Epileptic Syndromes in Childhood. A Practical Approach

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Abstract

Epileptic seizures are among the most frequent severe childhood neurological diseases and are defined as a transient, paroxysmal and involuntary event manifested by motor, sensory, autonomic and psychic signs, with or without alteration of consciousness, caused by synchronous and excessive neuronal activity in brain. Epilepsy is a brain disease characterized by (a) at least two unprovoked epileptic seizures or two reflex seizure at a minimum interval of 24 hours; or (b) an epileptic seizure or reflex seizure and risk of a new event estimated at least 60%; or (c) diagnosis of an epileptic syndrome. This article aims to review the main diagnoses and therapeutics of the most frequent epilepsy syndromes of the childhood and adolescence.

Keywords: Epilepsy, Diagnosis, Therapeutics, Treatment Outcome.
**INTRODUCTION**

Seizures have been reported in ancient societies for over 5,000 years. Before the Christian Era, seizures used to be attributed to demonic possession, divine punishment, or the influence of celestial objects, as reported in the Biblical Gospels of Matthew, Mark, and Luke. Hippocrates, in the chapter *On the Sacred Disease* of his *Corpus Hippocraticum*, challenges the belief that epilepsy was caused by magical forces, being a pioneer in relating seizures to changes in cerebral functioning.

Throughout ancient history, different approaches have been used to treat epilepsy, including prayers, exorcisms, physical punishment, herbal infusions, bloodletting, and even trepanation of the skull. The first ever known “scientific” treatment was supposedly proposed in 1857 by Charles Locock, who, imagining that seizures were related to “excess female sexuality,” developed a treatment approach involving a powerful antiaphrodisiac, potassium bromide.

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The present article aims to perform an objective update on the diagnosis and treatment of the main epilepsy syndromes in childhood and adolescence.

**DEFINITIONS AND EPIDEMIOLOGY**

There is no unique and universally accepted definition for “epilepsy,” a concept that has been changing in recent years with the constant evolution of neuroscience. Epileptic seizures can be defined as transient, paroxysmal, and involuntary events manifested by motor, sensory, autonomic, and psychic signs and symptoms, with or without alterations in consciousness, as a result of synchronous and excessive neuronal activity in brain tissue.

Epileptic seizures can be classified as “provoked or acute symptomatic” when temporally related to a condition that has transiently changed brain functioning, such as hemorrhage, ischemia, infection, metabolic change, or acute drug withdrawal. Epileptic seizures classified as “provoked” should not, a priori, be considered epilepsy because the triggering factor is transient in nature. On the other hand, “unprovoked” epileptic seizures, those that occur in the absence of an immediate precipitating factor, justify the use of the concept of “epilepsy.”

Seizures initially classified as “provoked” can arise from brain damage that will make the individual prone to present “unprovoked” seizures in the future.

Since 2014, an operational definition for epilepsy has been used due to its practicality: “Epilepsy is a brain disease characterized by (a) at least two unprovoked or reflex seizures occurring at least 24 hours apart; (b) one unprovoked or reflex seizure with an estimated recurrence risk of at least 60%; or (c) diagnosis of an epilepsy syndrome.”

The criteria of 60% recurrence risk and initiating treatment after the second seizure have been adopted according to an important study published nearly 20 years ago, which showed that, after the first unprovoked seizure, the maximum recurrence risk reached 40% in 5 years; after the second seizure, this risk reached 87%; and after the third seizure, the recurrence risk became invariable.

As for treatment time, epilepsy is considered “resolved” either when crises are related to a particular age range that has passed or in all patients with no recurrence for over 10 years and no continuous and regular use of AEDs for at least 5 years.

There are no reliable data on the incidence and prevalence of epilepsy in Brazil. In addition, the continental dimensions of our country cause significant epidemiological variation between regions. Between 2% and 3% of people will be diagnosed with epilepsy at some point in life, and the estimated prevalence of epilepsy in Brazil varies between 10 and 15 in 1,000 individuals.

