Module III.2

Childhood epilepsy

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Childhood epilepsy

1.1.2 Classification of epilepsies and epileptic syndromes according to age

1.1.2. 1 Epilepsy and neonatal period (birth to 2 months)

1.1.2.2 Childhood-onset epilepsy (2 months to 12 months)

1.1.2.3 Childhood-onset epilepsy (from one year to 12 years)

1.1.3. Neuropsychology of Epilepsy

1.1. Concept of Childhood Epilepsy

Epilepsy is one of the most frequent neurological disorders that occur during the neonatal and infantile stage and that has a greater biopsychosocial impact on the child who suffers from it.

Epilepsy can appear as a result of CNS disorders (brain infection, toxic, metabolic disorders, genetic malformations and acquired brain damage).

According to the International League Against Epilepsy (ILAE) epilepsy is classified:

Differentiating on the one hand the epileptic seizures and on the other hand

Categorizing the types of epilepsy and epileptic syndromes.

1.1. Concept of childhood epilepsy

Epilepsy could be defined by the presence of at least **two unprovoked or reflex epileptic seizures** (induced by a stimulus: light, auditory, tactile, etc.) that happen separately on different days.

Epileptic seizures are the abnormal transient discharge of synchronous neurons from the cerebral cortex that produces a **clear effect observable** by the person experiencing it or by an observer.

If the discharge affects a specific area of the cerebral cortex, this type of crisis will be focal, however, if the initial discharge simultaneously affects both hemispheres, the crisis will be generalized.

Childhood epilepsy

1.1. Concept of Childhood Epilepsy

Epileptic seizures are classified according to the onset of the abnormal discharge that gave rise to them, therefore, there are two types:

1. Focal: epileptic seizures that originate in a localized area of the cerebral cortex (epileptic focus)

to. Crises with motor, sensory or psychomotor manifestations that depend on the location of this focus.

b. They do not initially produce loss of consciousness.

2. Generalized: they affect simultaneously and from the beginning the entire cerebral cortex.

to. They cause loss of consciousness since the beginning of the crisis

b. The most common generalized crises are tonic-clonic seizures

1.1. Concept of Childhood Epilepsy

Epileptic syndromes refer to the association of one type or more types of seizures with interictal (during the seizure) or ictal (the epileptic seizure itself) electroencephalographic alterations that compromise the proper functioning of the central nervous system (CNS) or without it, the age of onset and other components, such as the severity, treatment and evolutionary course of these syndromes.

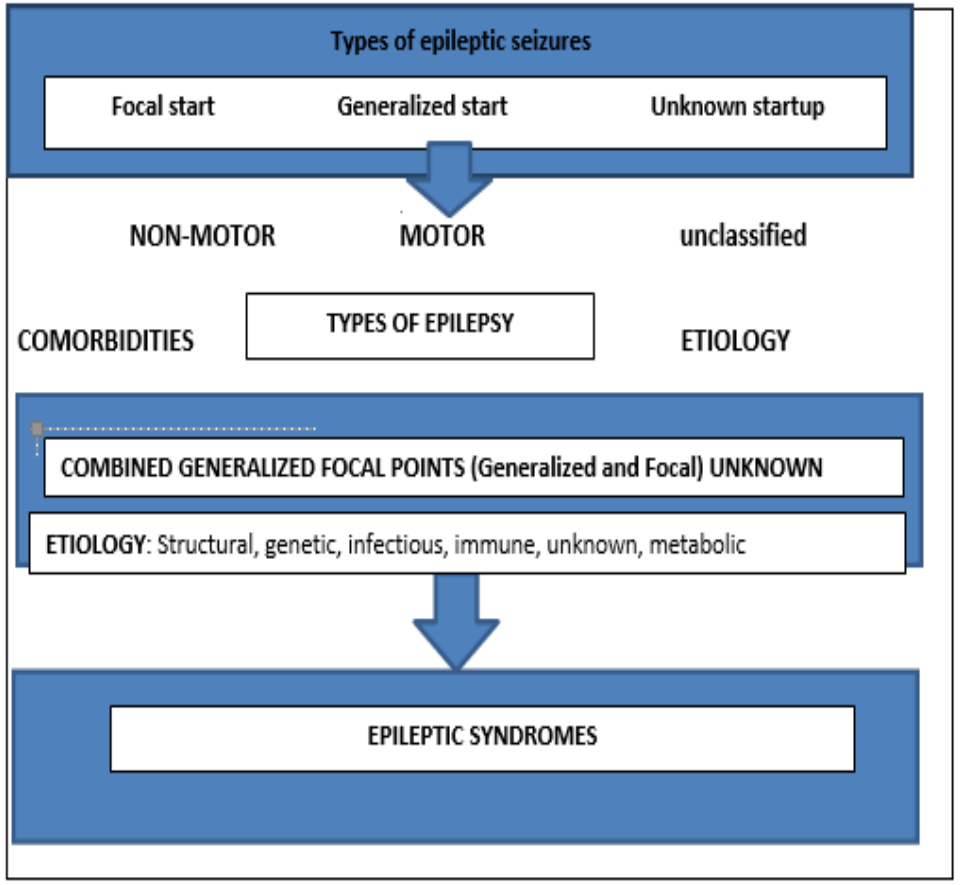
The most common forms of **Epileptic syndromes are age-dependent or self-limiting,** which means that these epileptic seizures will remit or disappear definitively with the maturation of the brain, these syndromes being the most frequent, particularly in school-age children and who also respond very well to treatment with antiepileptic drugs.

