) ESR/CRP Electrocardiogram - prolonged PR interval Echocardiogram - valvulitis	Throat culture for group A streptococci ASO titer - negative titers do not exclude the diagnosis Anti-DNase B titer
	Pregnancy - chorea gravidarum (usually resolves spontaneously)	Urine beta HCG	- Consider ARF - antiphospholipid antibody syndrome - Wilson's disease - hyperthyroidism - toxic/metabolic etiology
	Kernicterus - choreoathetosis - ballism - tremor - dystonia	CT/MRI brain - increased T2-weighted signal intensity in the basal ganglia (bilirubin deposition) CBC - hemolytic anemia Serum bilirubin	Hearing screen Genetic testing - G6PD - Crigler-Najjar - galactosemia
	Diabetes mellitus - hemichorea - hemiballism	Serial blood glucose measurements Serum electrolytes and osmolarity - rule out DKA Glycosylated hemoglobin (A1c)	CT/MRI brain (basal ganglia pathology)
	Hyperthyroidism - chorea	TSH, T4	
	Toxic metabolic encephalopathy	CBC with differential PT/PTT Serum electrolytes (including Ca, Mg, Phos) Liver function enzymes, ammonia Arterial blood gas CSF studies Blood and urine toxicologic screens CT/MRI brain (or cranial ultrasound in newborn)	TSH Cortisol Blood lead level

Table 5. Suggested Initial Workup for Acute Movement Disorders in Children (continued)

Movement Disorder	Differential Diagnosis	Initial Studies to Consider (immediately available results)	Other Suggested Evaluation (as inpatient or outpatient)
Myoclonus	Epileptic	CT/MRI brain (if first presentation) EEG	
	Neuroblastoma - opsoclonus-myoclonus-ataxia syndrome	CT or MRI chest/abdomen/pelvis - tumor evaluation	Urinary vanillylmandelic acid (VMA) and homovanillic (HVA) 123-I-metaiodobenzylguanidine (MIBG) scan
	Cofactor deficiency - myoclonus	CBC (exclude anemia)	Biotin Pyridoxine Cobalamin
	Serotonin syndrome	CBC with differential PT/PTT Creatinine kinase Liver function enzymes Serum electrolytes, BUN, creatinine Urinalysis - myoglobinuria Core temperature monitoring	
Rigidity	Neuroleptic malignant syndrome	CBC with differential Creatinine kinase Liver function enzymes Serum electrolytes, BUN, creatinine Urinalysis - myoglobinuria Core temperature monitoring	
Dystonia	Huntington disease - rigidity - bradykinesia - dystonia	CT/MRI brain (neurodegeneration)	Genetic testing
Any movement disorder with associated psychiatric/behavioral symptoms	NMDA receptor encephalitis	CBC with differential CT or MRI brain CSF studies	EEG (if concern for seizure)
Any movement disorder with associated fever/rigors	CNS infection - meningitis - encephalitis	CBC with differential PT/PTT Serum electrolytes, BUN, creatinine CT brain Blood cultures CSF studies	EEG (if concern for seizure)

to make due to a lack of established criteria, certain warning signs should alert providers to a potential psychopathology (in a patient without focal neurologic deficits and an otherwise normal workup):

- Abrupt onset with rapid progression;
- Multiple movement types with an inconsistent pattern, variability (e.g. changing from one movement type to another);
- Distractibility with performing tasks;
- Disability out of proportion to the movement disorder.

Note that a child who presents with abnormal movements and

personality changes should be evaluated for anti-N-methyl-D-aspartate receptor (NMDAR) encephalitis, especially in the setting of a viral prodrome (e.g., headache, fever, myalgias). A prospective study of 52 children with acute movement disorders found that 5 patients who presented primarily with behavioral alterations also demonstrated chorea, dystonia, or parkinsonism. These patients were diagnosed by positive NMDAR antibodies in the CSF and serum.[18]

Clearly, a psychogenic movement disorder is a diagnosis of exclusion and is not easily made in the emergency department, where the often extensive workup for a movement disorder starts. However, an awareness of this phenomenon is important as patients may require psychiatric consultation and follow-up. One small study identified a multidisciplinary approach and family involvement in treatment as integral to effectively treating a psychogenic movement disorder.[61]