**Benign familial neonatal seizures**

Described by Rett and Teubel in 1964, benign familial neonatal seizures constitute a rare, autosomal dominant syndrome caused by a mutation in genes encoding the subunits of voltage-gated potassium channels and are classified among the idiopathic generalized epilepsies and epilepsy syndromes. Seizures begin between the second and third days of life, being classified based on their signs and symptoms into clonic, focal, or generalized tonic; apnea; and seizures with autonomic phenomena. Virtually, all children have normal neurological examinations, although a slight motor delay can be identified in the first and second years of life.

The electroencephalogram (EEG) shows nonspecific findings and may be normal or present epileptogenic paroxysms with a generalized spike and spike-and-wave morphology. Magnetic resonance imaging (MRI) of the brain (head MRI) is a mandatory part of the neurological clinical investigation and shows no significant changes. Drug treatment with phenobarbital, sodium valproate, or divalproex sodium should be administered for a short period of time and discontinued after approximately 6 months.

**Nonfamilial neonatal seizures**

Also called benign idiopathic neonatal seizures or “fifth-day fits,” nonfamilial neonatal seizures were described by Dehan in 1977 and are considered a rare epilepsy syndrome among the idiopathic generalized epilepsies and epilepsy syndromes. The first seizure occurs between the fourth and sixth day of life, being classified into focal clonic seizures or apnea, with or without autonomic phenomena. There are cases of frequent recurrence and progression to neonatal status epilepticus (SE). Most patients have a normal neurological examination, although some may progress to a slight hypotonia in the first years of life.
The EEG may be normal, but around 60% of patients present a sharp, discontinuous, alternating theta pattern (theta pointu alternant), sometimes synchronous, other times asynchronous and monomorphic. Paroxysms of sharp morphology can be seen in different regions. Head MRI should be performed to rule out the possibility of structural damage, but it shows no significant changes1,5.

Drug treatment with phenobarbital, sodium valproate, or divalproex sodium should be administered; polytherapy may be necessary in difficult-to-control cases1,5.

Early or neonatal myoclonic encephalopathy

Also called Aicardi syndrome, neonatal myoclonic encephalopathy was described in 1978 by Aicardi and Goutières and is classified among the symptomatic generalized epilepsies and epilepsy syndromes. Considered a rare epilepsy syndrome, its incidence is unknown; it affects both sexes and all ethnicities1,6. It is clinically defined by epileptic seizures of early onset (first hours or days of life), which are classified based on their signs and symptoms into fragmentary (focal) or massive myoclonic (involving limbs and axial muscles). Focal clonic, simple partial, and tonic seizures as well as epileptic spasms and seizures with autonomic phenomena have also been described. Seizures tend to be frequent and may progress to SE. Neurological examination results are abnormal, with consistent developmental delay. The etiology is varied, with consistent presence of severe brain injury, combined or not with non-ketotic hyperglycinemia or other inborn errors of metabolism1,6.

The EEG shows disorganized and slow baseline activity and a classic burst/suppression pattern, with bursts consisting of polymorphic discharges of spikes, polyspikes, and sharp waves mixed with generalized slow waves. The cerebrospinal fluid should be collected for glycine determination1,6.

Its prognosis is poor, neurological morbidity is high, and mortality is estimated at 50% at the end of the first year of life. The treatment outcomes are discouraging, with usual progression to difficult-to-control multifocal epilepsy. The AEDs used are phenobarbital, sodium valproate, topiramate, and benzodiazepines. In specific cases, ACTH can reduce seizure frequency and intensity1,6.

West syndrome

Described in 1841 by W. J. West in a letter to The Lancet about his own son, West syndrome is the most frequent epileptic encephalopathy in the first year of life, characterized by this classic triad: (a) epileptic spasms, (b) developmental delay, and (c) hypsarrhythmia in the EEG1,6. It is predominant in boys, with an incidence ranging between 1.4% and 2.5% of all childhood epilepsies and between 2 and 3.5 in every 10,000 live births9.

