Paroxysmal movement disorders are a relatively uncommon but important subset of movement disorders. These disorders include paroxysmal dyskinesias, stereotypies, episodic ataxia, and benign paroxysmal torticollis. Some schemes of paroxysmal movement disorders include tics, which are discussed in Chapter 65, “Tic Disorders.” When approaching a patient with paroxysmal events that involve abnormal movements, one must include these disorders in the differential diagnosis. Evaluation of a paroxysmal movement disorder in children usually relies largely on the history because the abnormal movements are usually infrequent and the neurologic examination is normal between events. The history should include specific information with respect to frequency, timing, duration, and appearance of the clinical events in question.

In addition, it is important for the neurologist to see the movements. The ready availability of portable video cameras has improved the ability of the neurologist to actually witness infrequent episodes. Many parents have access to these devices and, very often, can provide the clinician with a videotape of representative events. Viewing an event in a natural setting will usually help distinguish a paroxysmal movement disorder from a seizure or transient ischemic event. This is especially true when the neurologic examination is unremarkable. Nevertheless, despite having witnessed an event, it can still be difficult to distinguish paroxysmal movement disorders from epileptic events. In these instances, a video electroencephalographic monitoring or ambulatory electroencephalographic monitoring can be used to determine whether an electrocortical correlate to the paroxysmal movement is present.

Assuming that a clinical episode can be captured, either in the office or by some form of video recording, it is of paramount importance to characterize the type of movement disorder exhibited by the patient. Tics, ataxia, stereotypies, tremor, chorea, dystonia, and athetosis may be present, either alone or in combination, in various paroxysmal movement disorders. Appropriate assignment of a clinical event to a specific movement disorder type will greatly aid in formulating the ultimate diagnosis. The following sections describe the most important paroxysmal movement disorders, with emphasis on diagnostic and treatment considerations.

**Paroxysmal Dyskinesias**

Paroxysmal dyskinesias are a relatively rare subset of hyperkinetic movement disorders defined by their episodic nature. Mount and Reback reported a familial form in which the proband had infantile-onset of periodic but prolonged dyskinesia induced by alcohol and other agents. The episodes were characterized by an aura of a tight sensation in the neck and abdomen and a sense of fatigue, followed by involuntary flexion of the arms and extension of the legs (dystonia). The spells progressed to involuntary choreo-
Paroxysmal dystonic choreoathetosis of Mount and Reback was subsequently used to help delineate it from the more common paroxysmal kinesigenic choreoathetosis. These historic terms, paroxysmal kinesigenic choreoathetosis and paroxysmal dystonic choreoathetosis are still used, but their utility is limited because of substantial overlap in characteristics.

The clinical manifestations of paroxysmal dyskinesias vary. The attacks are often not witnessed because of their brief duration. Earlier classifications were inaccurate because they used terms like paroxysmal choreoathetosis or paroxysmal dystonic choreoathetosis, implying that in all attacks movements are easily characterized. However, the type of dyskinesia observed is extremely variable. Furthermore, the duration of episodes is not a reliable characteristic for differentiating between paroxysmal kinesigenic choreoathetosis and paroxysmal dystonic choreoathetosis of Mount and Reback.

Demirkiran and Jankovic have proposed a classification scheme that is based primarily on the precipitating events, arguing that the precipitant is the best predictor of clinical course and response to specific medications (see Suggested Readings). For example, kinesigenic dyskinesias are far more responsive to nonbenzodiazepine anticonvulsants than are nonkinesigenic dyskinesias, regardless of duration of spell or etiology. They proposed the following classification of paroxysmal dyskinesias: (1) kinesigenic, (2) nonkinesigenic, (3) exertion induced, and (4) hypnogenic (paroxysmal nocturnal dystonia of sleep). Secondary categorization is made on the basis of etiology: idiopathic (familial versus sporadic) and secondary.

Paroxysmal kinesigenic dyskinesia is often inherited in an autosomal dominant fashion, but a quarter of the cases are sporadic. The attacks may precede motor manifestation. Most patients have dystonia, but some have a combination of chorea and dystonia and, rarely, ballism. The attacks may be limited to one side of the body or even one limb. The attacks decrease in frequency during adulthood. The patients respond well to anticonvulsants (Table 66-1).