1.1. Concept of Childhood Epilepsy

Classification of epilepsies and epileptic syndromes according to age:

The classification proposed by the ILAE, is created to respond to the categorization of epilepsy in a clinical context, being necessary for the diagnosis, the classification in three levels.

It is necessary to differentiate between the type of epileptic seizure, epileptic syndrome and the type of epilepsy



1.1. Concept of Childhood Epilepsy

Classification of epilepsies and epileptic syndromes according to age:

Once the seizure is identified, the next step is to classify the type of epilepsy that will be part of an epileptic syndrome.

According to the classification made by the ILAE (2017) epileptic syndrome is classified into four types of epilepsy which are:

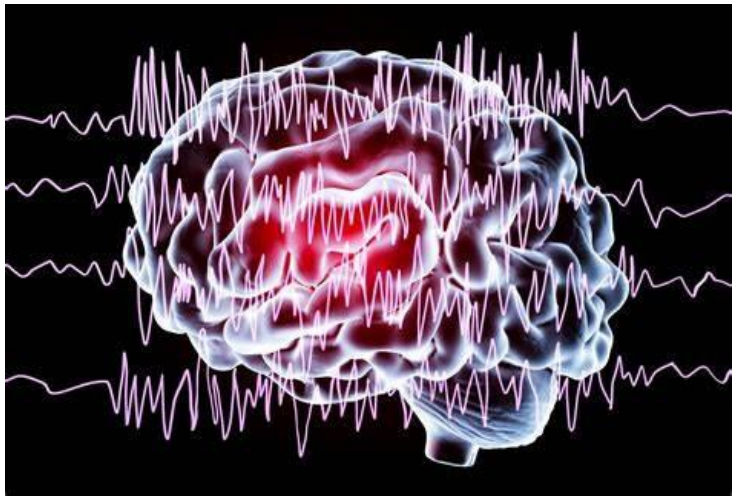
1. Focal epilepsy (motor onset or non-motor onset)

2. Generalized epilepsy

3. Focal or generalized epilepsy (combined)

4. Unknown, it is not known whether its origin is focal or generalized.

a. Symptomatic or probably symptomatic



1.1. Concept of epilepsy

Classification of the type of epileptic seizure (ILAE)

Focal epilepsy: is associated with the presence of abnormalities in a specific area of the brain and may include behavioral alterations analogous to the functions of the region where they originate.

Focal seizures they can be with motor start or with non-motor start.

Motor-onset symptoms involve automatisms, atonic spasms, clonic, epileptic, hyperkinetic, myoclonic, or tonic spasms.

Symptoms with non-motor onset are, behavioral detection, cognitive, emotional, sensory alteration.

Generalized epilepsy is characterized by generalized tonic-clonic seizures associated with paroxysms and a tip EEG or generalized wave polypuntas

Generalized seizures: are associated with the presence of diffuse abnormalities or bursts of abnormal brain activity that result in loss of consciousness, but without more specific sensory or behavioral characteristics.