The etiology is varied and can be determined in up to 75% of cases, with the main causes being brain injuries resulting from perinatal asphyxia, brain malformations, disorders of cortical development, and tuberous sclerosis complex. The prognosis is poor, with severe developmental delay and cognitive deterioration in 95% of children. One of the main physiopathogenic hypotheses relates spasms in West syndrome to an increased release of the neuropeptide corticotropin in the limbic system and regions of the encephalic trunk1,10.

The EEG shows the classic interictal hypsarrhythmia, characterized by slow and disorganized baseline activity, with polymorphic discharges of sharp waves, spikes, polyspikes, spike-and-waves, and polyspike-and-waves mixed with slow waves of high amplitude (over 200 microvolts), with a distinguishable lack of phase coherence in bursts and activation during the initial stages of non-rapid eye movement (NREM) sleep9,10.

Five variations of hypsarrhythmia are described: (a) hypsarrhythmia with increased interhemispheric synchronization, (b) asymmetrical hypsarrhythmia (c) hypsarrhythmia with consistent focal abnormalities, (d) hypsarrhythmia with episodes of generalized voltage attenuation, and (e) hypsarrhythmia with bilateral, high-
voltage, asynchronous slow activity. Brain MRI helps in defining the etiology of this syndrome.

The treatment of this syndrome has perhaps been one of the most discussed topics in pediatric neurology over many years. Since 1958, when Sorel and Dusaucy described satisfactory results with the use of ACTH, this hormone has been used in the treatment of West syndrome. Side effects such as increased blood pressure, osteoporosis, transient immunosuppression, and adrenocortical response changes may limit its use in some children. Several studies report the effectiveness of oral steroids, such as prednisone, considered an alternative to ACTH. The risk of seizure recurrence after discontinuation of oral corticosteroids is significantly higher than that after discontinuation of ACTH.

Vigabatrin is considered a first-line AED, particularly in cases secondary to tuberous sclerosis complex. Although the risk of concentric loss of vision limits its indication in some cases, vigabatrin is well tolerated by virtually all patients. Valproic acid and benzodiazepines can be prescribed in specific cases. For patients refractory to usual medications, topiramate, divalproex sodium, benzodiazepines, lamotrigine, topiramate, and rufinamide are indicated, usually requiring polytherapy. Levetiracetam can be used in combination with other AEDs. Palliative surgical procedures such as disconnection procedures, corpus callosotomy, and use of vagus nerve stimulation therapy can be useful in refractory patients. Focal cortical resections are indicated in specific and carefully selected cases. Protective helmets can reduce the risk of trauma to the skull.

Lennox–Gastaut syndrome

Described by Gastaut and colleagues in 1966, Lennox–Gastaut syndrome (LGS) is classified among the symptomatic generalized epilepsies and epilepsy syndromes, being more frequent in boys. The etiology is either structural or metabolic in most patients, although cryptogenic cases of lower neurological morbidity have been reported.

LGS is clinically defined by a triad of atonic, tonic, and atypical absence seizures. In later stages, the presence of complex partial, simple partial, and generalized tonic-clonic seizures (GTCS) is common. Seizures begin before 8 years of age, with a peak between 3 and 4 years of age. Two-thirds of patients will present nonconvulsive atypical absence SE at some point, and half of patients will present “drop attacks” with massive myoclonus, followed by generalized tonic seizures. Severe developmental, cognitive, and intellectual impairment is almost always present, as well as an association with behavioral disorders, hyperactivity, psychomotor agitation, and impulsivity. Comorbidity with autism spectrum disorder is common and the differential diagnosis must be established from West, Dravet, and Doose syndromes, in addition to atypical benign partial epilepsy (pseudo-Lennox syndrome).