Paroxysmal nonkinesigenic dyskinesia is usually inherited as an autosomal dominant trait. The attacks occur more often in males than females in a 2:1 ratio. The age of onset can be in early childhood, but attacks may not start until the early 20s. The frequency varies from three per day to two per year. The usual precipitating factors are fatigue, alcohol, caffeine, and emotional excitement. The attack may start with involuntary movements of one limb but may spread to involve all extremities and the face. The usual duration is minutes to several hours. During the attack, the patient may be unable to communicate, but the patient remains conscious and continues to breathe normally. Some families have predominant dystonia and others have predominant choreoathetosis. The attacks are relieved by sleep and in some cases respond to pharmacologic intervention.

Paroxysmal exertion-induced dyskinesia is usually inherited in an autosomal dominant fashion, although sporadic cases have been described. The attacks are triggered by prolonged exercise. The frequency varies from 1 per day to 2 per month. The usual duration is 5 to 30 minutes. Exercise limited to the upper extremity may provoke an attack in the upper extremity alone.

Paroxysmal hypnogenic dyskinesia is characterized by attacks of dystonia, chorea, or ballism during non-rapid eye movement sleep. The frequency may be from 5 times per year to 5 times per night. These attacks may be associated with electroencephalogram (EEG) signs of arousal, and the patient usually falls asleep after the attack. The usual duration of the attacks is 30 to 45 seconds but may be longer. Sometimes the patients may have daytime attacks of dyskinesia. Paroxysmal hypnogenic dyskinesia

<table>
<thead>
<tr>
<th>Type of Dyskinesia</th>
<th>First-Line Agent</th>
<th>Second-Line Agent</th>
<th>Other Agents</th>
</tr>
</thead>
<tbody>
<tr>
<td>Paroxysmal nonkinesigenic dyskinesia</td>
<td>Benzodiazepine</td>
<td>Phenytoin, carbamazepine</td>
<td>Levodopa, benzopine, trihexyphenidyl, acetylcholinesterase inhibitors</td>
</tr>
<tr>
<td>Paroxysmal kinesigenic dyskinesia</td>
<td>Carbamazepine</td>
<td>Phenobarbital, gabapentin and valproate</td>
<td>Acetazolamide</td>
</tr>
</tbody>
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**TABLE 66-1. Treatment of Idiopathic Paroxysmal Dyskinesias**

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BC Decker Inc Pages 427–431
appears to be a heterogeneous condition comprising attacks of different durations and clinical features. However, many cases appear to represent frontal lobe seizures. Some of the patients have paroxysmal hypnogenic dyskinesia attacks preceding tonic-clonic seizures.

Most cases of paroxysmal kinesigenic dyskinesia, paroxysmal nonkinesigenic dyskinesia, and paroxysmal exertion-induced dyskinesia are idiopathic. These conditions may be sporadic but are usually inherited in an autosomal dominant fashion. They may rarely be autosomal recessive. In addition to idiopathic or genetic paroxysmal dyskinesias, there are many other causes of dyskinesias. These include multiple sclerosis, central and peripheral trauma, strokes, and infections. Secondary dyskinesias should be considered in all atypical cases, such as older age at onset, prolonged duration of attacks, and when there are associated interictal neurologic deficits. Other reported etiologies include perinatal hypoxic encephalopathy, hypoparathyroidism, pseudohypoparathyroidism, thyrotoxicosis, nonketotic hyperglycemia, and methylphenidate ingestion.

The differential diagnosis of paroxysmal dyskinesias includes seizures, pseudoseizures, and tics. Seizures arising from supplemental motor area may resemble dyskinesias and sometimes may be precipitated by movement. The idiopathic paroxysmal dyskinesias have to be distinguished from symptomatic ones. Sometimes paroxysmal dystonias may be psychogenic.

Prognosis depends on the type of paroxysmal dyskine-
sia. Paroxysmal kinesigenic dyskinesia, even of prolonged duration, responds well to standard anticonvulsants, and the attacks tend to diminish during adulthood. Paroxysmal nonkinesigenic dyskinesia and paroxysmal exertion-induced dyskinesia have a variable prognosis. Even with repeated attacks, the neurologic function between the attacks is normal. In symptomatic dyskinesias, the prognosis depends on the underlying disease.

