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**Epileptic encephalopathies of infancy and childhood**

Epileptic encephalopathies are severe brain disorders that are characterized by: (1) electrographic paroxysmal, often aggressive, activity in the EEG; (2) seizures, usually polymorphic and resistant; (3) cognitive, behavioral, and neurological deficits, often persistent; and (4) sometimes death.

The concept of epileptic encephalopathies is based on the assumption that aggressive ictal (attack) and electrical (edectrographic) epileptogenic activity during the period of brain maturation is the main causative factor of progressive cognitive and neuropsychological deterioration or regression. On the other hand, this destructive epileptic activity is a specific age-dependent reaction of the brain (excessive neocortical excitability) to various pathological conditions, which can be diffuse or focal, symptomatic or idiopathic in etiology. This age-dependent epileptogenic reaction is inherent in the immature brain, its severity largely depends on the stage of maturation at the time of debut. Thus, in the neonatal period, EEG abnormalities are most often represented by a burst-suppression pattern, in infancy - by hypsarrhythmia, and slow generalized spike-wave discharges in early childhood. Accordingly, seizures and electrographic epileptogenic signs evolve with age, for example, the burst-suppression pattern may change to hypsarrhythmia, then to slow spike-wave discharges. All epileptic encephalopathies are characterized by a weakening or cessation of further progression at puberty, but often with severe residual neurocognitive deficit.

The following syndromes of epileptic encephalopathy with onset in neonatal, infancy or early childhood are distinguished:

* [Dravet syndrome (severe myoclonic epilepsy of infancy)](https://epidoc.ru/syndromes/encephalopathies/dravet_syndrome.html)
* [early myoclonic encephalopathy](https://epidoc.ru/syndromes/neonatal/early_myoclonic_encephalopathy.html)
* [epilepsy with continuous spike-waves of slow sleep](https://epidoc.ru/syndromes/encephalopathies/csws.html)
* [hypothalamic (gelastic) epilepsy](https://epidoc.ru/syndromes/encephalopathies/hypothalamic_epilepsy.html)
* [Landau-Kleffner syndrome](https://epidoc.ru/syndromes/encephalopathies/landau_kleffner.html)
* [Lennox-Gastaut syndrome](https://epidoc.ru/syndromes/encephalopathies/lennox_gastaut_syndrome.html)
* [myoclonic status in non-progressive encephalopathies](https://epidoc.ru/syndromes/encephalopathies/myoclonic_status.html)
* [Ohtahara syndrome](https://epidoc.ru/syndromes/neonatal/ohtahara_syndrome.html)
* [West syndrome](https://epidoc.ru/syndromes/encephalopathies/west_syndrom.html)

# Severe myoclonic epilepsy of infancy (Dravet syndrome)

**Prevalence**  
~ 6% of epilepsies that begin before age 3.

**Morbidity**  
~ 1/30,000 births.

**Age of disease onset**  
The first year of life with a peak at 5 months.

**Floor**  
Boys predominate slightly (66%).

**Neurological and mental status**  
Normal before the attacks started.

**Etiology**  
Mainly genetic - with mutations in the sodium channel genes. This is the most severe phenotype of autosomal dominant epilepsy with FS+.

**Clinical manifestations**  
Seizure tetrad: (1) early infantile febrile clonic seizures; (2) myoclonic seizures; (3) atypical absences; and (4) complex focal seizures. Three periods of evolution: (1) relatively mild with clonic seizures, usually during fever; (2) an increasingly aggressive period with numerous myoclonus, atypical absences, complex focal seizures, and status epilepticus that lasts for hours or days; (3) a static period when the seizures subside but severe residual mental and neurological impairment remains permanently. Seizures most often occur on awakening, activation, and in the waking state; seizures during sleep are uncommon (3%).

**Provoking factors**  
Diseases with high body temperature, hot baths, light stimulation (including pattern stimulation), movements, closing the eyes.