Crises of unknown cause seizures that cannot be classified due to lack of information or cannot fall into a certain diagnostic category.

Symptomatic or probably symptomatic

1.1. Concept of Epilepsy

Evolutionary development of epilepsy

Brain maturation is a process characterized by the presence of innumerable transformations, produced from conception, throughout gestation and later, until reaching maturity reaching an adult brain.

Within the age classification of epilepsy in the **neonate-infantile period,** we highlight:

1. Epilepsy and neonatal period (birth to 2 months)

2. Childhood-onset epilepsies (2 months to 12 months)

3. Childhood-onset epilepsies (from one year to 12 years)

1.1.Childhood epilepsy

1.1.1. 1. Epilepsy and neonatal period (birth to 2 months)

An especially vulnerable period for seizures to occur because of the combination of specific factors in a developing CNS.

Neonates have a highly excitable brain so the clinical expressiveness of a crisis at this age is of a focal type by neuronal discharges of erratic origin in one or another hemisphere.

The brain at this stage of neurodevelopment is manifested by presenting a bioelectric continuity, interhemispheric synergy, wake-sleep differentiation and reactivity to external stimuli in sleep.

Neonatal seizures are classified into clonic, tonic and myoclonic

1.1.1.1. Epilepsy and neonatal period (birth to 2 months)

Clonic crises: they are rhythmic jerks of muscle groups and can follow both a focal and multifocal pattern. Movements can oscillate from one part of the body to the other.

they can also affect events that affect the brain in a diffuse way, such as suffocation, subarachnoid hemorrhage, hypoglycemia and infections

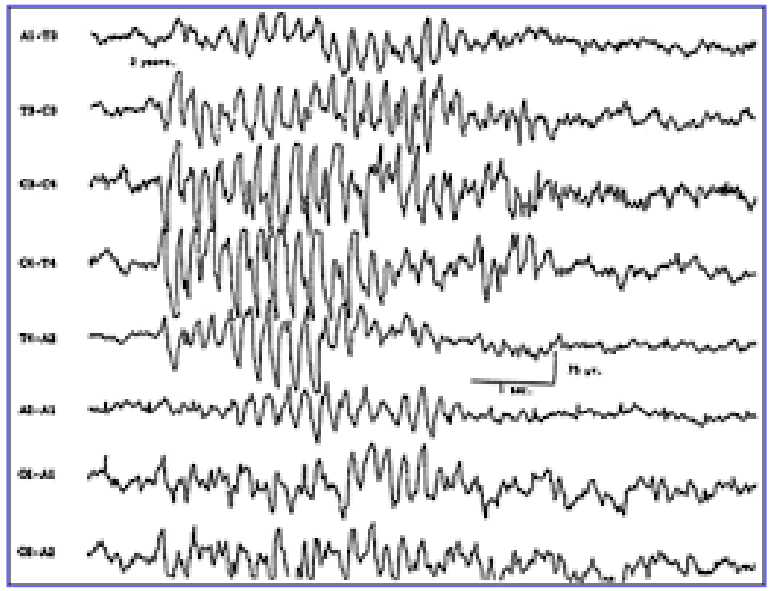
Tonic crises: the neonate adopts asymmetrical postures of the trunk or a deviation of the eyes to one side occurs

Myoclonic crises: crises very similar to those that affect older children and consist of the presence of rapid jerks of the muscles. These crises manifest themselves in the form of bilateral shocks, although occasionally a unilateral or focal myoclonus may appear.. .

1.1.1.1 Epilepsy and neonatal period (birth to 2 months)

Neonatal epileptic syndromes, as well as their electroclinic characteristics, are:

1. **Benign neonatal seizures (fifth-day seizures)**
2. **Benign neonatal epilepsy (ENBF)**
3. **Early childhood epileptic encephalopathy or Ohtahara syndrome.**
4. **Early myoclonic epileptic encephalopathy (EMP).**



1.1.1.1 Epilepsy and neonatal period (birth to 2 months)

Benign neonatal seizures (fifth-day seizures)

Unilateral, bilateral or migratory clonal movements of the limbs and face that last minutes, and apnea may appear. Seizures usually disappear in most cases and the evolution is favorable.

