

CHAPTER 180

Headache and Migraine

ETIOLOGY AND EPIDEMIOLOGY

Headache is a common symptom among children and adolescents. Headaches can be a primary problem such as migraines or tension-type headaches. Secondary headaches are most often associated with minor illnesses such as upper respiratory infections or sinusitis but may be the first symptom of serious conditions (e.g., meningitis, brain tumors), so a systematic approach is necessary.

CLINICAL MANIFESTATIONS

The temporal pattern (acute, recurrent episodic, chronic progressive, chronic nonprogressive) is useful in the differential diagnosis of headache (Table 180.1).

Tension-type headaches are the most common type of recurrent primary headaches in children and adolescents. Because they are generally mild, without associated symptoms, they typically do not disrupt daily activities. The pain is global and squeezing or pressing in character and can last for hours or days. There is no associated nausea, vomiting, phonophobia, or photophobia. Headaches can be related to environmental stresses or symptomatic of underlying anxiety or depression.

Migraine headaches are another common type of recurrent headaches and frequently begin in childhood. Headaches are stereotyped attacks of frontal, bitemporal, or unilateral, moderate to severe, pounding or throbbing pain that are aggravated by activity and last 1–72 hours. Associated symptoms include nausea, vomiting, pallor, photophobia, phonophobia, and an intense desire to seek a quiet, dark room for rest. Toddlers may be unable to verbalize the source of their discomfort and exhibit episodes of irritability, sleepiness, pallor, and/or vomiting. An aura can precede or coincide with the headache and typically persists between 5 and 60 minutes. Visual auras are common and consist of spots, flashes, or lines of light that flicker in one or both visual fields. Complex, atypical auras (e.g., hemiparesis, monocular blindness, vertigo, confusion) that are accompanied by a headache warrant careful diagnostic investigation, such as neuroimaging and electroencephalogram.

Common causes of **secondary headaches** include head trauma, viral illness, and sinusitis. Medication-overuse headaches may complicate primary and secondary headaches. Serious causes of secondary headache include increased intracranial pressure (ICP) caused by a mass (e.g., tumor, vascular malformation) or intrinsic increase in pressure (pseudotumor cerebri). Increased ICP should be suspected if the headache and associated vomiting are worse when lying down or on first awakening, if they awaken the child from sleep, if they remit on arising, or if they are exacerbated by coughing, Valsalva maneuver, or bending over. Papilledema (Fig. 180.1) or focal neurologic deficits should trigger an intensive etiologic investigation.

DIAGNOSTIC STUDIES

For most children, a thorough history and physical examination provide an accurate diagnosis and obviate the need

TABLE 180.1 Four Temporal Patterns of Childhood Headache

Acute: Single episode of pain without a history of such episodes. The “first and worst” headache raises concerns for aneurysmal subarachnoid hemorrhage in adults, but is commonly due to febrile illness related to upper respiratory tract infection in children. Regardless, more ominous causes of acute headache (hemorrhage, meningitis, tumor) must be considered.

Acute recurrent: Pattern of attacks of pain separated by symptom-free intervals. Primary headache syndromes, such as migraine or tension-type headache, usually cause this pattern. Recurrent headaches are occasionally due to specific epilepsy syndromes (benign occipital epilepsy), substance abuse, or recurrent trauma.

Chronic progressive: Implies a gradually increasing frequency and severity of headache. The pathologic correlate is increasing ICP. Causes of this pattern include pseudotumor cerebri, brain tumor, hydrocephalus, chronic meningitis, brain abscess, and subdural collections.

Chronic nonprogressive or chronic daily: Pattern of frequent or constant headache. Chronic daily headache generally is defined as >3-mo history of ≥ 15 headaches/mo. Affected patients have normal neurologic examinations; psychologic factors and anxiety about possible underlying organic causes are common.

ICP, Intracranial pressure.

