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Chapter 46

Disorders of Movement

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Key Points

- Disorders of movement can present to the emergency department as new complaints or as an exacerbation of a known condition.
- Without a prior diagnosis, the etiologies of movement disorders are difficult to differentiate.
- For children with disorders of movement, the gait is often the most revealing part of the physical examination.
- The etiologies of movement disorders in children are diverse.

Selected Diagnoses

Postinfectious acute cerebellar ataxia
Acute disseminated encephalomyelitis
Acute dystonia
Huntington's disease
Sydenham's chorea
Tourette's syndrome

Discussion of Individual Diagnoses

Postinfectious Acute Cerebellar Ataxia

Postinfectious acute cerebellar ataxia is the most common cause of acute ataxia in children.[1] Most cases occur in the second year of life.[2] Seventy-five percent of patients report an antecedent febrile illness within the 3 weeks preceding the onset of ataxia. About half of the patients will have nystagmus. A staggering gait, dysarthria, and truncal ataxia are the characteristic features.[2] Peripheral white blood cell counts are expected to be normal. Cerebrospinal fluid studies typically yield a mild pleocytosis with normal protein. A computed tomographic scan of the head will typically be normal. In contrast, magnetic resonance imaging of the head typically shows abnormal signal intensity in the cerebellum, cerebral white matter, or globus pallidus.[3] The pathophysiology of postinfectious acute cerebellar ataxia is poorly understood. The offending agent may produce the ataxia via direct invasion of the cerebellar tissues or via an autoimmune-mediated effect.[4] Cerebral and cerebellar antibodies have been found after varicella infections.[5] Parvovirus B19 has been associated with cerebellar vascular injuries.[6] Single-photon emission computed tomography of the head may demonstrate reduced regional cerebellar blood flow.[7]

Treatment is controversial and predominantly supportive. For severe or persistent cases, glucocorticoids and intravenous immune globulin have been used with some success.[4] Fortunately, most children experience a benign course and recover fully.

Acute Disseminated Encephalomyelitis

Acute disseminated encephalomyelitis, also know as parainfectious encephalitis, is the most common demyelinating condition in children.[8] This demyelinating disease of childhood has an acute onset, and children with acute disseminated encephalomyelitis typically present with ataxia, abnormal motor control, and altered mental status. This condition is typically considered a self-limited, monophasic disorder that is thought to be immune mediated[9] (Table 46–1). The incidence of acute disseminated encephalomyelitis is estimated to be as high as 5 in 10,000 hospital admissions. The association with an antecedent viral infection is reported to be between 54% and 77%. However, the true cause-and-effect relationship has not been definitively established due to the high background frequency of viral illnesses in children.[8,10-12] The average age of onset is 8 years, with a reported age range of 3 months to 18 years of age. A common reported prodrome is headache, fever, nausea, vomiting, and malaise. There are no clinical, laboratory, or computed tomographic findings pathognomonic for acute disseminated encephalomyelitis. The diagnosis is made based on the neurologic examination and the presence of large, multifocal, hyperdense areas in the brain and spinal cord on T2-weighted magnetic resonance imaging. Electroencephalograms typically reveal nonspecific findings and are not diagnostic. Cerebrospinal fluid findings are also nondiagnostic and may reveal mild pleocytosis.

Acute disseminated encephalomyelitis is thought to be due to an immune-mediated process. Treatment is primarily aimed at modulating the immune response and includes the use of glucocorticoids, intravenous immune globulin, and plasmapheresis. To date there have been no clinical trials that compare the effectiveness of these modalities to each other

Table 46–1 Known Causes of Acute Disseminated Encephalomyelitis

Viral	Bacterial	Immunizations
Hepatitis A virus	*Clostridium tetani*	Influenza (killed)
Herpes simplex virus	β-Hemolytic streptococci	Measles (live)
HIV	*Legionella*	Meningococcal A and C
Human herpesvirus 6	*Mycoplasma*	Rabies (killed)
Cytomegalovirus	Rickettsiae	Rubella (live)
Coronavirus		
Mumps virus		
Varicella-zoster virus		

Abbreviation: HIV, human immunodeficiency virus.

Table 46–2 Causes of Dystonia by Rapidity of Symptom Onset

Acute	Gradual
Carbon monoxide	Ataxia-telangiectasia
Cerebral abscess	Brain tumor
Cerebrovascular accidents	Glutaric aciduria type I
Cisapride	Hallervorden-Spatz disease
Dextromethorphan	Leigh disease
Disulfram	Mitochondrial disorders
Droperidol	Neonatal hypoxia
Encephalitis	Wilson's disease
Hemolytic-uremic syndrome	
Infantile bilateral striatal necrosis	
Methanol	
Metoclopramide	
Postinfectious	
Promethazine	

or to placebo. This is mostly due to the relative rarity of these cases. Treatment recommendations are usually based on small case series.[10] Response to these interventions has been reported to be rapid and dramatic in many cases, with complete recovery in 57% to 81% of patients.[8] In general, the clinical outcome is good, with complications mainly involving mild cognitive deficits. Unfortunately, relapses may occur in as many as one third of patients.