The EEG features classic slow (often less than 3 Hz) generalized spike-and-wave discharges in a moderate to markedly disorganized and slow baseline activity. Generalized epileptic “recruiting” rhythm (burst of fast activity with a 10 to 20 Hz frequency and medium amplitude) is frequent during sleep. Due to the high intensity of discharges and the intense EEG disorganization, distinguishing interictal from ictal activity may be impossible.

Head MRI complements the etiological research, with radiological findings being quite varied, including structural lesions resulting from perinatal hypoxia, brain malformations, dysplasia, and neuronal migration disorders. High-resolution head MRI enables the identification of minor dysplasia in patients with LGS previously classified as cryptogenic, whereas positron emission tomography scan can identify hypometabolic areas in patients with normal head MRI.

Treatment generally fails to provide satisfactory control of seizures, and progression to tonic or atypical absence SE is common. Carbamazepine and phenytoin may exacerbate myoclonic and atypical absence seizures, and myoclonic SE induced by these AEDs has been reported. Sodium valproate, divalproex sodium, benzodiazepines, lamotrigine, topiramate, and rufinamide are indicated, usually requiring polytherapy. Levetiracetam can be used in combination with other AEDs. Palliative surgical procedures such as disconnection procedures, corpus callosotomy, and use of vagus nerve stimulation therapy can be useful in refractory patients. Focal cortical resections are indicated in specific and carefully selected cases. Protective helmets can reduce the risk of trauma to the skull.

Landau–Kleffner syndrome

Also called acquired epileptic aphasia, Landau–Kleffner syndrome was described by Landau and Kleffner in 1957 and is considered a rare epilepsy syndrome. It is diagnosed between ages 3 and 7 years, with a male predominance.

The etiology is not completely known yet, but brain injuries can possibly be found, including malformations of cortical development and injuries resulting from trauma or central nervous system infections. There seems to be genetic involvement in a smaller percentage of the cases.

Epileptic seizures are not mandatory criteria for the diagnosis of Landau–Kleffner syndrome, although they are present in up to 80% of cases. In half of patients, seizures predate aphasia. Seizures are activated by NREM sleep, with a predominance of partial seizures, GTCS, atypical absence seizures and, more rarely, myoclonic seizures. Progression is heterogeneous, and the epilepsy becomes either easy to control or refractory. Receptive aphasia is present in all patients, and its cause is not fully known yet. Functional changes resulting from epileptogenic discharges in the neural circuitry responsible for speech are identified as the direct cause of verbal behavior disorders. Intense epileptic activity in the left temporal lobe appears to contribute to the establishment of hypometabolic encephalopathy manifested.
by changes in verbal and social behavior. However, even after the temporal discharges disappear, most children remain aphasic. After a short time, receptive aphasia is followed by expressive aphasia and mutism. Behavioral disorders and autism spectrum disorder affect almost 80% of these patients, usually lasting their entire lives.1,19,20-22

The EEG shows sharp wave discharges in temporal regions, with an occasional left predominance. Extratemporal discharges may also be present, and a significant proportion of these patients present continuous spike-and-wave during sleep, which aggravates neurological morbidity. Head MRI may be normal or may present nonspecific findings, such as brain atrophy, or specific structural changes, such as malformations of cortical development and destructive lesions.1,20-22

Drug treatment, despite generally leading to the control of epileptic seizures, appears incapable of changing verbal and social behavior. ACTH and oral steroids are effective in some patients. Sodium valproate, divalproex sodium, topiramate, and benzodiazepines can assist in antiepileptic maintenance therapy. Ethosuximide is indicated in cases of refractory absence seizures. Levetiracetam can be used in combination. Phenobarbital and phenytoin are contraindicated due to reports of seizure aggravation and SE. The involvement of the eloquent cortex prevents surgical procedures aimed at cortical resection. Palliative surgeries such as Frank Morrell’s multiple subpial transection can be indicated in selected cases.1,19-21