**Diagnostic procedures**  
MRI shows mild and nonspecific abnormalities. Metabolic tests, muscle and skin biopsy are normal; mitochondrial cytopathy is uncommon.

**Interictal EEG**  
Slowing of background rhythms, with frequent generalized polyspike-slow-wave and 2 Hz spike-slow-wave complexes. Multifocal disturbances in the form of spikes and slow oscillations are often recorded.

**Ictal EEG**  
Depends on the type of attack

**Forecast**  
Adverse, persistent mental and neurological deficits, often fatal.

**Differential diagnostics**  
[Febrile seizures](https://epidoc.ru/syndromes/idiopathic_epi_infancy/febrile_seizures.html), syndrome[Lennox-Gastaut](https://epidoc.ru/syndromes/encephalopathies/lennox_gastaut_syndrome.html), epilepsy with[myoclonic-astatic seizures](https://epidoc.ru/syndromes/ige/doose_syndrome.html), benign[myoclonic epilepsy of infancy](https://epidoc.ru/syndromes/idiopathic_epi_infancy/benign_myoclonic_infancy.html),[progressive myoclonus epilepsy](https://epidoc.ru/syndromes/pme/index.html).

**Treatment**  
Attacks are not controlled by AEDs. Carbamazepine, phenytoin and lamotrigine are contraindicated. Early treatment of infectious diseases and hyperthermia (factors that provoke status epilepticus) is mandatory.

# Early myoclonic encephalopathy

**Prevalence**  
Unknown. Approximately 100 cases have been described, but this figure is undoubtedly an underestimate, and many cases of this severe and fatal infant disease are simply not diagnosed.

**Age of disease onset**  
The first hours and days of life.

**Floor**  
Boys and girls suffer equally.

**Neurological and mental status**  
Violated.

**Etiology**  
Multifactorial disease. Inborn errors of metabolism are the most common cause (nonketotic hyperglycinemia, propionic aciduria, methylmalonic acidemia, D-glyceric acidemia, sulfite-xanthine oxidase deficiency, Menkes disease, Zellweger syndrome, molybdenum cofactor deficiency). Metabolic causes explain the high incidence in siblings. Structural brain disorders are rare.

**Clinical manifestations**  
Triad of intractable seizures - initially chaotic myoclonus appears, followed by simple focal seizures and then tonic epileptic (infantile) spasms.

Myoclonus is chaotic, moving from one part of the body to another in a random order. Often they are limited to a small area - a finger, tongue, eyelids, lips, repeated in the same muscle group, then migrate to another part of the body. Myoclonus is short, single or repeated, very frequent, almost continuous. Massive, usually bisynchronous, axial myoclonic jerks can occur at the onset of the disease or join later, often interspersed with chaotic myoclonus.

Simple focal seizures (eye deviation, autonomic symptoms such as facial flushing, apnea), focal clonic seizures of any part of the body, asymmetric tonic posture of the trunk. Tonic seizures are frequent; epileptic spasms occur less frequently and usually in the late stages of the disease.

**Diagnostic procedures**  
Metabolic screening is mandatory, mainly measuring serum amino acids, particularly glycine and glycerol metabolites, organic acids, and amino acids in the cerebrospinal fluid. Neuroimaging is initially normal, but may subsequently reveal signs of atrophy.

**Interictal EEG**  
Repetitive burst-suppression pattern, physiological rhythms are absent. Three to four months after the onset of the disease, the EEG pattern evolves into a pattern of atypical hypsarrhythmia or multifocal spikes.

**Ictal EEG**  
Chaotic myoclonus usually does not have a direct correlate in the EEG and may occur after “flashes”.

**Forecast**  
Unfavorable. More than half of patients die within weeks or months of onset, and the rest develop persistent neurological and mental deficits

**Differential diagnostics**  
The main differential diagnosis is with[Ohtahara syndrome](https://epidoc.ru/syndromes/neonatal/ohtahara_syndrome.html).