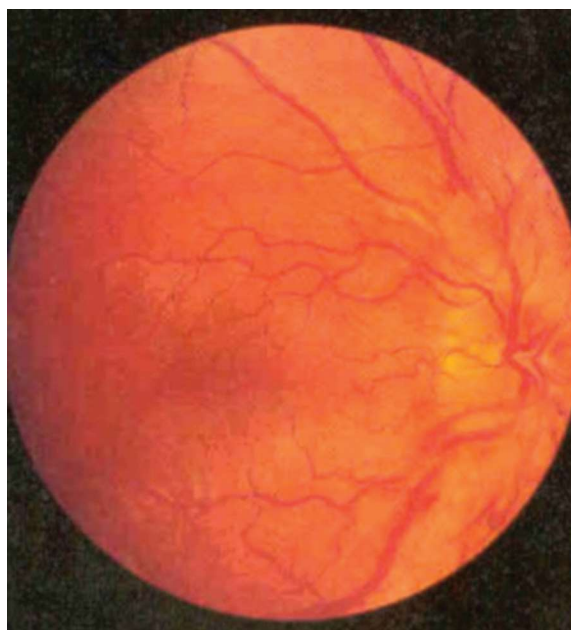


FIGURE 180.1 Papilledema with dilation of the vessels, obliteration of the optic cup, loss of disc margin, and hemorrhages around disc. From Kliegman RE, Behrman RM, Jenson HB, eds. *Nelson Textbook of Pediatrics*. 18th ed. Philadelphia: Saunders; 2007:2107.

for further testing. Neuroimaging is usually not necessary. Imaging is warranted, however, if the patient has an abnormal neurologic examination, symptoms of increased ICP, unusual neurologic features during the headache (atypical aura), or progressive worsening of headaches. In these cases, brain magnetic resonance imaging is the study of choice, providing the highest sensitivity for detecting posterior fossa lesions and other, more subtle abnormalities. When the headache has a

sudden, severe onset, emergent computed tomography (CT) can quickly evaluate for intracranial bleeding. If the CT is negative, a lumbar puncture should be performed to measure opening pressure and evaluate for pleocytosis, elevated red blood cells, and xanthochromia to evaluate for a subarachnoid bleed.

TREATMENT

Treating migraines requires an individually tailored regimen to address the frequency, severity, and disability produced by the headaches. Intermittent symptomatic (abortive) analgesics are the mainstay for treatment of infrequent episodes of migraine. Symptomatic therapy requires early analgesic administration, often accompanied by rest in a quiet, dark room. Acetaminophen or a nonsteroidal antiinflammatory drug such as ibuprofen or naproxen sodium is often effective. Hydration and antiemetics are useful adjunctive therapies. If these first-line medications are insufficient, triptan agents may be considered. Triptans, available in injectable, nasal spray, oral disintegrating, and tablet form, are serotonin receptor agonists that can alleviate migraine symptoms promptly. Based on theoretical concerns regarding increased risk for stroke, it is generally recommended that triptans be avoided for patients with hemiplegic migraine or migraine with brainstem aura.

Many children suffer severe and frequent migraines that disrupt their daily lives. Before initiating daily medications, lifestyle modifications including increasing fluid intake, avoiding skipping meals, engaging in good sleep hygiene, and exercising regularly, as well as identification and elimination of any precipitating or aggravating influences (e.g., caffeine, certain foods, stress, missed meals, dehydration), are beneficial. Other adjunctive treatment options include psychologic support, stress management, and biofeedback. Although there is limited data on the effectiveness of nutraceuticals, riboflavin and magnesium may be helpful for migraine prophylaxis with a low side-effect profile. Children with more than one disabling headache per week may require **daily preventive agents** to reduce both attack frequency and severity. Preventive medications include tricyclic antidepressants (e.g., amitriptyline, nortriptyline), anticonvulsants (e.g., topiramate, valproic acid), antihistamines (e.g., cyproheptadine), β blockers (e.g., propranolol), and calcium channel blockers (e.g., verapamil). The use of calcitonin gene-related peptide receptor antagonists is currently being studied in children.

PEARLS FOR PRACTITIONERS

See *Neurology: Pearls for Practitioners* at the end of this section.