Differential Diagnosis and Workup

Perhaps one of the most important distinctions to make early is whether one is dealing with a potential seizure, even after completing a comprehensive history and physical. While it may be difficult to distinguish seizure from hyperkinetic movements simply based on phenomenology, there are some features that lend more weight to one or the other. First, alteration of mental status in the setting of abnormal movements usually indicates seizure, although an acute process (such as CNS infection causing myoclonus or chorea) may also present with abnormal mentation. In addition, the presence of a premonitory aura (such as sensory symptoms or nausea) is more indicative of seizure, as is a post-ictal state or Todd's paralysis.[62]

Hyperkinetic movements may also present similar to cerebellar pathology, causing a variety of ataxias. Cerebellar ataxia can usually be distinguished by abnormal coordination and balance testing (e.g., as seen with the "drunken sailor" gait characteristic of truncal ataxia). A positive Romberg's test is a strong indicator of sensory ataxia due to a cerebellar or dorsal column abnormality.[63] A child with a movement disorder may appear to have abnormal cerebellar tests on examination, although this may be a result of inability to control the baseline movements.

Management

As for all patients, emergency department management for a new movement disorder begins with assessing for stability of the airway, breathing, and circulation before proceeding with workup. In the context of movement disorders, patients with upper airway dystonia should be immediately assessed for emergent intubation while receiving positive pressure ventilation via bag-valve mask. This can be done while administering treatment (e.g. IV diphenhydramine) to reverse the dystonia (especially if a drug-induced cause is suspected). A patient with altered mental status should also be considered for intubation for airway protection. A child with SS or NMS may require intubation for hypoxic respiratory failure if there is concern for life-threatening pulmonary embolism (a known complication of these disorders). As stated previously, use of succinylcholine as a paralytic agent should be avoided in these patients due to possible rhabdomyolysis. Circulation can be maintained with intravenous crystalloids, particularly for those children with SS or NMS who may have rhabdomyolysis from severe muscle rigidity. Children who appear septic should be given several weight-based fluid boluses while obtaining blood cultures and administering broad-spectrum antibiotics. Consider vasopressors if fluid resuscitation fails to maintain adequate mean arterial pressure.

While movement disorders are diagnosed clinically, the suspected underlying cause may require laboratory and radiologic investigation. Table 5 lists a general differential diagnosis for each movement disorder and suggested workup, including studies that are widely available and can be obtained quickly in the emergency department. In most cases, brain imaging (computed tomography and/or magnetic resonance imaging) is a reasonable means to exclude structural pathology. Head computed tomography (CT) is readily available at many institutions and provides rapid results. However, certain lesions are better characterized using magnetic resonance imaging (MRI), which may require hospital admission to complete. It should be noted, however, that not every child with abnormal movements requires imaging in the emergency department. Patients with certain movement disorders, such as tics, are unlikely to have abnormal brain imaging.[64]

Children with abnormal movements who also present with fever or rigors should be evaluated for CNS infection and sepsis, including blood cultures (and subsequently empiric antibiotics if bacterial meningitis is suspected),[65] CT brain, and lumbar puncture with CSF studies. Any child who presents with abnormal movements after head trauma should be risk-stratified for clinically important traumatic brain injury according to the validated prediction rules established by the PECARN group.[66] It is also important to consider evaluation for non-accidental trauma in any child presenting with an acute neurological problem, especially in the setting of multiple injuries or inconsistent history. If one is unable to distinguish between a movement disorder and seizure, EEG (particularly with video monitoring) may be considered as part of the workup.

A child without altered mental status or constitutional signs/symptoms (such as fever or vomiting) who has an unremarkable history, is functioning normally in their daily activities, and who isn't at risk for injury due to their movements may be better candidates for outpatient follow-up and further evaluation.

Once the movement disorder has been confidently diagnosed clinically, it is reasonable to begin therapy to minimize or stop the movements, particularly for patients who are