Familial benign neonatal epilepsy:

Group of autosomal dominant benign epileptic syndromes that begin on the second or third day of life (full-term newborns)

Composed of tonic crises, with autonomic symptoms (vegetative system)

Seizures begin with tonic phase (symmetrical or asymmetrical) associated with apnea/cyanosis and followed by clonic movements, unilateral or bilateral, symmetrical or not.

Brief and very frequent seizures (up to 30 episodes a day). It can also constitute a "fixed gaze"

EEG activity shows small focal or multifocal abnormalities. Remission of seizures occurs around 4 to 6 months of age.

Neurodevelopment is usually normal and in some percentages of these children, they may have febrile or afebrile seizures in childhood after a period without seizures. (Fons-Estupiña, 2018).

1.1.1.1 Epilepsy and neonatal period (birth to 2 months)

Early childhood epileptic encephalopathy (Ohtahara syndrome)

Rare epileptic syndrome that **has an unfavorable prognosis.**

The onset of seizures can occur in the fetal period or after birth.

The most frequent type of seizure is tonic, symmetrical or asymmetric seizures, although focal motor seizures can also appear in approximately 30% of these infants.

Among the most frequent causes are malformations in cortical development, genetic alterations related to channelopathies and synaptopathies.

The evolution to infantile spasms or multifocal epilepsy is very frequent. (Fons-Estupiña, 2018).

1.1.1.1 Epilepsy and neonatal period (birth to 2 months)

Early myoclonic encephalopathy

Syndrome similar to Ohtahara's but differentiating the type of crises that are predominantly myoclonus (axial, segmental or erratic).

The frequency of crises can be variable, but it is usually continuous.

The onset of crises is usually early, in the first hours or days of life and in some cases intrauterine.

Seizures are focal or subtle clonic and may be followed by myoclonus. (Fons-Estupiña, 2018).

1.1.1.2 Epilepsy in lactation and early childhood (2 months to 12 months)

Epileptic and specific syndromes that begin between 2 to 12 months, **are symptomatic and probably symptomatic focal epilepsies,** of which are mesial, lateral, frontal, parietal and occipital temporal epileptic syndromes). (Browne et al, 2009).

As for generalized/symptomatic epilepsies, highlight West syndrome, tonic seizures and atonic seizures.

With regard to epilepsies **idiopathic and symptomatic generalized**, there are three types of epilepsy:

. Benign childhood epilepsy with centrotemporal tips,

. Benign early-onset childhood epilepsy (with vegetative symptoms)

. Late-onset childhood occipital epilepsy (with visual symptoms).

Seizures that do not necessarily lead to a diagnosis of epilepsy, are the crisis febriles.

1.1.1.3 Epilepsy in lactation and early childhood (2 months to 12 months)

Within focal and probably symptomatic epilepsies, they can appear at any age.

Three types of crisis

1. Simple focal seizures (focal)

2. Complex focal seizures (psychomotor, temporal lobe)

3. Tonic-clonic seizures (great evil)

Five syndromes where the type of crisis will respond to the location of the epileptogenic zone (temporal lobe, temporal mesial, frontal, occipital, parietal)

Within this group, highlight symptomatic focal epilepsy called **hemiconvulsion-hemiplegia syndrome**, A rare form of epilepsy that begins during the first two years of life.

- Sudden and prolonged unilateral clonic crisis followed by unilateral hemiparesis. (Browne, 2009).

1.1.1.3 Epilepsy in lactation and early childhood (2 months to 12 months)

As for generalized/symptomatic epilepsies, there are **four types of epilepsies** (West Syndrome, Tonic Seizures, Atonic Seizures, and Dravet Syndrome)

West syndrome- most common form in children during **the first year of life,** being the peak age of onset between 4 and 6 months. It is characterized **by a symptomatic triad** of infantile spasms (EI), intellectual disability, and hypsarrhythmic EEG

In the EEG record, a hypsarrhythmic path is observed causing a halt to the neurological maturation process at the beginning of the critical manifestations (during the crisis) infantile spasms appear (sudden, symmetrical and bilateral contraction) that affect the axial muscles and extremities, which determine the appearance of the spasm either in flexion, extension or mixed, being able to be of different intensity (mild or massive) (Browne, 2009).

1.1.1.3 Epilepsy in lactation and early childhood (2 months to 12 months)

Tonic crises: They are brief seizures that consist of the sudden appearance of an increase in tone in the extensor muscles.