TABLE 181.1 Paroxysmal Disorders of Childhood

Seizures/epilepsy
Migraine and variants • Paroxysmal torticollis of infancy • Benign paroxysmal vertigo • Cyclic vomiting
Syncope • Vasovagal syncope • Breath-holding spells • Cardiac syncope (arrhythmias, long QT syndrome)
Transient ischemic attack
Metabolic disorders • Hypoglycemia • Inborn errors of metabolism
Sleep disorders • Narcolepsy, cataplexy • Night terrors
Paroxysmal dyskinesias and other movement disorders • Tics • Stereotypies
Shudder attacks
Sandifer syndrome (gastroesophageal reflux causing paroxysmal stiffening and posturing)
Functional neurologic symptom disorder
Normal behavior • Neonatal jitteriness • Benign sleep myoclonus • Daydreaming • Self-stimulatory behavior

ETIOLOGY AND EPIDEMIOLOGY

The differential diagnosis of paroxysmal disorders includes **seizures** and **nonepileptic events** (Table 181.1). Nonepileptic paroxysmal events are often normal or benign phenomena, although some are clinically significant and consequential. A thorough medical history from the patient and primary witnesses is the most reliable tool for establishing the correct diagnosis.

Acute symptomatic seizures are secondary to an acute problem affecting brain excitability, such as electrolyte imbalance or infection (Table 181.2). In the classification of seizures, “symptomatic” refers to seizures that are temporally correlated with an acute neurologic or systemic insult. In contrast, **epilepsy** is defined as recurrent, unprovoked seizures. Epileptic seizures are generally classified as focal, arising from one region of the cortex, or generalized, which arise from both hemispheres simultaneously (Table 181.3). Approximately 4–10% of children experience at least one seizure. The incidence of childhood epilepsy is 1–2%.

FOCAL SEIZURES

Clinical symptoms of focal seizures may include motor (tonic, clonic, myoclonic), sensory, psychic, or autonomic abnormalities.

Focal seizures with retained awareness (previously termed *simple partial seizures*) arise from a specific anatomic focus and may or may not spread to surrounding brain regions. The location and extent of spread of the seizure focus determine the clinical symptoms.

CHAPTER 181

Seizures

A seizure is a transient occurrence of signs or symptoms resulting from abnormal excessive or synchronous neuronal activity in the brain.

TABLE 181.2 Causes of Seizures

PERINATAL CONDITIONS
Malformation of cortical development
Intrauterine infection
Hypoxic-ischemic encephalopathy*
Trauma
Hemorrhage*
INFECTIONS
Encephalitis*
Meningitis*
Brain abscess
METABOLIC CONDITIONS
Hypoglycemia*
Hypocalcemia
Hypomagnesemia
Hyponatremia
Hypernatremia
Storage diseases
Degenerative disorders
Porphyria
Pyridoxine dependency and deficiency
POISONING
Lead
Drug toxicity
Drug withdrawal
NEURO CUTANEOUS SYNDROMES
Tuberous sclerosis
Neurofibromatosis
Sturge-Weber syndrome
Incontinentia pigmenti
SYSTEMIC DISORDERS
Vasculitis
Systemic lupus erythematosus
Hypertensive encephalopathy
Hepatic encephalopathy
OTHER CAUSATIVE DISORDERS/CONDITIONS
Febrile illnesses*
Accidental trauma*
Child abuse*
Familial*
Cryptogenic* (no underlying disorder found)
Increased intracranial pressure
Tumor
Remote brain injury

*Common.

TABLE 181.3 Classification of Seizures and Epilepsy Syndromes

FOCAL ONSET
With retained awareness (focal aware seizures)
Motor onset (can be tonic, clonic, myoclonic, atonic, or epileptic spasms)
Nonmotor onset (e.g., autonomic, visual, auditory, olfactory, gustatory, vertiginous, sensory)
With impaired awareness (dyscognitive features)
Motor or nonmotor onset (see above)
Focal to bilateral tonic-clonic seizures
GENERALIZED ONSET
Motor (can be tonic, clonic, myoclonic, atonic, or epileptic spasms)
Nonmotor (absence)
UNKNOWN ONSET
Motor (can be tonic-clonic, epileptic spasms)
Nonmotor (behavioral arrest)

Focal seizures with impaired awareness (previously termed *complex partial seizures*) can have similar sensorimotor signs but also have associated alteration of consciousness. Although the child may not be completely unresponsive, subtle slowing or alteration of mental status (dyscognitive features) may occur. Along with altered responsiveness, patients may have automatisms or stare during these seizures. Automatisms are automatic semi-purposeful movements of the mouth (lip smacking, chewing) or extremities (rubbing of fingers, shuffling of feet). Focal seizures that manifest only with psychic or autonomic symptoms, such as a feeling of déjà vu during a temporal lobe seizure, can be difficult to recognize.