Management of paroxysmal dyskinesia depends on the type (see Table 66-1). Paroxysmal kinesigenic dyskinesia typically responds well to antiepileptic drugs, including phenytoin, carbamazepine, phenobarbital, and leviteracem. The dosage required is usually less than the standard anticonvulsant dosage. For paroxysmal nonkinesigenic dyskinesia, pharmacotherapy is less successful. The most effective medications include clonazepam, haloperidol, alternate-day oxazepam, and anticholinergics. Antiepileptic drugs are ineffective in most cases. Avoidance of precipitating factors like alcohol, caffeine, and stress is important. For paroxysmal exertion-induced dyskinesia, avoidance of prolonged exercise may help diminish the frequency of attacks. Drug therapy is often ineffective, but there are isolated reports of improvement with levodopa or acetazolamide. Paroxysmal hypnogenic dyskinesia is best viewed as frontal lobe epilepsy until proved otherwise.

Stereotypies

Stereotypies are repetitive, rhythmic, patterned movements that are most common when the child is excited, anxious, or concentrating on something. Stereotypies frequently take the form of hand flapping, arm waving, complex hand movements, and facial grimacing, as well as hopping, jumping, or rocking. In contrast to tics, however, stereotypies are relatively invariant and do not migrate or evolve significantly in their appearance over time. Furthermore, patients do not go through periods where these movements are more or less apparent. Indeed, they seem most exquisitely sensitive to the child’s emotional state.

Much of the literature dealing with stereotypical behaviors has focused on specific patient populations, namely those who are mentally retarded or blind and those with specific diagnostic entities, such as autism or Rett syndrome. In these populations, it has been inferred that stereotypical behaviors are some form of self-stimulatory act. However, stereotypies may be seen in children with no physical or neurologic impairment.

The neurologic basis of stereotypies is unknown. It is interesting to note that many parents will often give a history that stereotypical behaviors began within the first year of life. Hand waving or hand flapping usually starts at 10 months of age in normal children, and it is interesting to speculate that stereotypies may be a persistence of this developmental milestone. With respect to the putative self-stimulatory aspect of stereotypies, this hypothesis is essentially untestable in mentally retarded or autistic populations.

As with most other paroxysmal movement disorders, it greatly aids in the diagnostic evaluation if stereotypies can be witnessed by the examiner. Although this may seem difficult, parents often will be aware of some specific trigger that may precipitate an event. Likewise, parents can identify activities at home that are associated with stereotypies so they can easily videotape the patient.

In children with no other neurologic impairment, stereotypies seldom require any specific form of therapy. Young children are typically unaware they are making stereotypical movements. There are few published reports of effective pharmacologic interventions in otherwise normal children who exhibit stereotypies. Reassurance and counseling should usually suffice, as these behaviors become much less prominent with maturity, although they may persist into adulthood. The best strategy is for parents to ignore the behavior or redirect the child. Punishment is not appropriate. In neurologically impaired populations, a large body of literature exists regarding cognitive-behavioral and pharmacologic interventions. In those who are institutionalized, pharmacologic strategies may be directed toward anxiolytics or neuroleptics.
Episodic Ataxia

Rarely, children may exhibit paroxysmal events best characterized as ataxic, with features suggestive of cerebellar dysfunction. Paroxysmal tonic up-gaze or associated chorea or dystonia have been reported as concomitants to the ataxia. Attacks may be precipitated by movement, stress, or fatigue, and typically resolve within minutes to hours. In these instances, a videotape of an event is invaluable, as parents often may only describe events as “shaking” with no loss of consciousness. Indeed, witnessed events usually demonstrate a relatively pure ataxia affecting gait and appendicular movements. There are two described entities with a presumed hereditary basis. Episodic ataxia with myokymia (EA1) describes a syndrome of paroxysmal ataxia precipitated by movement or startle associated with rippling of muscles in the body. This condition has been linked to a gene on chromosome 12 encoding a potassium channel. Episodic ataxia without myokymia (EA2) is non-kinesigenic and is allelic on chromosome 19 with the calcium channel gene associated with familial hemiplegic migraine.