**Treatment**  
The attacks are resistant to existing therapy.

# Epilepsy with continuous spike-waves of slow-wave sleep (Epilepsy with electrical status epilepticus of slow-wave sleep)

**Prevalence**  
~ 0.2% to 0.5% of all childhood epilepsies.

**Age of disease onset**  
The attacks begin between the ages of 2 months and 12 years, with a peak at 4-5 years. The EEG pattern of continuous spike-wave slow sleep (CSWS) usually develops 1-2 years after the onset of attacks.

**Floor**  
The majority of patients are male (62%).

**Neurological and mental status**  
Violations in more than 1/3 of cases.

**Etiology**  
Symptomatic, probable symptomatic and idiopathic.

**Clinical manifestations**  
Three stages of the disease: - The first stage before ESMS: infrequent nocturnal focal motor seizures, often in the form of epileptic status hemiclonus, absences, atonic, complex focal seizures, and generalized tonic-clonic seizures (GTCS). - The second stage against the background of ESMS: seizures are more frequent, complicated by typical or more often atypical absences, myoclonic absences, epileptic status absences, atonic or clonic seizures, and GTCS. Tonic seizures are uncommon. Noticeable psychomotor deterioration and behavioral disorders. - The third stage (after several months, lasts from 2 to 7 years) with remission of seizures and general improvement.

**Diagnostic procedures**  
MRI abnormalities in more than half of patients. Unilateral or diffuse cortical atrophy, porencephaly, malformations of cortical development, and mainly polymicrogyria (18%).

**Interictal EEG**  
Initially multifocal and bisynchronous sharp waves (mainly in the frontal areas). In the ESMS phase - continuous (from 85% to 100%) mainly bisynchronous with a frequency of 1.5-2 Hz (or 3-4 Hz) spike-waves during NREM sleep.

**Ictal EEG**  
Depends on the type of attacks.

**Forecast**  
In all cases, remission of attacks occurs, cognitive abilities improve, behavioral disorders decrease, but recovery is always slow and often incomplete; only less than a quarter of patients achieve an acceptable level of social adaptation - these are patients with normal premorbid cognitive development and a short period of ESMS

**Differential diagnostics**  
(1) Syndrome[Landau-Kleffner](https://epidoc.ru/syndromes/encephalopathies/landau_kleffner.html); (2) benign childhood focal seizures with atypical evolution; and (3) syndrome[Lennox-Gastaut](https://epidoc.ru/syndromes/encephalopathies/lennox_gastaut_syndrome.html).

**Treatment**  
Attacks in this disease are not the main problem, since remission occurs one way or another. For their treatment, drugs such as valproic acid, lamotrigine, levetiracetam and sultiame can be used. Treatment of ESMS itself, which causes neuropsychological disorders, is empirical and has a temporary effect. The following regimens or combinations are suggested:a. Benzodiazepines orally in combination with valproates.b. ACTH (80 IU/day) or prednisone (2-5 mg/kg per day) with a gradual reduction over 3 months. The earlier the treatment is started, the shorter the duration of steroid use and the better the final result. In cases with severe speech disorders, the surgical technique of subpial transcortical incisions is sometimes successfully used.

# Hypothalamic (gelastic) epilepsy

An epileptic disease - hypothalamic hamartoma - manifests itself with gelastic seizures. Often develops into generalized epileptic encephalopathy with severe seizures, cognitive and behavioral disorders.

**Prevalence**  
An extremely rare disease, approximately 0.1% of all patients with seizures.

**Age of disease onset**  
Neonatal or early childhood, peak at 2-3 years.

**Floor**  
Among boys, it is twice as common.

**Etiology**  
Hypothalamic hamartoma, which has epileptogenic potential and is directly involved in the pathogenesis of gelastic and dacrystic seizures.