Dravet syndrome

Also called severe myoclonic epilepsy in infancy or severe myoclonic epilepsy of infants, Dravet syndrome was described by Charlotte Dravet in 1978 and is a rare epilepsy syndrome, with a slight male predominance and genetic etiology. Seizures begin in the first year of life and are classified as clonic, involving half the body, and possibly progressing to GTCS, with or without fever. Generalized or fragmentary myoclonic seizures, tonic seizures, GTCS, and atypical absence seizures appear later, usually with fever as a trigger. The epilepsy has a refractory behavior and is accompanied by developmental stagnation and regression, behavioral disorders, hyperactivity, impulsivity, and autism spectrum disorder.1,23

The EEG changes progressively and may be normal in the early stages. Baseline activity progresses to become disorganized and slow; in half the cases, synchronous theta rhythm with a frequency of 4–5 Hz emerges in the central and parietal regions.

Discharges of spike, spike-and-wave, polyspike, and polyspike-and-wave appear sometimes in a generalized or multifocal pattern. Head MRI is normal in most patients; nonspecific changes such as brain atrophy may be present. Mesial temporal sclerosis is present in 2%-70% of patients. Approximately 75% of patients have mutations in the SCN1A gene, which encodes the alpha-1 subunit of the sodium channels.1,23,24

Treatment is discouraging and rarely leads to satisfactory and lasting seizure control. Because it is an epilepsy syndrome with polymorphic seizures, the choice of AED should be aimed at the predominant types of seizure. Phenobarbital, sodium valproate, divalproex sodium, benzodiazepines, topiramate, ethosuximide, and zonisamide can be used. Carbamazepine, phenytoin, and lamotrigine should be avoided due to reports of seizure aggravation. The combination of sodium valproate, stiripentol, and clobazam appears to be effective for a significant part of these patients. Refractory cases can benefit from ketogenic diet.1,25

Doose syndrome

Also called myoclonic-astatic epilepsy, Doose syndrome was described by Herman Doose in 1970 and is classified among the generalized cryptogenic or symptomatic epilepsy syndromes, with seizure polymorphism being frequent. Its estimated incidence is 1 case for every 10,000 live births, corresponding to 1%-2% of all childhood-onset epilepsies. It is more common in boys, except in cases of seizures with onset before the age of 1 year, for which incidence is equal in both sexes.1,26

Although the diagnosis is based on the presence of myoclonic seizures, GTCS, atonic, tonic, and atypical absence seizures have also been described. For 94% of patients, seizures begin in the first 5 years of life. Children initially present normal psychomotor development, later progressing to cognitive and behavioral deterioration. The differential diagnosis from LGS can be made by noting that in Doose syndrome, children are neurologically healthy before the onset of seizures and myoclonic seizures are markedly predominant.1,26

The EEG is initially normal, but baseline activity soon becomes slow and disorganized, and generalized polymorphic discharges with focal sharp waves appear. Bursts of slow spike-and-wave discharges with a 2 to 2.5 Hz frequency can appear in the interictal period. Some patients show “Doose rhythm,” consisting of theta, monomorphic activity of medium amplitude and bilateral projection to the parietal regions.1,26-28

Treatment should include broad-spectrum AEDs such as sodium valproate, divalproex sodium, topiramate, lamotrigine, and benzodiazepines. Good results have been reported with the use of ethosuximide, levetiracetam, primidone, ACTH, oral steroids, and acetazolamide. Ketogenic diet is reserved for refractory cases. The prognosis is quite variable, from cases with complete remission of symptoms and seizure control to cases progressing to refractory seizures and developmental impairment. Episodes of SE with tonic-vibratory seizures and myoclonic SE suggest a worse prognosis.27,28

Benign Rolandic epilepsy

Benign childhood epilepsy with centrotemporal spikes, also called benign Rolandic epilepsy, has a slight predominance in males and a strong genetic predisposition. Its annual incidence is estimated at 45:100,000 people, being one of...
the most frequent childhood epilepsies. This is an age-related epilepsy, with seizures beginning at the age of 7-10 years, stereotyped, and characterized according to its signs and symptoms by sensory changes (paresthesia) in the tongue and lips, followed by intense salivation, speech impairment, facial clonuses, lateral deviation of the eyes and head, and increased muscle tone in the upper and lower limbs ipsilateral to the eye deviation. GTCS may occur. Seizures predominate during sleep; can start with vocal emission, grunting, or screaming; and last less than 3 minutes.