The duration of crises is longer than that of myoclonic crises.

Atonic crises:consist of the sudden loss of muscle tone, involves head, trunk, jaw or the muscles of the extremities, which causes falls that causes trauma and injuries from this type of crisis.

1.1.1.3 Epilepsy in lactation and early childhood (2 months to 12 months)

Dravet syndrome (formerly known as severe myoclonic epilepsy of childhood)

It occurs in the first year of life in a normal child with prolonged, febrile and afebrile, focal (usually hemiclonics) and generalized tonic-clonic seizures.

Seizures are usually intractable, and starting in the second year of life, children show cognitive and behavioral impairments. (ILAE, 2017). Over time they may develop ataxia and pyramidal signs

A syndrome characterized by seizures typically around 6 months of age.

Most babies have had an onset of seizures before 15 months of age, however, a small minority of cases begin in the second year of life. The first seizure is associated with fever in about 60% of cases. Not all babies start with febrile seizures. The sensitivity of seizures to fever may persist throughout life.

1.1. Childhood-onset epilepsies (from one year of life)

Among the epileptic syndromes that occur at this stage that begin in childhood, we find:

Symptomatic and probably symptomatic focal epilepsies

Idiopathic focal and generalized epilepsies

Epileptic encephalopathies

Those seizures that do not necessarily lead to a diagnosis of epilepsy.

1.1. Childhood-onset epilepsies (from one year of life)

- **Symptomatic and probably symptomatic focal epilepsies**, the five mesial, lateral, frontal and occipital temporal epileptic syndromes are found.

  These types of syndromes lead to **three types of crisis:** simple focal seizures (focal) crises; complex focal (psychomotor, temporal lobe) and tonic-clonic (grand mal)

Idiopathic focal epilepsies can be the significant component of three major syndromes:

Benign childhood epilepsy with centrotemporal tips

Benign occipital epilepsy of childhood

Epileptic encephalopathy Lennox-Gastau and Landau Kleffner

Childhood epilepsy

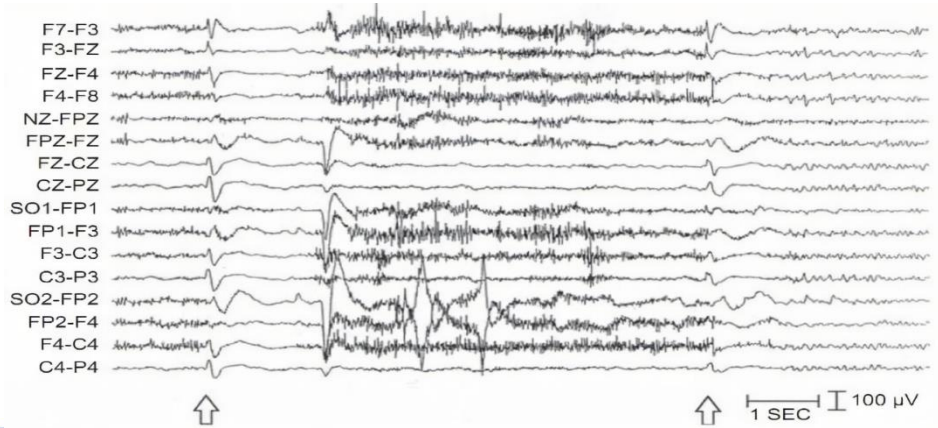
1.1. Childhood-onset epilepsies (from one year of life)

- **Benign partial epilepsy of childhood with centrotemporal (rolandic) tips begins between 3-10 years in healthy children**
- Sensory-motor focal seizures affect the face, oropharynx and upper limb (oro-facio-brachial clonies, speech block and oral paresthesias). Infrequent crises and nocturnal predominance. The EEG shows focus of tips in the center-temporal, uni or bilateral region. (Martínez et al, 2014).
- **Childhood epilepsy with occipital paroxysms,** early onset, known as Panayiotopoulos syndrome, in young children with a peak age of 5 years. Main symptoms: ictal vomiting, deviation of the eyes, and deterioration of consciousness. Infrequent and often solitary crises. The prognosis is excellent and usually resolves within a few years of its appearance.
- ***Epilepsy with absence in childhood:*** typical absence crises, myoclonic and tonic-clonic of generalized onset. Genetic/idiopathic generalized epilepsy in an otherwise normal child with multiple daily absence seizures associated with generalized spikes and waves of 2.5 to 3.5 Hz.
- Crises are caused by hyperventilation, ELI. Frequent absence crises between 2 and 12 years (maximum 5-6 years). Development and cognition are typically normal. ADD and learning difficulty may occur (ILAE, 2017).