Table 6. Suggested Oral Pharmacotherapy for Childhood Movement Disorders

Movement Disorder	Drug	Clinical Considerations
Tics	Clonidine: Start 0.05 mg/day, max 0.3 mg/day TID Haloperidol: Start 0.25 mg/day, max 5-10 mg/day	Clonidine: Avoid abrupt discontinuation (risk of withdrawal syndrome – agitation, increased tics, tachycardia, diaphoresis) Haloperidol: Increased risk of drug-induced dystonic reaction; chronic use limited by risk of developing tardive dyskinesia
Dystonia	Diphenhydramine IV: 1-2 mg/kg per dose (max 50 mg) Levodopa: Start 100 mg BID or TID, with carbidopa Trihexyphenidyl: Start 2-4 mg/day, max 60 mg/day Pimozide: Start 1 mg/day, max 6-12 mg/day	Diphenhydramine: Indicated for suspected drug-induced dystonia Levodopa: Trial indicated in all patients with limb-onset dystonia Consider anticholinergic (with or without pimozide) if no response to levodopa
Tremor	Primidone: 10-25 mg/kg/day BID or TID Propranolol: Start 0.5-1 mg/kg/day TID, max 20-40 mg/kg/day TID Clonazepam: 0.01-0.3 mg/kg/day BID or TID	
Chorea/hemichorea Ballism	Haloperidol: 0.5-20 mg/day Pimozide: 1-10 mg/day Reserpine: 0.1-0.3 mg/day BID Tetrabenazine: 12.5-100 mg/day	Haloperidol and pimozide: Avoid in patients with QT interval prolongation Reserpine and tetrabenazine: For patients with debilitating chorea
Myoclonus	If epileptic: Sodium valproate: 250-4200 mg/day Clonazepam: 4-10 mg/day If posthypoxic or postencephalitic: Levetiracetam (specific dose not studied in children)	
Adapted from: Edgar TS. Oral pharmacotherapy of childhood movement disorders. *J Child Neurol* 2003;18:S40-S49.		

injured or distressed. Table 6 lists some suggested drug therapies. If possible, however, the movements should be documented as best as possible prior to treatment (ideally, on video with consent). A pediatric neurologist should be consulted for specific drug recommendations prior to starting medication, as he or she will follow up with the patient after discharge.

Disposition

Most children with new movement disorders can safely be discharged home after a thorough evaluation. Certain situations may require longer ED observation or brief hospital admission, such as the need for further workup and control of symptoms. Admission is necessary in conditions for which airway monitoring is needed, for patients with disabling movements or those suffering physical injury due to the movement disorder, and for treatment of reversible and potentially harmful movement disorders (e.g., SS, NMS). In addition, children may benefit from consultation with a physical and/or occupational therapist in adaptive gait training and activities of daily living (ADLs) to function as independently as possible, including return to school.

Regardless of disposition, every child with a new movement disorder should have consultation and follow-up with a pediatric neurologist in addition to their primary physician, even when a reversible cause is identified, and particularly if drug therapy is initiated.

Reassurance and education are important interventions for patients and their families. While most movement disorders are easily distinguished from seizure on the basis of classic

movement patterns and maintenance of consciousness, it is important to reassure parents and caregivers who may be most concerned about a seizure. Furthermore, the fact that most movement disorders in previously healthy children are not life-threatening and not permanent should be established, although the expected timeline of recovery will vary based on the specific movement disorder.

Educating patients and families about the spectrum of etiologies for most movement disorders should occur early in the evaluation, which may take several hours in the ED. For patients discharged home, personalized discharge instructions are another way in which emergency physicians can educate about movement disorders, especially in the case of drug-induced syndromes to provide a reminder about offending agents and recommend notifying other healthcare providers about avoiding specific medications.

Additionally, it is important to address patients' and families' expectations about treatment of a movement disorder. If drug therapy is started in the emergency department, discharge instructions should contain detailed medication instructions and potential adverse effects (including the risk of developing a drug-induced dystonic reaction with a neuroleptic agent). If a reversible cause is found (e.g. drug-induced dystonia, acute rheumatic fever), patients may expect complete resolution of the abnormal movements but may not achieve this immediately. Sydenham chorea, even with appropriate antibiotic and immunomodulatory treatment, may take several months to fully resolve. Furthermore, children with a previously diagnosed movement disorder are at higher risk for recurrence. Appropriate follow-up is essential for long-term monitoring and management.