**Clinical manifestations**  
Laughter is a characteristic, initial manifestation. The nature of laughter is sad, quiet or loud, and, as a rule, unmotivated. In 13% of patients, dacrystic attacks (crying) are observed. Attacks are usually short (from 10 to 30 seconds), daily. Ictal disturbances of consciousness occur in half of patients. Ictal vegetative symptoms are observed in 1/3. More than half (66%) of patients also suffer from generalized attacks, such as tonic, atonic, generalized tonic-clonic, absences. Also, cognitive and behavioral disorders develop in most patients. At the same time, cognitive and behavioral disorders do not occur in children with hypothalamic hamartoma and early puberty, but without attacks.

**Diagnostic procedures**  
MRI is required to detect hypothalamic hamartoma.

**Interictal EEG**  
It may be normal, but non-specific and non-lateralizing episodic disturbances are more often recorded.

**Ictal EEG**  
Low-voltage fast rhythms with simultaneous suppression of background activity.

**Forecast**  
In most patients, the disease is progressive, with worsening attacks and progressive cognitive and behavioral impairment. More than half (59%) of patients experience early puberty.

**Differential diagnostics**  
Nonepileptic events and gelastic seizures originating from other areas of the brain (temporal and frontal lobes).

**Treatment**  
Drug therapy is often ineffective, and polytherapy may do more harm than good. There is a report of successful use of gonadotropin-releasing hormone (GnRH) with remission of attacks in 2 patients - similar to patients with early puberty. Surgical removal of hamartoma is a technically very complex procedure, but very effective. Complete resection allows achieving complete remission of attacks and prevents the development of persistent behavioral and cognitive disorders.

# Landau-Kleffner syndrome (acquired epileptic aphasia)

**Prevalence**  
Low; 1-2 cases per year in specialized epileptology centers.

**Age of disease onset**  
Usually up to 6 years; peak at 3-6 years.

**Floor**  
Twice as common in boys.

**Neurological and mental status**  
Normal before the attacks started.

**Etiology**  
A dysfunctional disorder of childhood that probably results from epileptogenic "functional damage" to the speech cortex during a critical period of child development. Symptomatic cases are rare.

**Clinical manifestations**  
Speech disorders (100%) and seizures (75%). It begins with verbal auditory agnosia (inability to respond to speech and later non-speech sounds). The course is subacute or intermittent. Symptoms of all types of aphasia may occur. Finally, the child becomes completely "numb" - mutism, unable to respond even to non-verbal sounds. The disease often occurs with remissions and exacerbations. Cognitive and behavioral problems develop in more than 3/4 of cases. Also, 3/4 of patients occasionally experience epileptic seizures - generalized tonic-clonic (GTC), focal motor, atypical absences and atonic seizures, as well as epileptic status. Seizures most often occur at night.

**Diagnostic procedures**  
MRI often reveals no changes; when using functional neuroimaging methods, disturbances in the temporal lobe of the dominant hemisphere are noted.

**Interictal EEG**  
Sharp-slow wave discharges in the posterior temporal areas, often multifocal and bisynchronous. At a certain stage of the disease, ESMS almost always develops - electrical status of slow sleep with almost continuous spike-wave activity in the slow sleep phase. However, it should be noted that recording the electrical status of slow sleep is not a prerequisite for making a diagnosis.

**Ictal EEG**  
Depends on the type of attacks

**Forecast**  
Usually unfavorable. Remission of seizures and EEG abnormalities occurs on average at the age of 15, speech and neuropsychological disorders gradually disappear. However, only 10%-20% of patients achieve complete normalization. The rest have a permanent and quite serious speech and cognitive deficit. The prognosis does not depend on the frequency and type of seizures.