Paroxysmal ataxias have been described in patients with amino acidopathies, disorders of lactate and pyruvate metabolism, repeated exposures to known cerebellar toxins, and vascular headaches. Appropriate historical information and laboratory studies should be performed before the true nature of the behavior is recognized.

Benign Paroxysmal Torticollis

Benign paroxysmal torticollis is an episodic disorder occurring in the first few months of life. It is characterized by head tilts to one side for a few hours or days. Rarely, spells can last up to 2 weeks. The torticollis may occur without any associated symptoms or may be accompanied by pallor, vomiting, irritability, or ataxia. Episodes typically recur with some regularity, up to twice a month initially and becoming less frequent as the child grows older. The spells abate spontaneously, usually by 2 to 3 years of age but always by age 5. The child is normal between spells. Intercital and ictal EEGs are normal.

The differential diagnosis is broad and diagnosis of benign paroxysmal torticollis is one of exclusion. Torticollis can be seen as an acute dystonic reaction to medication, as a symptom of a posterior fossa or cervical cord lesion, or as cervical vertebral abnormalities. In the case of structural lesions, the torticollis tends to be persistent and not paroxysmal. Torticollis can also be a sign of trochlear nerve palsy. Congenital muscular torticollis is present from birth, is nonparoxysmal, and is associated with palpable tightness or fibrosis of the sternocleidomastoid muscle unilaterally.

It has been suggested that benign paroxysmal torticollis is a migraine variant. There is often a family history of migraine. Some older children complain of headache during a spell, and many children go on to develop typical migraine after they have “outgrown” the paroxysmal torticollis. Treatment of benign paroxysmal torticollis is not required.

Other Disorders

A number of other paroxysmal movement disorders distinctly affect children. They are uncommon but still bear some discussion. Shuddering attacks are brief events that occur in infancy and are characterized by the child appearing to be cold and shivering. These events may occur multiple times per day and do not have an electrocerebral signature. The cause is unknown and the child is typically otherwise normal. Although shuddering attacks resolve spontaneously, there is a reported association with familial essential tremor. Beta-blockade is an effective treatment.

Another movement with paroxysmal features is that of spasmus nutans, accurately captured by the clinical triad of head nodding, nystagmus, and head tilt. This is another benign entity, although care must be taken to rule out an intracranial lesion, as spasmus nutans has been seen in the context of gliomas involving the optic chiasm or third ventricle. This disorder usually presents in the first year of life and remits by 3 to 4 years of age.

Masturbation is a normal behavior that occurs in both boys and girls. Although masturbation occurs at all ages and has even been observed in utero, it is most common at about 4 years of age and during adolescence. Masturbation in young children may involve unusual postures or movements that may be mistaken for abdominal pain or seizures. Masturbatory movements in boys are usually obvious to the observer owing to direct genital manipulation. In girls, they are more subtle and often involve adduction of the thighs or sitting on a hand or foot and rocking. When the movements are accompanied by posturing of the limbs, they are often mistaken for paroxysmal dystonia. Several characteristic features of masturbating girls who present for diagnosis have been identified: (1) onset after 2 months of age and before 3 years of age, (2) stereotyped posturing with pressure applied to the pubic area, (3) quiet grunting, diaphoresis, or facial flushing, (4) episode duration of less than a minute to several hours, (5) no alteration of consciousness, (6) normal findings on examination, and (7) cessation with distraction or engagement of the child in another activity. Unnecessary diagnostic tests are commonly performed before the true nature of the behavior is recog-
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Suggested Readings


Practitioner and Patient Resources

Dystonia Medical Research Foundation
One East Wacker Drive, Suite 2430
Chicago, IL 60601-1905
Phone: (312) 755-0198
In Canada: 800-361-8061
Fax: (312) 803-0138
E-mail: dystonia@dystonia-foundation.org
http://www.dystonia-foundation.org/

The mission of the Dystonia Medical Research Foundation is to advance research for more treatments and ultimately a cure; to promote awareness and education; and to support the needs and well being of affected individuals and families. The Web site features specific pages for patients, physicians, and researchers.