1.1. Childhood-onset epilepsies (from one year of life)

Lennox-Gastaut syndrome*:* A severe form of epileptic encephalopathy that begins in childhood.
Children have frequent seizures of various types.
Seizures usually begin between the ages of 2 and 6 and are usually accompanied by intellectual disability.

Tonic seizures are the main component and have a slow-wave peak EEG pattern.
These children present a mixture of different types of seizures, (tonic-clonic, myoclonus, typical absences and head drop) that constitute a form of atonic, tonic or myoclonic crisis.
This syndrome is characterized by having very frequent seizures and it is common for atypical absence crises to go unnoticed by parents and the child (Browne, 2009).



1.1. Childhood-onset epilepsies (from one year of life))

Landau-Kleffner syndrome: subacute onset of acquired aphasia in a child with normal previous development and cognition. The **syndrome begins between 2 and 8 years of age** (maximum between 5 and 7 years)
Seizures may not occur in all cases are usually infrequent and self-limiting.

This syndrome is characterized by a subacute onset of progressive aphasia in a child with previous age-appropriate language development.
Initial presentation may be with progressive aphasia (40%), seizures, or both.
Children become progressively unable to understand the spoken word, stop understanding when spoken to and respond verbally.
Psychiatric and cognitive disorders are commonly observed in addition to language impairment.
Language impairment typically fluctuates. Seizures and EEG abnormalities resolve with age in most cases, however, in most (>80%) residual language impairment is observed that can be severe (especially if onset is earlier). (ILAE, 2017).

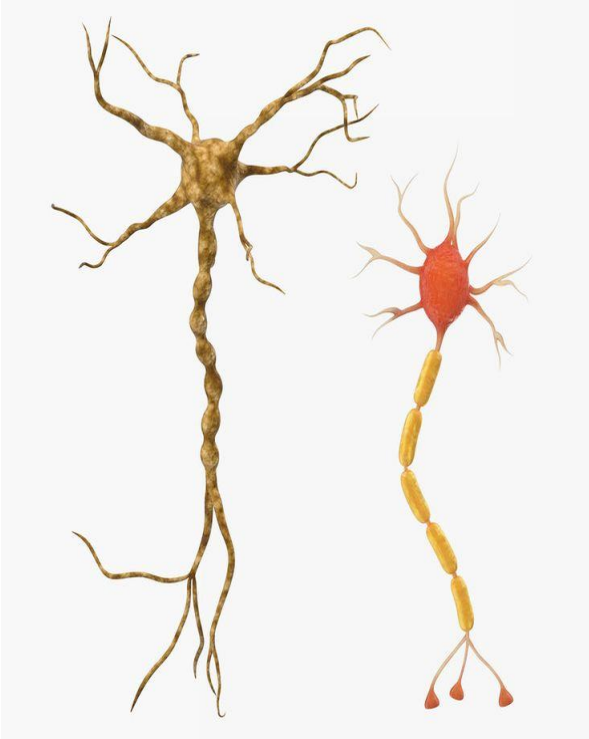
1.1.2. Neuropsychology of Epilepsy:

Epilepsy as such does not produce cognitive impairment, however, if it appears it may be due to the presence of epileptic encephalopathy or an underlying brain injury (Ronconi, 2019).

The process of describing the possible neuropsychological alterations found must be adapted to each specific child.

Another interesting issue is to assess that both behavioral and cognitive difficulties may be due to the impact of this underlying lesion or of the epileptiform activity itself (electric shocks) on maturing neural networks in a developing brain.