When focal seizures spread to involve both cerebral hemispheres, they are said to have evolved to a bilateral tonic-clonic seizure (previously called a *secondarily generalized seizure*). It is often difficult to distinguish primary generalized tonic and clonic seizures from those that evolved from focal seizures on purely clinical grounds. Clarifying any premonitory symptoms suggestive of an aura can be helpful. This distinction is important because evaluation and treatment for focal or generalized epilepsy syndromes are quite different.

GENERALIZED SEIZURES

Tonic, clonic, and biphasic **tonic-clonic** seizures may occur alone or in association with other seizure types. Typically, the seizure begins abruptly but occasionally is preceded by a series of myoclonic jerks. During a tonic-clonic seizure, consciousness and control of posture are lost, followed by tonic stiffening and upward deviation of the eyes. Pooling of secretions, pupillary dilation, diaphoresis, and hypertension are common. Clonic jerks follow the tonic phase. In the postictal phase, the child might be hypotonic. Irritability and headache are common as the child awakens.

TABLE 181.4 Differentiating Absence Seizures and Focal Seizures with Impaired Awareness

FEATURE	ABSENCE SEIZURE	FOCAL SEIZURE WITH IMPAIRED AWARENESS
Duration	Seconds	Minutes
Provoking maneuver	Hyperventilation Photoc stimulation	Variable, but often none
Postictal phase	None (return immediately to baseline)	Confusion, sleepiness
Number of seizures	Many per day	Infrequent (rarely >1/day)
EEG features	Interictal: normal except bursts of generalized spike wave and sometimes occipital intermittent rhythmic delta activity Ictal: 3-Hz generalized spike-wave	Interictal: normal or focal slowing, sharp waves, or spikes Ictal: focal discharges (with or without spread to contiguous regions or the contralateral hemisphere)
Neurologic examination	Normal	Normal, or focal deficits
Neuroimaging	Normal*	Normal, or focal abnormalities (mesial temporal sclerosis, focal cortical dysplasia, neoplasm, encephalomalacia)
First-line treatment	Ethosuximide or valproic acid [†]	Often levetiracetam, oxcarbazepine, or lamotrigine are used

*In the proper clinical context, and with an appropriate EEG, a diagnosis of absence epilepsy should be made without neuroimaging.

[†]Oxcarbazepine and carbamazepine are relatively contraindicated for children with typical absence epilepsy, as their seizures can be exacerbated by these medications.

EEG, Electroencephalographic.

Absence Seizures

Seizures in which the primary clinical feature is staring can be either absence (generalized) or focal seizures. The clinical hallmark of absence seizures is a brief (<15 seconds) loss of awareness accompanied by eyelid fluttering or simple automatisms, such as fumbling with the fingers and lip smacking. Absence seizures usually begin between 4 and 6 years of age. Neurologic examination and brain imaging are normal. The characteristic electroencephalographic (EEG) patterns consist of **generalized 3-Hz spike-and-wave activity**. An absence seizure can often be provoked by **hyperventilation**. Differentiating absence from focal seizures with impaired awareness can be difficult but is essential for appropriate evaluation and treatment (Table 181.4).