References

1. Fernández-Alvarez E. Prevalence of paediatric movement disorders. In: Thénenin M, ed. *Paediatric Movement Disorders: Progress in Understanding*. Montrouge, France: John Libbey Eurotext; 2005:1-18.
2. Alexander GC, et al. Increasing off-label use of antipsychotic medications in the United States, 1995-2008. *Pharmacoepidemiology and Drug Safety* 2011;20:177-184.
3. Olfson M, et al. Trends in antipsychotic drug use by very young, privately insured children. *J Am Acad Child Adolesc Psychiatry* 2010;49;1:13-23.
4. Comer JS, et al. National trends in child and adolescent psychotropic polypharmacy in office-based practice, 1996-2007. *J Am Acad Child Adolesc Psychiatry* 2010;49;10:1001-1010.
5. Helm M. Acute cerebellar ataxia. *UpToDate* 2012.
6. Fernandez-Alvarez E, Aicardi J. *Movement Disorders in Children*. London: High Holborn House. 2001:19.
7. Sanger TD, et al. Definition and classification of hyperkinetic movement disorders in childhood. *Movement Disorders* 2010;25;11:1538-1549.
8. Schlaggar BL, Mink JW. Movement disorders in children. *Pediatrics in Review* 2003;24:39-51.
9. Mahone EM, et al. Repetitive arm and hand movements (complex motor stereotypies) in children. *J Pediatr* 2004;145:391-395.
10. Jacobsen RC. Out-of-hospital lingual dystonia resulting in airway obstruction. *Prehosp Emerg Care* 2011;15:537-540.
11. Esen I, et al. Cetirizine-induced dystonic reaction in a 6-year-old boy. *Pediatric Emerg Care* 2008;24;9:627-628.
12. Jamuar SS, et al. Rhabdomyolysis, acute renal failure, and cardiac arrest secondary to status dystonicus in a child with glutaric aciduria type I. *Molecular Genetics and Metabolism* 2012;106:488-490.
13. Robottom BJ, Shulman LM, Weiner WJ. Drug-induced movement disorders: Emergencies and management. *Neurol Clin* 2012;30:309-320.
14. Gibofsky A, Zabriskie JB. Clinical manifestations and diagnosis of acute rheumatic fever. *UpToDate* 2010.
15. Przekop A, Sanger TD. Birth-related syndromes of athetosis and kernicterus. *Handbook of Clinical Neurology* 2011;100:387-395.
16. Wijdicks EFM. Neuroleptic malignant syndrome. *UpToDate* 2012.
17. Boyer EW. Serotonin syndrome. *UpToDate* 2012.
18. Dale RC, et al. A prospective study of acute movement disorders in children. *Dev Med Child Neurol* 2010;52:739-748.
19. Agrawal D. Approach to the child with acute ataxia. *UpToDate* 2012.
20. Wilfong A. Overview of the classification, etiology, and clinical features of pediatric seizures and epilepsy. *UpToDate* 2012.
22. Cheung MC, Shahed J, Jankovic J. Malignant Tourette Syndrome. *Movement Disorders* 2007;22;12:1743-1750.
21. Mink JW, Zinner SH. Movement disorders II: Chorea, dystonia, myoclonus, and tremor. *Pediatrics in Review* 2010;31:287-295.
22. Cheung MC, Shahaed J. Jankovic J. Malignant Tourette syndrome. *Movement Disorders* 2007;22;1743-1750.
23. Pinchichero ME. PANDAS: Pediatric autoimmune neuropsychiatric disorder associated with group A streptococci. *UpToDate* 2012.
24. Lewis MH, Bodfish JW. Repetitive behavior disorders in autism. *Ment Retard Dev Disabil Res Rev* 1998;4:80-89.
25. Goldman S, Temudo T. Hand stereotypies distinguish Rett syndrome from autism disorder. *Movement Disorders* 2012;27;8:1060-1062.
26. Swett Jr. CS. Drug-induced dystonia. *Am J Psychiatry* 1975;132;5:532-534.
27. Patel A, et al. Ondansetron-induced dystonia, hypoglycemia, and seizures in a child. *Ann Pharmacotherapy* 2011;45:e7.
28. Boulloche J, et al. Dystonic reactions with metoclopramide: Is there a risk population? *Helv Paediatr Acta* 1987;42;5-6:425-432.
29. Singh MK, et al. Acute dystonia associated with aripiprazole in a child. *J Am Acad Child Adolesc Psychiatry* 2007;46;3:306-307.
30. Ayd Jr. FJ. A survey of drug-induced extrapyramidal reactions. *JAMA* 1961;175;12:1054-1060.
31. Derinoz O, Caglar AA. Drug-induced movement disorders in children at paediatric emergency department: 'Dystonia.' *Emerg Med J* 2012 March 7 (Epub ahead of print).
32. Weiner SG, Normandin PA. Sydenham chorea: A case report and review of the literature. *Pediatric Emerg Care* 2007;23;1:20-24.
33. Panamonta M, et al. The relationship of carditis to the initial attack of Sydenham's chorea. *International J Cardiology* 2004;94:241-248.
34. Lee MJ, Hickenbottom S. Neurologic disorders complicating pregnancy. *UpToDate* 2011.
35. Robottom BJ, Weiner, WJ. Chorea gravidarum. *Handbook of Clinical Neurology* 2011;100:231-235.
36. Quintero-Del-Rio AI, Miller V. Neurologic symptoms in children with systemic lupus erythematosus. *J Child Neurology* 2000;15(12):803-807.
37. Parikh S, et al. Neurologic characteristics of childhood lupus erythematosus. P*ediatr Neurol* 1995;13:198-201.
38. Gonzalez-Alegre P, et al. Moyamoya-induced paroxysmal dyskinesia. *Mov Disord* 2003;18:1051-1056.
39. Suwanwela NC. Moyamoya disease: Etiology, clinical features, and diagnosis. *UpToDate* 2012.
40. Robinson RO, et al. Choreic syndrome after cardiac surgery. *Arch Dis Child* 1988;63:1466-1469.