**Differential diagnostics**  
Acquired deafness or selective mutism; epilepsy with continuous spike-waves during slow-wave sleep ([CSWS](https://epidoc.ru/syndromes/encephalopathies/csws.html)),[benign](https://epidoc.ru/syndromes/bfec/index.html)or[severe focal](https://epidoc.ru/syndromes/severe_neocortical/index.html)childhood epilepsy

**Treatment**  
Seizures are fairly easily controlled with AEDs. The goal is to reduce the number of epileptiform discharges in the EEG. Valproate, ethosuximide, clobazam, and sulthiame are used. Lamotrigine, levetiracetam, topiramate, and zonisamide may also be tried. ACTH or prednisone is often the drug of choice, especially in younger patients with newly diagnosed disease. There is empirical evidence that results depend on early treatment with a high initial dose of steroids for at least 3 months. Continuation of treatment after this period depends on the observed effect and side effects of the drugs. Steroids are usually used with valproate or benzodiazepine, which can be continued after steroids are discontinued. Quite a good effect is achieved with a surgical method - subpial intracortical transections.

# Lennox-Gastaut syndrome

**Prevalence**  
~ 5% to 10% of children with seizures.

**Morbidity**  
~ 2.8 per 10,000 births.

**Age of disease onset**  
~ from 1 to 7 years; peak at 3-5 years.

**Floor**  
Boys slightly predominate (60%).

**Neurological and mental status**  
Motor, cognitive and behavioral impairments are observed in approximately 60% even before the onset of attacks.

**Etiology**  
Extensive and varied. These are often severe brain lesions and malformations of cortical development (70%). 1/3 of cases are idiopathic or probably symptomatic.

**Clinical manifestations**  
Clusters of seizures of various types, mental retardation. The following types of seizures predominate - tonic, atonic, atypical absences. Tonic seizures tend to occur at night. Falls are also noted, often traumatic. Half of the patients develop nonconvulsive epileptic status of atypical absences, tonic or atonic seizures, myoclonus.

**Provoking factors**Sleep, inactive state

**Diagnostic procedures**  
Neuroimaging data may reveal brain lesions, malformations of cortical development. Biochemical, hematological and other laboratory tests are usually within normal limits.

**Interictal EEG**  
Slowing of background activity, paroxysms of fast rhythms and spike-wave complexes with a frequency of <2.5 Hz.

**Ictal EEG**  
Depends on the seizure type. Tonic seizures are paroxysms of fast activity (~20 Hz); atypical absences are slow spike-wave complexes with a frequency of <2.5 Hz; myoclonic seizures are polyspikes; atonic seizures are spikes/polyspikes/slow spike-waves and paroxysms of fast activity. A pattern of flattening of EEG activity is often recorded, sometimes in combination with fast activity.

**Forecast**  
Poor, fatal outcome in 5%, 80%-90% continue to have attacks into old age. Almost all (85%-92%) have severe neurological and mental deficits. Normal development is the exception.

**Differential diagnostics**  
Non-epileptic events, other[epileptic encephalopathies](https://epidoc.ru/syndromes/severe_neocortical/index.html), epilepsy with[myoclonic-astatic seizures](https://epidoc.ru/syndromes/ige/doose_syndrome.html).

**Treatment**  
Complete seizure control is unlikely. Valproate (for all types of seizures), clonazepam (myoclonic), and phenytoin (tonic). Lamotrigine, levetiracetam, and topiramate may be more effective than older drugs. Felbamate may be helpful in some cases. Steroids are sometimes helpful. New data on the effectiveness of[ketogenic diet](https://epidoc.ru/diagnose_treat/ketogenic.html). There is no data on effectiveness yet.[vagus nerve stimulation](https://epidoc.ru/diagnose_treat/vns.html). In severe cases, the possibility can be considered.[surgical treatment](https://epidoc.ru/diagnose_treat/surgery/index.html)– resective surgery of localized structural brain lesions, callosotomy to reduce generalization and, accordingly, drop attacks. In case of epileptic status, intravenous benzodiazepines are used, sometimes with steroids, assisted breathing. Intravenous diazepam and lorazepam can cause tonic seizures. In the treatment of patients with Lennox-Gastaut syndrome, a multidisciplinary approach and family support are necessary.