Atypical Absence, Myoclonic, and Atonic Seizures

Atypical absence seizures manifest as episodes of impaired consciousness with automatisms, autonomic phenomena,

and motor manifestations, such as eye opening, eye deviation, and body stiffening. They are associated with slower EEG discharges (2 Hz) and accompany other seizure types. **Myoclonus** is a sudden jerk of all or part of the body; not all myoclonus is due to epilepsy. Nonepileptic myoclonus may be benign, as in sleep myoclonus, or indicate serious disease. Myoclonic epilepsy usually is associated with multiple seizure types. The underlying disorder producing myoclonic epilepsy may be static (e.g., **juvenile myoclonic epilepsy** [JME]) or progressive and associated with neurologic deterioration (e.g., neuronal ceroid lipofuscinosis). **Myoclonic absence** refers to body jerks that commonly accompany typical or atypical absence seizures. Atonic seizures involve a sudden loss of postural tone that often results in falls and injuries; though typically brief (lasting 1–2 seconds), they are very disabling.

Febrile Seizures

Seizures in the setting of fever may be caused by central nervous system infections (meningitis, encephalitis, brain abscess), first presentation of epilepsy triggered by fever, or **febrile seizures**. The latter represents the most common cause of seizures among children between 6 months and 5 years of age, occurring in about 4% of all children. By definition, a febrile seizure occurs in the presence of fever. **Simple febrile seizures** are generalized at onset, last less than 15 minutes, and occur only once in a 24-hour period. If the seizure has focal features, lasts longer than 15 minutes, or recurs within 24 hours, the seizure is referred to as a **complex febrile seizure**.

The prognosis of children with simple febrile seizures is excellent. Although febrile seizures recur in 30–50% of children, intellectual achievements are normal. The risk of subsequent epilepsy is not substantially greater than that of the general population (~2%). Factors that increase the risk for the development of epilepsy include abnormal neurologic examination or development, family history of epilepsy, and complex febrile seizures.

Since simple febrile seizures are brief and the outcome is benign, most children require no treatment. Rectal diazepam can be administered during a seizure to abort a prolonged convulsion; it is appropriate to provide a rescue medication for children with a history of prolonged febrile seizures. Since antiseizure medications have side effects and children with febrile seizures have an excellent prognosis, daily antiseizure medication to prevent febrile seizures is not recommended. Administration of antipyretics during the same fever episode may reduce the risk of recurrence within that illness, but it does not seem to impact febrile seizure recurrence during subsequent fever episodes.

Psychogenic Nonepileptic Events

Psychogenic nonepileptic events (PNEE) (previously termed *pseudoseizures*) may be the manifestation of functional neurologic symptom disorder (i.e., somatic symptom disorder or conversion disorders). Additionally, children with epilepsy may, consciously or subconsciously, exhibit concurrent PNEE. PNEE semiology typically differs from epileptic seizures. In PNEE, the patient's eyes are often closed and the movements are tremulousness or thrashing. In contrast, during seizures, the eyes are typically open and movements are tonic or clonic.

Verbalization and pelvic thrusting are seen more commonly in PNEE, urinary and fecal continence is usually preserved, and injury does not usually occur. If the tongue is bitten, more often the tip of the tongue is injured (as opposed to the sides of the tongue in epileptic seizures). PNEE can often be initiated or terminated by suggestion. An EEG performed during PNEE does not show epileptiform patterns. The diagnosis is critical because appropriate mental health care, and not anti-convulsant medications, is the crux of treatment. Careful assessment and intervention for physical or sexual abuse, as well as other social stressors, is a key component of the care of children with PNEE.

EPILEPSY SYNDROMES

Epilepsy syndromes have distinctive EEG and clinical features (age of seizure onset, specific seizure types). Recognition of epilepsy syndromes provides useful information regarding the underlying etiologies that must be considered, treatments that may be helpful (or harmful), and long-term prognostic information. Comorbid learning difficulties, especially language-based learning disabilities, and attention-deficit/hyperactivity disorder (ADHD) are common. There is no contraindication to treating these children's ADHD with stimulant medication.

Childhood epilepsy with centrotemporal spikes, also known as **benign Rolandic epilepsy**, is among the most common epilepsy syndromes and usually begins between ages 5 and 10 years. The seizures typically occur only during sleep or on awakening. Affected children usually have focal motor seizures involving the face and arm (abnormal movement or sensation around the face and mouth, drooling, rhythmic guttural sound, impaired speech and swallowing). Seizures sometimes evolve to bilateral convulsions. The interictal EEG classically demonstrates independent bilateral centrotemporal sharp waves but is otherwise normal. With a classic history and EEG and a normal neurologic examination, the diagnosis can be made with confidence and neuroimaging is not required. Seizures usually respond promptly to antiseizure medication. Intellectual outcome is typically normal and the epilepsy resolves after puberty.