41. Mihai CM, et al. Atypical onset of diabetes in a teenage girl: A case report. *Cases Journal* 2008;1:425.
42. Dooley JM, Hayden JD. Benign febrile myoclonus in childhood. *Can J Neurol* Sci 2004;31:504-505.
43. Pranzatelli MR. Myoclonus in childhood. *Semin Pediatr Neurol* 2003;10;1:41-51.
44. Edgar TS. Oral pharmacotherapy of childhood movement disorders. *J Child Neurol* 2003;18:S40-S49.
45. Russell HV, et al. Clinical presentation, diagnosis, and staging evaluation of neuroblastoma. *UpToDate* 2012.
46. Brunklaus A. Investigating neuroblastoma in childhood opsoclonus-myoclonus syndrome. *Arch Dis Child* 2011 Apr 3 (Epub ahead of print).
47. Dalmau J, Rosenfeld MR. Paraneoplastic syndromes causing movement disorders. *Handbook of Clinical Neurology* 2011;100:315-321.
48. Tarsy D. Tardive dyskinesia: Clinical features and diagnosis. *UpToDate* 2012.
49. Correll CU, Kane JM. One-year incidence rates of tardive dyskinesia in children and adolescents treated with second-generation antipsychotics: A systematic review. *J Child Adol Psychopharm* 2007;17;5: 647-655.
50. McDonagh M, et al. Drug class review: Atypical antipsychotic drugs: Final update 3 report. Portland (OR): Oregon Health & Science University; July 2010.
51. Tarsy D. Tardive dyskinesia: Prevention and treatment. *UpToDate* 2012.
52. Jankovic J, et al. Essential tremor among children. *Pediatrics* 2004;114;5: 1203-1205.
53. Ferrara J, Jankovic J. Epidemiology and management of essential tremor in children. *Pediatr Drugs* 2009;11;5:293-307.
54. Pranzatelli MR, et al. Clinical spectrum of secondary parkinsonism in childhood: A reversible disorder. *Pediatric Neurology* 1994;10;2:131-140.
55. Chutorian AM, et al. Toxic pediatric parkinsonism: Report of a child with metabolic studies and response to treatment. *J Child Neurol* 2003;18:812-815.
56. Mott SH, et al. Encephalopathy with parkinsonian features in children following bone marrow transplantations and high-dose amphotericin B. *Ann Neurol* 1995;37:810-814.
57. Ty EB, Rothner DA. Neuroleptic malignant syndrome in children and adolescents. *J Child Neurol* 2001;16;3:157-163.
58. Marechal C, et al. Serotonin syndrome induced by tramadol intoxication in an 8-month-old infant. *Pediatric Neurology* 2011;44;1:72-74.
59. Phan H, et al. Serotonin syndrome following a single 50 mg dose of sertraline in a child. *Clinical Toxicology* 2008;46:845-849.
60. Graudins A, et al. Treatment of the serotonin syndrome with cyproheptadine. *J Emerg Med* 1998;16;4:615-619.
61. Faust J, Soman TB. Psychogenic movement disorders in children: Characteristics and predictors of outcome. *J Child Neurol* (online):1-5. 2011.
62. Helms A, Shulman L. Chapter 12: Movement disorders that imitate epilepsy. In: Kaplan PW, Fisher RS. *Imitators of Epilepsy.* 2nd ed. New York: Demos Medical Publishing. 2005:163-184.
63. Subramony SH. Chapter 22: Ataxic disorders. In: Daroff: *Bradley's Neurology in Clinical Practice.* Philadelphia: Elsevier Saunders. 2012.
64. Scahill L, et al. Contemporary assessment and pharmacotherapy of Tourette syndrome. *NeuroRx* 2006;3:192-206.
65. Talan DA, et al. Role of empiric parenteral antibiotics prior to lumbar puncture in suspected bacterial meningitis: State of the art. *Rev Infect Dis* 1988;10;2: 365-376.
66. Kuppermann N, et al. Identification of children at very low risk of clinically-important brain injuries after head trauma: A prospective cohort study. *Lancet* 2009;374:1160-1170.