Acute Dystonia

Dystonic reactions are characterized by opisthotonus, lateral neck flexion, oculogyric spasm, tightening of the extremities, and pain.[13] Onset of dystonia may occur acutely or gradually, and the causes are heterogeneous (Table 46–2). Because the causes of dystonia are varied, there is not a single overall approach that is appropriate to these children. The evaluation and management is typically guided by the likely cause based on the history and physical examination. Although clinical experience suggests acute dystonia is relatively rare in children, one of the more common causes is exposure to medications. Commonly implicated medications include dextromethorphan, cisapride, and metoclopramide. Treatment for medication-induced acute dystonia is often accomplished by administering 1.25 mg/kg (up to 50 mg) of intravenous or intramuscular diphenhydramine. Unless there are other extenuating circumstances, children who respond well to the diphenhydramine can usually be discharged home from the emergency department after a short period of observation. Continuing oral diphenhydramine at home for a few days is probably prudent. Coordinating the discontinuance of the inciting medication with the prescribing physician is a reasonable courtesy in many cases.

Huntington's Disease

Huntington's disease is an inherited neurodegenerative disorder that is characterized by movement, cognitive, and behavioral problems. Huntington's disease is autosomal dominant and affects 1 in 10,000 people.[14] Huntington's disease is primarily a disease of middle-aged adults; fewer than 10% of cases are diagnosed in children younger than 20 years of age. The term *juvenile Huntington's disease* is sometimes used to designate these early-onset cases. While adults will often have uncontrolled choreic movements, children are much more likely to manifest rigid akinetic symptoms.[15] The initial presentation of juvenile Huntington's disease is often very subtle and nonspecific, making early diagnosis difficult. Common early symptoms include personality changes, school performance problems, rigidity, slowness, stiffness, an awkward gait, clumsiness, speech difficulties, drooling, frequent choking episodes, and seizures.[16-19]

There is no specific emergency department evaluation for children suspected of having juvenile Huntington's disease. If suspected, the decision to pursue an inpatient or outpatient diagnostic evaluation for children with suspected juvenile Huntington's disease is made on a case-by-case basis. Because Huntington's disease is a progressive disorder, patients may present for a variety of issues over the course of their disease. Seizures, poorly controllable myoclonus, increased rigidity, feeding difficulties, deteriorating mental status, and complications associated with medications are a few of the reasons a child with known Huntington's disease may present to the emergency department. Coordinating care with the family and the child's neurologist is usually prudent.

Sydenham's Chorea

First described by Thomas Sydenham, MD, in 1685, Sydenham's chorea is a hypotonic, hyperkinetic syndrome, characterized by spontaneous involuntary and uncoordinated movements. Other features may include muscular weakness, frequent falls, dysarthria, difficulty concentrating, impaired writing, slurred speech, and emotional lability.[20,21] In particular, Sydenham's chorea has been shown to be associated with neuropsychiatric disorders.[20-22] Sydenham's chorea is currently thought to be part of a somewhat controversial grouping of disorders known as "pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections" (PANDAS).[23,24] The disorders of movement in Sydenham's chorea are usually bilateral, but hemichorea may be seen in as many as 20% of these patients.[21] There is a clear relationship between Sydenham's chorea and rheumatic fever. The presence of Sydenham's chorea is one of the major criteria used to clinically diagnose rheumatic fever. Sydenham's chorea is seen in as many as 25% of patients diagnosed with rheumatic fever. School-age children are the pediatric group most commonly affected. A single episode of chorea can last 2 to 6 months.[22,25,26] If performed, magnetic resonance imaging of the brain typically reveals injury to the caudate nucleus.[20] The etiology of Sydenham's chorea is unknown, but an immune-mediated process is likely.[20]

A variety of treatments for Sydenham's chorea have been tried over the years, including haloperidol, barbiturates, chlorpromazine, benzodiazepines, and valproic acid.[27,28] Immune modulators such as corticosteroids and intravenous immune globulin have also been tried with variable success. Currently, the most promising treatment is valproic acid, which has been shown to be safe and relatively effective.[25] Valproic acid has been shown to both control the motor movements and stabilize mood swings.[25]

Tourette's Syndrome

Tourette's syndrome is the most common and well-known tic disorder.[29] The onset is usually between 2 and 28 years of age, with the peak around 11 years. Tourette's syndrome is characterized by repetitive, stereotypical tics that are intermittent and have a compulsive quality to them. The range of symptoms can be divided between vocal, motor, and behavioral. Boys tend to manifest motor and vocal symptoms, while girls tend to manifest more behavioral symptoms. The most well-known and dramatic of the vocal manifestations is coprolalia, the expression of "dirty" words or phrases. Most patients do not demonstrate this feature. Between 5% and 30% of patients with Tourette's syndrome manifest coprolalia. The vocal tics range from simple sounds to complex phrases or speech patterns such as echolalia (repetition of words) or palilalia (rapid repetition of words or phrases). The motor manifestations may be simple, fast, meaningless muscle movements or they may be slower, stereotyped movements that look purposeful. The behavioral symptoms include those labeled attention-deficit/hyperactivity disorder, obsessive-compulsive disorder, self-mutilation, aggression, and various learning disabilities.[30-33]

There is no specific emergent treatment for the manifestations of Tourette's syndrome. Coordinated care with the child's neurologist is prudent. Most children essentially "outgrow" their tics, but about 10% of children have persistence and worsening symptoms into adulthood.[29,34,35]

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