# Myoclonic status in non-progressive encephalopathies

**Prevalence**  
Unknown, among children with severe forms of epilepsy it is approximately 0.5%-1%.

**Age of disease onset**  
From the first day of life until 5 years, peak at 12 months.

**Floor**  
Girls suffer twice as often.

**Neurological and mental status**  
Violations (76%).

**Etiology**  
(1) Chromosomal abnormalities, mainly Angelman syndrome (49%); (2) fetal or neonatal brain hypoxia (20%); and (3) malformations of cortical development and other lesions (31%).

**Clinical manifestations**  
Atypical status epilepticus is a type of myoclonic seizures and recurrent absences that last for a long time (sometimes for several days). Myoclonus affects the eyelids, face, and limbs, is chaotic, asynchronous, and becomes more rhythmic and synchronous during absences. Myoclonus is often subtle, and may give the impression of apathy or ataxia in an infant. Myoclonic status may be the first manifestation of seizures; in other cases, it begins with focal motor seizures, myoclonic absences, massive myoclonus, or, less commonly, generalized or unilateral clonic seizures, which in some cases occur only against the background of high fever. Tonic seizures are not observed. Many patients experience frequent and unexpected spontaneous massive startle attacks in the form of a sudden, short-term drop in postural tone, as well as prolonged bursts of intention myoclonus or tremor.

**Diagnostic procedures**  
Chromosomal analysis, neuroimaging and EEG.

**Interictal EEG**  
Disturbances in background rhythms in the form of focal or diffuse slowing, asymmetrical (more in the frontal-central areas) paroxysms of oscillations in the range of 3-6 Hz, sometimes with a spike component.

**Ictal EEG**  
It varies, and is mainly represented by paroxysms of slow waves with a frequency of 3 to 6 Hz with superimposed spikes, or paroxysms of slow spike-waves with a frequency of <2 Hz.

**Forecast**  
Unfavorable, with progressive deterioration, formation of severe neurological and mental deficit. Myoclonic status weakens with age.

**Differential diagnostics**  
Progressive encephalopathies - such as late infantile ceroid lipofuscinosis,[migratory focal seizures in infants](https://epidoc.ru/syndromes/severe_neocortical/migrating_focal_seizures.html),[Dravet syndrome](https://epidoc.ru/syndromes/encephalopathies/dravet_syndrome.html).

**Treatment**  
There is currently no effective treatment. Benzodiazepines may temporarily interrupt status epilepticus myoclonus.

# Ohtahara syndrome

**Prevalence**  
Unknown. Approximately 100 cases have been described, but this figure is undoubtedly an underestimate, and many cases of this severe and fatal infant disease are simply not diagnosed.

**Age of disease onset**  
Mostly in the first 10 days of life, sometimes in utero or up to 3 months of age. Epileptic spasms generally occur in 1.5-5 per 1000 newborns in the postnatal period.

**Floor**  
Boys get sick slightly more often.

**Neurological and mental status**  
Violations.

**Etiology**  
The most common cause is malformations of the brain - hemimegalencephaly, porencephaly, Aicardi syndrome, olivary-dentate dysplasia, agenesis of the mammillary bodies, linear sebaceous nevus syndrome, cerebral dysgenesis, focal cortical dysplasia. Less often, the cause of the disease is other structural and metabolic disorders.

**Clinical manifestations**  
Mainly tonic spasms, which are usually represented by a tonic flexion lasting from 1 to 10 seconds, can be single or repeated in clusters, from 10 to 300 times during the day. They can be generalized and symmetrical, or lateralized. They occur both in sleep and in wakefulness. Somewhat less often, in about a third of infants, chaotic focal motor clonic seizures or hemiconvulsions are observed. Alternating hemiconvulsions or generalized tonic-clonic seizures (GTCS) are extremely rare. Myoclonic seizures, including chaotic myoclonus, are also rare.