Physician CME Questions

1. A 16-month-old boy presents with sudden-onset myoclonus with ocular involvement and an ataxic gait. His mother also recently noticed an abdominal mass while bathing the child. What diagnosis needs to be excluded?
   A. intussusception
   B. neuroblastoma
   C. Meckel's diverticulum
   D. Wilms' tumor
   E. acute cerebellar ataxia

2. A couple brings their 6-month-old infant to the ED with generalized abnormal movements after finding the infant unresponsive in his crib. The infant is breathing spontaneously with intact airway-protective reflexes, but is somnolent. The remainder of the history is unremarkable, and the mother states that the child was last seen normal the previous night before she left for work. Her boyfriend, who cared for the child at that time, states the infant was acting normally prior to going to sleep. He denies trauma. A CT of the head demonstrates a subarachnoid hemorrhage. In addition to stabilizing the child and speaking with the appropriate consultants, which of the following diagnoses must be considered in this child?
   A. cerebral aneurysm
   B. hypertensive encephalopathy
   C. non-accidental trauma
   D. meningitis
   E. sudden infant death syndrome

3. What is the most common acquired cause of chorea in children?
   A. moyamoya disease
   B. drug-induced chorea
   C. chorea gravidarum
   D. acute rheumatic fever (Sydenham chorea)
   E. trauma

4. Which of the following characteristics distinguishes tics from other hyperkinetic movement disorders?
   A. premonitory urge
   B. intermittent frequency
   C. distractibility
   D. stereotyped movements
   E. shock-like nature

5. Which of the following is the recommended first-line treatment for drug-induced dystonic reactions?
   A. lorazepam
   B. succinylcholine
   C. metoclopramide
   D. atropine
   E. diphenhydramine

6. The most common type of abnormal movements in a child is:
   A. chorea
   B. athetosis
   C. seizure
   D. ataxia
   E. dystonia

7. With appropriate supportive treatment (including discontinuation of the offending agent), serotonin syndrome generally resolves within:
   A. 7 days
   B. 24 hours
   C. 2 weeks
   D. 1 month
   E. 3 days

8. The most common acquired cause of parkinsonism in children is:
   A. Parkinson's disease
   B. cerebral palsy
   C. drug-induced
   D. encephalitis
   E. trauma

9. A 17-year-old boy presents with uncontrolled stereotyped movements of his tongue. He has a history of schizophrenia and has been taking the same dose of risperidone for several years. What is the most likely diagnosis?
   A. tardive dyskinesia
   B. drug-induced dystonia
   C. simple motor tics
   D. stroke
   E. neuroleptic malignant syndrome

10. A 5-year-old girl presents with chorea. After completing a history and physical examination, you decide to perform a workup for acute rheumatic fever. Which of the following conditions must be evaluated for in a child with suspected Sydenham chorea?
    A. subacute bacterial endocarditis
    B. carditis/valvulitis
    C. myocardial infarction
    D. congestive heart failure
    E. QT interval prolongation

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Pediatric Emergency Medicine Reports