Childhood absence epilepsy is another common syndrome. Absence seizures typically begin in the early school years and usually resolve by late childhood or adolescence. If absence does not remit, nearly half will go on to develop JME (see later discussion). Ethosuximide is usually the first-choice medication. A subset of patients also has generalized tonic-clonic seizures. For these children, valproic acid is the first choice as it can prevent both absence and convulsive seizures.

JME is the most common generalized epilepsy among adolescents and young adults. Onset is typically in early adolescence with myoclonic jerks (exacerbated in the morning, often causing the patient to drop objects), generalized tonic-clonic seizures, and absence seizures. Seizures usually resolve promptly with antiseizure medication. JME is classically treated with valproic acid; however, there are concerns about side effects and teratogenicity. Levetiracetam has a much lower risk of teratogenicity and is generally well tolerated. As such, levetiracetam is now favored, particularly for girls and women since treatment of JME is usually lifelong.

Infantile spasms are brief contractions of the neck, trunk, and arm muscles, followed by a phase of sustained muscle contraction lasting less than 2 seconds. Spasms occur most frequently when the child is awakening or going to sleep. Each jerk is followed by a brief period of relaxation with repeated spasms in clusters of variable duration. When flexion of the thighs and crying are prominent, the syndrome may be mistaken for colic or gastroesophageal reflux. **West syndrome** is the triad of infantile spasms, developmental regression, and a dramatically abnormal EEG pattern (**hypsarrhythmia**—a pattern of chaotic high-voltage slow waves, spikes, and polyspikes). The peak age at onset of infantile spasms is 3–8 months. The underlying etiology of the spasms dictates the prognosis. More than 200 different etiologies have been identified, including tuberous sclerosis, malformations of cortical development (e.g., lissencephaly), genetic syndromes (e.g., trisomy 21), acquired brain injury (e.g., stroke, perinatal hypoxic-ischemic encephalopathy), and metabolic disorders (e.g., phenylketonuria). Infants for whom an etiology is determined are classified as having “symptomatic” infantile spasms and are at very high risk for long-term neurodevelopmental difficulties. With advances in genetic testing, few children have infantile spasms of uncertain etiology, but those with uncertain etiology have a somewhat better long-term prognosis.

Infantile spasms are a neurologic emergency since rapid initiation of effective therapy offers the best chance for favorable neurodevelopmental outcomes. First-line treatment options for infantile spasms include adrenocorticotrophic hormone, high-dose oral corticosteroids, and vigabatrin. Other antiseizure medications are much less effective. For infants with underlying tuberous sclerosis, vigabatrin is the treatment of choice. For other patients, treatment determinations are made on a case-by-case basis.

Lennox-Gastaut syndrome is a severe epilepsy syndrome with variable age of onset although most children present before age 5 years. Frequent, multiple seizure types including atonic, focal, atypical absence, and generalized tonic, clonic, or tonic-clonic semiologies characterize the disorder. Many children have underlying brain injury, malformations, or genetic etiologies. The seizures are typically difficult to control, and most patients have significant intellectual disability.

Benign (self-limited) neonatal convulsions are an autosomal dominant genetic disorder linked to abnormal neuronal potassium channels. Otherwise well newborns present with focal seizures toward the end of the first week of life, leading to the colloquial term *fifth-day fits*. Response to treatment is generally excellent, and the long-term outcome is typically favorable.

Acquired epileptic aphasia (Landau-Kleffner syndrome) is characterized by the abrupt loss of previously acquired language in young children. The language disability is an acquired cortical auditory deficit (auditory agnosia). The EEG is highly epileptiform in sleep, the peak area of abnormality often being in the dominant perisylvian region (language areas). This diagnosis should be considered for young patients with clear language regression, as it is a potentially treatable entity.