The EEG shows normal baseline activity and sharp wave discharges projected in the central, temporal, and parietal regions, sometimes to the right or the left. When discharges during sleep are frequent, “pseudo”-focal slowing can occur. Head MRI is always normal.

Treatment usually leads to satisfactory seizure control, and AEDs can be discontinued in adolescence. The first-choice drugs are carbamazepine and oxcarbazepine, although other AEDs, such as levetiracetam, topiramate, and lamotrigine, can have a similar efficacy. Benzodiazepines are a treatment option in patients with frequent nocturnal recurrences.

Benign occipital epilepsy, Panayiotopoulos type
Also called early-onset benign childhood occipital epilepsy, benign occipital epilepsy is an age-related syndrome affecting both sexes, presenting a strong genetic component, and being the second most common childhood benign partial epilepsy. Seizures rarely occur, and many patients have only one event during their entire life. Neurological examination results are normal, and neuropsychomotor development is not impaired.

Seizures begin at around 5 years of age and are characterized by behavioral changes, irritability, aggressiveness, followed by autonomic phenomena, pallor, lip cyanosis, nausea, vomiting, and head and eye deviation. Visual changes typical of the occipital lobe may be present as well as ictal amaurosis. Seizures may be short or progress to partial SE.

The EEG shows normal baseline activity and the presence of sharp wave discharges in the occipital region, although discharges can also be observed in other regions. One-third of these children present normal EEG. Head MRI should be performed to rule out structural lesions in the posterior cortex, but it should be normal in this syndrome.

Because this syndrome has a low risk of seizure recurrence, the use of AEDs should be considered with caution. When continuous treatment is chosen, carbamazepine or oxcarbazepine should be prescribed, although phenobarbital, sodium valproate, and topiramate possibly have a similar effectiveness.

Occipital epilepsy, Gastaut-type
Also called late-onset childhood occipital epilepsy, Gastaut-type occipital epilepsy is considered a rare, age-related syndrome that affects both sexes and has a strong genetic component. Seizures are characterized by elementary visual hallucinations, with colored or bright dots, colorful geometric shapes, black circles, or dots that move anarchically. Ictal amaurosis, eye deviation, forced palpebral closure, and blinking are less frequent. Progression to GTCS is relatively common, as well as intense holocranial headache.

The EEG shows normal baseline activity and sharp wave discharges in the posterior cortex, predominantly in the occipital region, sometimes to the right or the left. Fast beta rhythms are described in the posterior cortex. Head MRI should be performed to rule out structural damage and should be normal.

The recommended AEDs are carbamazepine or oxcarbazepine, although topiramate, sodium valproate, and levetiracetam present a similar efficacy. The prognosis is usually good, with complete remission of the crises after 2-4 years.

Childhood absence epilepsy
Childhood absence epilepsy (CAE) is classified among the idiopathic generalized epilepsies, being the most frequent childhood genetic generalized epilepsy. Although it affects both sexes, it is more common in girls, has a strong genetic component, and affects neurologically healthy children. Absence seizures start at around 6-8 years of age and are characterized by sudden and complete consciousness impairment, lasting a few seconds. They end with complete and sudden recovery of consciousness. Seven different types of typical absence seizures are recognized: (a) absence seizure with consciousness impairment, (b) absence seizure with a slight clonic component, (c) absence seizure with an atomic component, (d) absence seizure with a tonic component, (e) absence seizure with automatisms, (f) absence seizure with autonomic phenomena, and (g) mixed forms (a combination of aforementioned ictal manifestations). It is important to note that children diagnosed with CAE may progress to myoclonic seizures and GTCS.