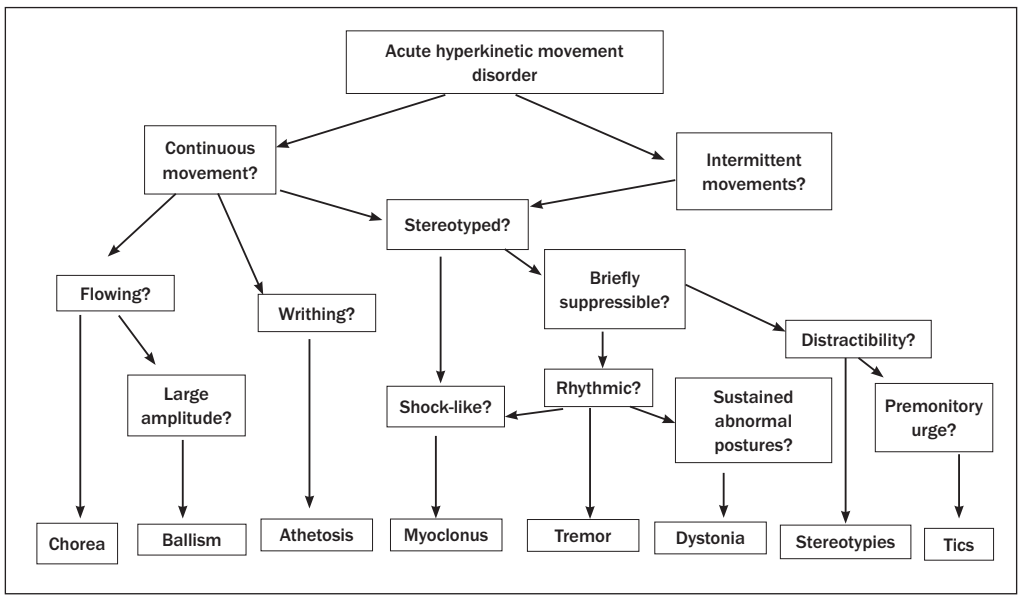
The Practical Journal of Pediatric Emergency Medicine

Acute Movement Disorders

Acute Movement Disorders in Children

Hyperkinetic/ dyskinetic	Definition and key features	Clinical considerations
Tics	Repeated, individually recognizable (stereotyped), intermittent movements or movement fragments that are almost always briefly suppressible and are usually associated with awareness of an urge to perform the movement	Screen for signs of malignant Tourette syndrome
Stereotypies	Repetitive, simple movements that can be voluntarily suppressed	If concern for autism disorder or Rett Syndrome ("hand-wringing," developmental regression), consultation/follow up with a pediatric neurologist
Dystonia	Involuntary sustained or intermittent muscle contractions causing twisting and repetitive movements, abnormal postures, or both	Close monitoring/intervention for upper airway dystonia resulting in laryngospasm In severe dystonia or status dystonicus, evaluate for rhabdomyolysis and acute renal failure
Tremor	Rhythmic oscillation of a body part, produced by either alternating synchronous contractions of reciprocally innervated antagonistic muscles	Evaluate for parkinsonism
Chorea/ hemichorea	An ongoing random-appearing sequence of one or more discrete involuntary movements or movement fragments	Check blood glucose level Consider testing for acute rheumatic fever, including evaluation for carditis
Ballism	Chorea that affects proximal joints such as shoulder or hip leading to large amplitude movements of the limbs, sometimes with a flinging or flailing quality	Check blood glucose level Consider workup for cerebrovascular cause (e.g. subthalamic nucleus CVA)
Athetosis	Slow, continuous, involuntary writhing movement that prevents maintenance of a stable posture	In neonates, consider birth complications (trauma, asphyxia) and kernicterus
Myoclonus	A sequence of repeated, often nonrhythmic, brief shocklike jerks due to sudden involuntary contraction or relaxation of one or more muscles	Initiate seizure precautions if concern for associated epilepsy
Hypokinetic/ bradykinetic Parkinsonism (bradykinesia, rigidity, tremor)	Syndrome of slowness of movement (bradykinesia), tremor in the hands or legs, rigidity of muscles, shuffling gait, and postural instability	Consider a trial of levodopa
Special Cases Neuroleptic malignant syndrome (NMS)	Syndrome of fever, rigidity, mental status change, autonomic dysfunction, and movement disorder resulting from exposure to dopamine receptor-blocking drugs Acute onset (contrast to serotonin syndrome) Elevated creatine kinase Transaminitis Leukocytosis	Treatment aimed at preventing life-threatening complications (dehydration, electrolyte imbalance, acute renal failure, pulmonary embolism, cardiac arrest, seizure, sepsis) Supportive care includes lowering core temperatures (cooling or dantrolene for severe hyperthermia), hydration, and control of agitation with benzodiazepines
Serotonin syndrome (SS)	Syndrome of mental status change, abnormal neuromuscular tone, and autonomic dysfunction resulting from exposure to serotonergic drugs Subacute onset (contrast to neuroleptic malignant syndrome)	Treatment aimed at preventing life-threatening complications (seizure, disseminated intravascular coagulation, metabolic acidosis, cardiac arrest) In contrast to NMS, dantrolene is NOT recommended for severe hyperthermia - consider paralysis and endotracheal intubation