STATUS EPILEPTICUS

Status epilepticus is a neurologic emergency defined as (1) ongoing seizure activity for 5 or more minutes or (2) repetitive

seizures without recovery of consciousness. Status epilepticus carries an approximately 14% risk of new neurologic deficits and 4–5% mortality, both related to the underlying etiology. Etiologies include new-onset epilepsy, drug intoxication, drug withdrawal (especially missed anticonvulsant doses among children with epilepsy), hypoglycemia, electrolyte imbalance, acute head trauma, infection, ischemic stroke, intracranial hemorrhage, metabolic disorders, and hypoxia.

The first priority of treatment is to ensure an adequate airway, breathing, and circulation (see Chapter 38). Vital signs should be obtained and oxygen administered as needed. If respirations are inadequate, positive pressure ventilation may be required. Intravenous (IV) access should be obtained, and laboratory evaluation should be undertaken (see later).

Several pharmacologic options exist for management of status epilepticus (Table 181.5). Initial management is with a benzodiazepine and should be administered if the seizure persists longer than 5 minutes. Lorazepam, diazepam, and midazolam are all effective agents and should be given as an adequate single dose (rather than successive smaller doses), which can be repeated one time. IV administration is preferred. When IV access is not available, benzodiazepines may be administered via rectal, intranasal, buccal, or intramuscular routes.

If the seizure does not resolve after two doses of benzodiazepine, a second-line agent must be administered. There are few data from clinical trials to guide subsequent management. For newborns and infants, phenobarbital is reasonable. For older children, IV fosphenytoin (preferred over phenytoin due to tolerability), valproic acid, or levetiracetam may be considered. If the seizures persist, options include repeating one of the second-line medications or beginning a continuous infusion of midazolam or pentobarbital (with appropriate airway protection, blood pressure support, and strong consideration of video-EEG monitoring to evaluate for ongoing subclinical seizures) (see Table 181.5). If this approach is ineffective, preparations for general anesthesia are undertaken. When status epilepticus stops, maintenance therapy is initiated with the appropriate anticonvulsant.

LABORATORY AND DIAGNOSTIC EVALUATION

Children with simple febrile seizures who have recovered completely require little or no laboratory evaluation other than studies necessary to evaluate the source of the fever. Outside of simple febrile seizures, it is important to consider possible provoking factors for a first lifetime seizure, such as meningitis, sepsis, head trauma, metabolic derangements (including glucose, sodium, and calcium abnormalities), and toxins. Laboratory evaluation may include a complete blood count and measurement of glucose, calcium, sodium, potassium, chloride, bicarbonate, urea nitrogen, creatinine, magnesium, and phosphorus, as well as blood or urine toxicology screening. For an otherwise healthy child with a single self-resolved seizure and a normal examination, in the absence of history to suggest a provoking etiology, laboratory evaluation may not be necessary. Children with clinical signs and symptoms of meningitis (e.g., neck stiffness, Kernig sign, Brudzinski sign), or history or physical examination suggestive of intracranial infection, should undergo a lumbar puncture. Cerebrospinal

TABLE 181.5 Management of Status Epilepticus

STABILIZATION
ABCs (airway, breathing, circulation)
Cardiac monitoring
Oxygen and pulse oximetry
Intravenous access
Immediate laboratory tests
Glucose
Basic metabolic panel—sodium, calcium, magnesium
Anticonvulsant drug levels
Toxicology studies as appropriate
Complete blood counts, platelets, and differential
PHARMACOLOGIC MANAGEMENT
Benzodiazepine
Lorazepam IV, IN
Diazepam IV Rectal diazepam
Midazolam IV, IM, IN, buccal
Standard anticonvulsant medications
Fosphenytoin IV, IM
Phenobarbital IV
Levetiracetam IV
Valproic acid IV
Continuous infusions
Pentobarbital
Midazolam
General anesthesia

IM, Intramuscularly; IN, intranasally; IV, intravenous.

fluid (CSF) should be analyzed for cell counts, culture, protein, and glucose levels. In children less than 18 months old, particularly young infants, the clinical symptoms of meningitis may be subtle. Neonates also may require testing for inborn errors of metabolism, blood ammonia, CSF glycine and lactate, and a clinical trial of pyridoxine. Evaluation for infections, such as urine and stool cultures, and polymerase chain reactions for herpes simplex virus, cytomegalovirus, and enterovirus should be considered.