The EEG shows a typical pattern of generalized spike-and-wave discharges with a frequency of 3-4 Hz. Baseline activity should be normal, and occipital intermittent rhythmic delta activity may occur, which is considered an EEG marker of a better prognosis. Head MRI does not show any abnormalities.

AEDs usually lead to complete absence control, with sodium valproate, divalproex sodium, ethosuximide, and lamotrigine being recommended. In refractory cases, combination with benzodiazepines may be necessary.

Juvenile myoclonic epilepsy
Described in 1957 by Janz and Christian, juvenile myoclonic epilepsy is classified among the idiopathic generalized age-dependent epilepsies and epilepsy syndromes, being considered the most frequent generalized genetic epilepsy syndrome in adolescence, with an annual incidence of...
0.5-6.3 per 100,000 individuals. It affects neurologically healthy individuals of both sexes, with a female predominance\(^1,38,39\).

Seizures start at around 14-16 years of age, being characterized by bilateral, symmetrical myoclonus, either in isolation or in bursts, predominantly affecting the upper limbs. Myoclonic seizures are most common upon awakening and in early morning; they are activated by sleep deprivation, alcohol intake, physical fatigue, and emotional disorders. Myoclonic seizures may be accompanied by GTCS (90% of cases) and absence seizures (15% of cases). As myoclonus is very brief, the level of consciousness remains unaltered, but changes can be triggered by light stimulation (photosensitive epilepsy)\(^1,38-40\).

The EEG shows normal baseline activity and generalized discharges of spikes, spike-and-waves, polyspikes, and polyspike-and-waves most acute in the anterior cortex. Focal or multifocal spikes during sleep are a usual finding in idiopathic generalized epilepsies. Photic stimulation is mandatory during the EEG because almost half of patients are photosensitive\(^38-40\).

Detailed guidance on triggering factors is considered an essential part of treatment. The main AEDs are sodium valproate, divalproex sodium, and lamotrigine. Clonazepam is effective but should be considered a supplemental drug. Topiramate and zonisamide can be used after failure of first-line drugs. Carbamazepine, oxcarbazepine, phenytoin, vigabatrin, and gabapentin may cause myoclonic and absence SE\(^1,39,40\).

**Rasmussen’s encephalitis**

Rasmussen’s encephalitis (RE) was described in 1958 by Rasmussen, Olszewski, and Lloyd-Smith. It is characterized by refractory partial seizures, hemiparesis, and progressive intellectual impairment\(^4\). Seizures begin during childhood and are classified based on their signs and symptoms into complex partial or simple partial (clonic) seizures, with possible progression to GTCS. Focal SE can affect up to 20% of these patients. The etiology and pathophysiology of RE is only partially known, with the participation of factors related to the immune response\(^4\).

EEG findings vary during the disease progression and may be normal in its initial stages. Baseline activity becomes progressively asymmetric, with the affected cerebral hemisphere having a slow, disorganized, depressed baseline activity. The interictal record is marked by the presence of focal or multifocal discharge of sharp waves with a clear hemispheric predominance. Head MRI shows hemispheric cerebral atrophy, most prominent in the temporal and insular regions. Magnetic resonance spectroscopy can demonstrate a relative reduction in the neuronal marker N-acetyl aspartate in the affected hemisphere\(^1,42\).

Treatment is discouraging and usually fails to stop the severe progression of this disease. Although plasmapheresis and high doses of steroids, immunoglobulins, interferon, and ACTH can be prescribed, cognitive, mental, intellectual, and motor deterioration is almost always present. Both traditional and more recent AEDs are ineffective as well. Surgical treatment should be indicated at an early stage, aiming to preserve the child’s neurological state, with hemispherectomy being the most common procedure. Frank Morrell’s multiple subpial transection is an alternative surgical approach when the eloquent cortex is involved\(^1,42\).

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