**Diagnostic procedures**  
As with neonatal seizures, neuroimaging is necessary to determine the etiology and possible treatment. This usually reveals significant disturbances and malformations of cortical development. If neuroimaging data are normal, metabolic screening is mandatory.

**Interictal EEG**  
Pseudo-rhythmic repetitive burst-suppression pattern, physiological rhythms are absent. With age, the EEG shows evolution to the hypsarrhythmia pattern of West syndrome, then to the slow spike-wave pattern of Lennox-Gastaut syndrome.

**Ictal EEG**  
The burst-suppression pattern, namely burst, is associated with tonic spasms of varying duration. Tonic spasms may also be associated with the following EEG changes: - diffuse desynchronization with the disappearance of burst-suppression activity, when tonic spasms are grouped into clusters with an interval between spasms of 5-10 seconds - the burst-suppression pattern becomes more frequent and diffuse, higher in amplitude than in the interictal period

**Forecast**  
It is a devastating syndrome, with high mortality and complications. Half of the patients die within weeks or months of the onset of the disease, while the rest develop permanent neurological and mental deficits.

**Differential diagnostics**  
The main differential diagnosis is with the syndrome[early myoclonic encephalopathy](https://epidoc.ru/syndromes/neonatal/early_myoclonic_encephalopathy.html).

**Treatment**  
There is currently no effective drug treatment. In some cases, if the etiology is focal cortical dysplasia, surgical treatment may be considered.

# West syndrome (infantile spasms)

**Prevalence**  
The most common type of epileptic encephalopathy.

**Morbidity**  
From 3 to 5 per 10,000 births.

**Age of disease onset**  
From 3 to 7 months (77%), less often up to 3 months or at the age of 1-5 years.

**Floor**  
Boys (60%) get sick slightly more often.

**Neurological and mental status**  
Developmental delay, mild or severe, in 2/3 the delay is noted even before the onset of attacks.

**Etiology**  
Severe lesions predominate. Tuberous sclerosis is a common cause.

**Clinical manifestations**  
Epileptic (infantile) spasms are a characteristic type of seizure. The seizure may be accompanied by crying. Spasms occur in clusters up to 30 times a day, with 20-100 spasms in each cluster. Seizures occur more often during wakefulness than during sleep.

**Diagnostic procedures**  
A clinical assessment of the neurodevelopmental status is performed. Laboratory screening for electrolyte, metabolic, or other abnormalities is usually normal. In unclear cases, cerebrospinal fluid testing, neurometabolic testing, and chromosomal analysis can be performed to identify the etiology. Computed tomography (CT) and especially magnetic resonance imaging (MRI) are mandatory before initiating steroid therapy.

**Interictal EEG**  
The classic epileptiform pattern of hypsarrhythmia is recorded in 2/3 of patients. Asymmetric and modified hypsarrhythmia occur in 1/3 of cases.

**Ictal EEG**  
Up to 11 different ictal patterns may be recorded, lasting from 0.5 sec to 2 min. The most common pattern (72%) consists of a high-amplitude generalized slow wave, an episode of low-amplitude fast activity, and diffuse flattening of the EEG.

**Forecast**  
In 5%, the outcome is fatal, 65% develop epilepsy with resistant seizures, in half of cases persistent motor deficit remains, in 2/3 severe cognitive/psychological disorders occur. Normal development is observed only in 5%-12% of patients.

**Differential diagnostics**  
Nonepileptic events: normal startle reactions, "colic, abdominal pain", benign nonepileptic infantile spasms, benign neonatal sleep myoclonus, Sandifer syndrome or gastroesophageal reflux, torticollis, abnormal dystonic posture of the trunk, opisthotonus. Benign or other severe forms of epilepsy in this age group.

**Treatment**  
Vigabatrin or adrenocorticotropic hormone (ACTH) begin to control spasms in 2/3 of patients within a few days. The final result is not always determined by treatment. In some cases with intractable seizures and focal structural lesions, resective neurosurgery may be used.