The EEG is the most useful neurodiagnostic test for distinguishing seizures from nonepileptic paroxysmal disorders and for classifying seizures as having focal or generalized onset. The EEG must be interpreted in the context of the clinical history; some normal children have focal or epileptiform EEG patterns. Conversely, children with seizures may have normal interictal EEGs. When the diagnosis is unclear, EEG with prolonged recordings and simultaneous

videomonitoring in an attempt to capture a typical event may be necessary.

Magnetic resonance imaging (MRI) is superior to computed tomography (CT) in showing most brain pathology, but in the emergency department setting, CT can be performed rapidly and often shows acute intracranial hemorrhage more clearly than MRI. MRI is unnecessary in patients with the primary generalized epilepsies, such as typical childhood absence and JME. Lesions (e.g., tumors, arteriovenous malformations, cysts, strokes, gliosis, focal atrophy) may be identified in 25% of patients with other epilepsy types, even when the clinical examination and EEG do not suggest focal features. Identification of some lesions, such as focal cortical dysplasia, hamartoma, and mesial temporal sclerosis, can assist in consideration of surgical treatment of pharmacoresistant epilepsy.

LONG-TERM THERAPY

The decision to institute daily seizure medications for a first unprovoked seizure is based on the likelihood of recurrence balanced against the risk of long-term drug therapy. Determination of the recurrence risk is based on the clinical history and neurodiagnostic testing. Absence seizures, infantile spasms, atypical absence seizures, and atonic seizures are universally recurrent at the time of diagnosis, so treatment should be initiated immediately.

The goal of treatment is to maintain an optimal functional state. Medication toxicity should be weighed against the risk of the seizures. Initial drug selection is based on the seizure type (Fig. 181.1). Using a single agent limits toxicity, contains cost, and improves compliance. Approximately 60% of children obtain satisfactory seizure control and minimal side effects with the initial drug. If seizure control is not achieved, despite good compliance, addition of a second drug is considered.

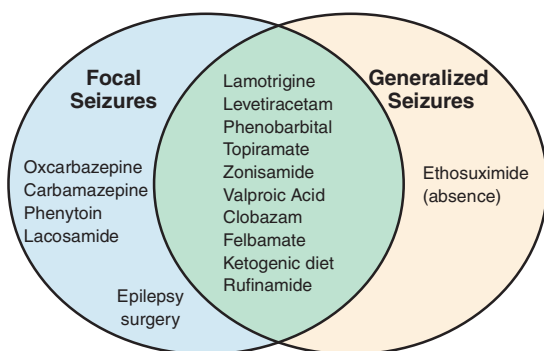
The duration of antiseizure treatment varies according to seizure type and epilepsy syndrome. For most children, medications can be weaned off after 2 years without seizures.

There are some exceptions. For example, children with JME, progressive myoclonic epilepsy, atypical absence seizures, and Lennox-Gastaut syndrome usually require lifelong treatment. Children who have other neurologic comorbidities, seizures that were initially difficult to control, or persistently epileptiform EEGs are at highest risk for recurrence when therapy is discontinued.

There are important safety considerations for people with epilepsy. The risk of drowning is high, so swimming and bathing must only occur under direct adult supervision. Children should use appropriate helmets for sports such as bicycling or ice skating. There is no contraindication to participation in contact sports, but scuba diving, hang gliding, and free climbing are not safe for people with epilepsy. Each country and individual states have specific laws regarding driving for people with epilepsy. Most require a period of seizure freedom before driving.

PEARLS FOR PRACTITIONERS

See *Neurology: Pearls for Practitioners* at the end of this section.



Special circumstances:

1. For absence epilepsy, ethosuximide is the first line treatment.
2. Adrenocorticotrophic hormone, oral corticosteroids, or vigabatrin are used to treat infantile spasms.

FIGURE 181.1 Treatment for epilepsy.