Algorithm for Distinguishing Between Hyperkinetic Movement Disorders



Suggested Initial Workup for Acute Movement Disorders in Children

Movement Disorder	Differential Diagnosis	Initial Studies to Consider (immediately available results)	Other Suggested Evaluation (as inpatient or outpatient)
Tics	Simple motor tics Complex motor tics Tourette Syndrome	CT/MRI brain (if focal neurologic deficits)	
Stereotypies	Physiologic Autism disorder Rett syndrome	CT/MRI brain (if focal neurologic deficits)	Genetic testing - Autism disorder - Rett syndrome
Chorea Hemichorea Hemiballism Choreoathetosis	Wilson's disease - chorea - choreoathetosis - "wing-beating" tremor - progressive dystonia	Liver function enzymes CT/MRI brain (may see ventriculomegaly and brain atrophy in advanced cases) Slit lamp examination - Kayser-Fleischer rings (neurologic disease)	Serum/urine copper level Ceruloplasmin levels
	Systemic lupus erythematosus - chorea	CT/MRI brain (if focal neurologic deficits or seizure) CBC, reticulocyte count - hemolytic anemia, leukopenia, thrombocytopenia Urinalysis — cellular casts Electrocardiogram — pericarditis	ANA Anti-DNA antibody Anti-Sm antibody Antiphospholipid antibody Lupus anticoagulant
	Acute rheumatic fever (ARF) - Sydenham chorea	CBC with differential Throat swab (rapid streptococcal antigen test) ESR/CRP Electrocardiogram - prolonged PR interval Echocardiogram - valvulitis	Throat culture for group A streptococci ASO titer - negative titers do not exclude the diagnosis Anti-DNase B titer
	Pregnancy - chorea gravidarum (usually resolves spontaneously)	Urine beta HCG	- Consider ARF - antiphospholipid antibody syndrome - Wilson's disease - hyperthyroidism - toxic/metabolic etiology
	Kernicterus - choreoathetosis - ballism - tremor - dystonia	CT/MRI brain - increased T2-weighted signal intensity in the basal ganglia (bilirubin deposition) CBC - hemolytic anemia Serum bilirubin	Hearing screen Genetic testing - G6PD - Crigler-Najjar - galactosemia
	Diabetes mellitus - hemichorea - hemiballism	Serial blood glucose measurements Serum electrolytes and osmolarity - rule out DKA Glycosylated hemoglobin (A1c)	CT/MRI brain (basal ganglia pathology)
	Hyperthyroidism - chorea	TSH, T4	
	Toxic metabolic encephalopathy	CBC with differential PT/PTT Serum electrolytes (including Ca, Mg, Phos) Liver function enzymes, ammonia Arterial blood gas CSF studies Blood and urine toxicologic screens CT/MRI brain (or cranial ultrasound in newborn)	TSH Cortisol Blood lead level

Movement Disorder	Differential Diagnosis	Initial Studies to Consider (immediately available results)	Other Suggested Evaluation (as inpatient or outpatient)
Myoclonus	Epileptic	CT/MRI brain (if first presentation) EEG	
	Neuroblastoma - opsoclonus-myoclonus-ataxia syndrome	CT or MRI chest/abdomen/pelvis - tumor evaluation	Urinary vanillylmandelic acid (VMA) and homovanillic (HVA) 123-I-metaiodobenzylguanidine (MIBG) scan
	Cofactor deficiency - myoclonus	CBC (exclude anemia)	Biotin Pyridoxine Cobalamin
	Serotonin syndrome	CBC with differential PT/PTT Creatinine kinase Liver function enzymes Serum electrolytes, BUN, creatinine Urinalysis - myoglobinuria Core temperature monitoring	
Rigidity	Neuroleptic malignant syndrome	CBC with differential Creatinine kinase Liver function enzymes Serum electrolytes, BUN, creatinine Urinalysis - myoglobinuria Core temperature monitoring	
Dystonia	Huntington disease - rigidity - bradykinesia - dystonia	CT/MRI brain (neurodegeneration)	Genetic testing
Any movement disorder with associated psychiatric/ behavioral symptoms	NMDA receptor encephalitis	CBC with differential CT or MRI brain CSF studies	EEG (if concern for seizure)
Any movement disorder with associated fever/rigors	CNS infection - meningitis - encephalitis	CBC with differential PT/PTT Serum electrolytes, BUN, creatinine CT brain Blood cultures CSF studies	EEG (if concern for seizure)

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