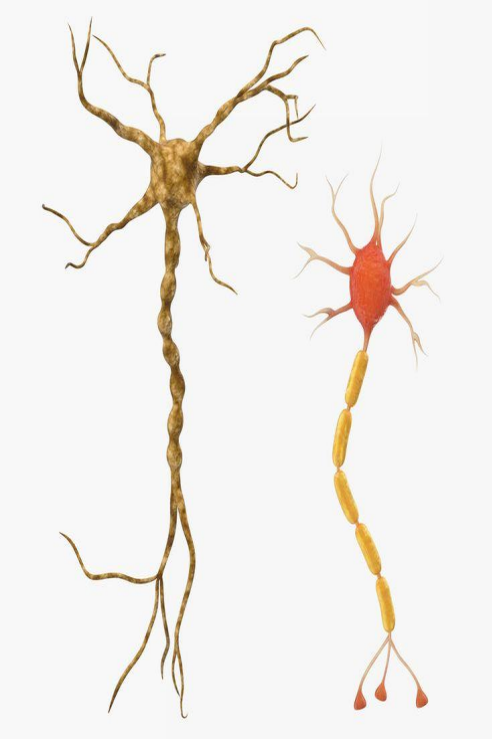
The literature shows data showing decreased neuropsychological performance of students with epilepsy in multiple domains, including general intelligence (Salinas et al, 2018).



1.1. Neuropsychology of epilepsy

Cognitive functioning: important variability within groups, with a higher percentage in children outside the normative values when they are valued with the general child population.

Attention: certain antiepileptic drugs can cause less attention span, concentration, and fatigue. There are also some epileptic syndromes associated with a specific alteration in attentional control, such as, for example, childhood absence epilepsy and ADHD (combined type). Importantly, attention problems may precede the onset of epilepsy.



1.1. Neuropsychology of epilepsy

Executive functions: the effect of epilepsy on the development of executive functions

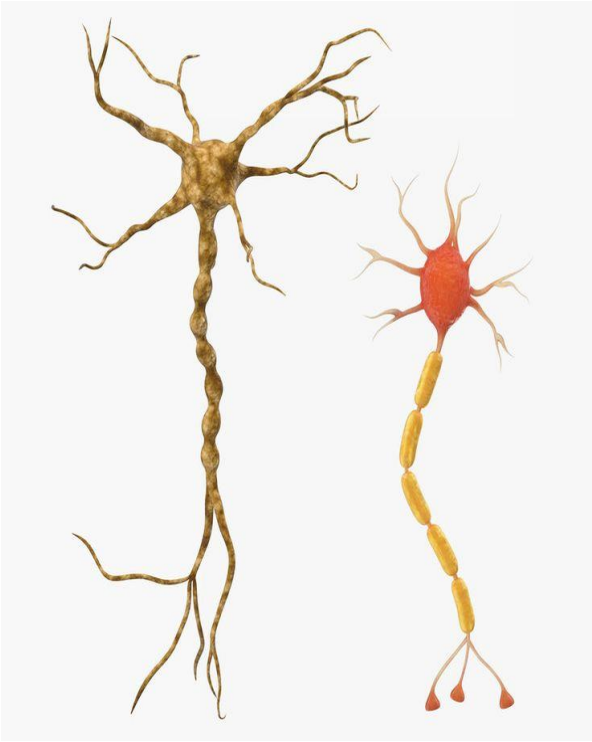
Worse executive performance in generalized epilepsy than in focal-onset epilepsy. The most frequent alterations are:

working memory, processing speed and difficulty in solving problems.

problems in executive functions predictor of adaptability and quality of life in children with epilepsy as significant as the variables related to the disease and its severity

Memory: type of epilepsy and its location (onset versus focal epilepsy) (hemispheric lateralization) affect mnesic performance (retrieval of previously stored information).

Also as a prognostic factor of this performance, assess that focal onset epilepsy that originates in the frontal or temporal region may be a risk factor for observing low performance in memory.



1.1. Neuropsychology of epilepsy

Learning difficulties: consider the presence of low academic performance both of the level expected for their age and course and an academic performance below what is expected at the cognitive level in the child with epilepsy.

In terms of specific learning disorders, math problems are the most prevalent and verbal performance in both reading difficulty and the presence of poor performance on semantic memory tests.

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Images

Image 1: https://babyviewer.blogspot.com/2018/06/how-is-eeg-done-on-baby.html
Image 2: : https://www.canadianepilepsyalliance.org/ilae-2017-new-classification-of-seizure-types
Image 3 https://blog.amigopanda.com.br/epilepsia-infantil/
Image 4: https://babyviewer.blogspot.com/2018/06/how-is-eeg-done-on-baby.html
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