Seizures in childhood

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LEARNING OBJECTIVES:
1. Define seizures and epilepsy.
2. Summarize the classifications of epilepsy.
3. Know the features of the most common types of childhood epilepsies.
4. Understand the importance of EEG in the diagnosis of epilepsy.
5. **Understand the principles of epilepsy management**.

Definitions:

Seizure: is the changes in motor activity and/or behavior that results from abnormal excessive neuronal activity in the brain.

Convulsion: refers to motor manifestation of seizure.

Epilepsy : is a condition in which seizures recur , usually spontaneously (epilepsy is considered to be present when 2 or more unprovoked seizures occur at an interval greater than 24 hr apart) .

The clinical event is accompanied by characteristic EEG changes of either spike or sharp wave (the epileptic discharge); these discharges may be focal or diffuse.

Acute symptomatic seizure: occurs secondary to an acute problem affecting brain excitability such as electrolyte imbalance or meningitis.

Epilepsy syndrome: is a disorder that manifests one or more specific seizure types and has a specific age of onset, specific EEG abnormalities, and a specific prognosis.

Focal (partial) epileptic seizures: arise in specific loci in one part of the cerebral cortex that carry with them identifiable clinical features either subjective or observed. Consciousness may or may not be retained or there may be partial loss of awareness.

Generalized epileptic seizures: involve large areas of brain, usually both hemisphere, and are associated with early impairment of consciousness.

Epidemiology:

Seizure may signal a potentially serious underlying systemic or CNS disorder that requires thorough investigation and management.

Febrile seizures

Is the most common seizure disorder during childhood, occur in approximately 3-4% of young children.

Febrile seizures criteria:

1. Age dependent (occur between the age of 6 to 60 month), the peak age of onset is about 14-18 mo of age.
2. Occur with a temperature of 38 Cº or higher.
3. Not the result of CNS infection or any metabolic imbalance.
4. Occur in the absence of a history of prior afebrile seizures.

Genetic factors:
The genetic contribution of febrile seizure is manifested by a positive family history for febrile seizures. In many families the disorder is inherited as an autosomal dominant trait.

Clinical manifestations:

Febrile seizures can be classified into:

Simple (typical) which characterized by:

1. Represent 75% of all febrile seizures.
2. Manifested by a generalized tonic –clonic seizures.
3. Lasting less than 15 min.
4. Brief postictal period of drowsiness.
5. Occurs only once in 24 hr.

Complex (complicated, atypical) which characterized by:

1. Account for 25% of cases.
2. Focal seizure activity.
3. Lasting over 15 min.
4. Focal findings during the postictal period.
5. Repeated during the same febrile episode.

Febrile status epilepticus: is a febrile seizure lasting > 30 min.

Approximately 30-50% of children have recurrent seizures with later episodes of fever (30% of those experiencing a first episode, in 50% after two or more episodes).

Risk factors associated with increased recurrence of febrile seizures include:

Major

1. Age < 12 mo.
2. Duration of fever < 24 hr
3. Fever 38-39 Cº

Minor

1. Family history of febrile seizures
2. Family history of epilepsy
3. Complex febrile seizure
4. Male gender
5. Lower serum sodium at time of presentation

Having no risk factors carries a recurrence risk of approximately 12%; one risk factor , 25-50%; two risk factors, 50–59%; three or more risk factors, 73–100%.

Most children with febrile seizures have only a slightly greater risk of later epilepsy than the general population. Only 5% of children who experience febrile seizures proceed to develop epilepsy later in life.

Risk Factors for Occurrence of Subsequent Epilepsy after a Febrile Seizure:

1. Complex features.
2. Positive family history of epilepsy.
3. Fever < 1 hr before febrile seizure.
4. Neurodevelopmental abnormalities .

Work-up
1. Lumbar puncture:

The most important responsibility is to determine the cause of fever and to rule out meningitis or encephalitis. Lumbar puncture should be performed for all infants younger than 6 mo of age who present with fever and seizure, if the child is ill-appearing, or at any age if there are clinical signs or symptoms of concern.
2. EEG:
Not indicated if the child has simple febrile seizure and is neurologically healthy because EEG would not predict the future recurrence of febrile seizure or epilepsy even if the result is abnormal.
3. Blood studies: e.g. electrolytes, CBC, blood glucose are not routinely indicated in children with first simple febrile seizure.
4. Neuroimaging:
CT or MRI is not recommended in the evaluation of child with first simple febrile seizures.

Treatment
Routine management of a normal patient with simple brief febrile seizures includes:
1. Careful search for the cause of fever.
2. Reassurance and education of the parents.
Antipyretics can decrease the discomfort of the child but do not reduce the risk of having a recurrence febrile seizure.

Prolonged anticonvulsant prophylaxis for preventing recurrent febrile convulsions is controversial and no longer recommended for most children because of the potential risks of many drugs and the excellent prognosis regardless of treatment.
Also preventive anticonvulsant treatment has not been shown to reduce the risk of later epilepsy in higher risk patients.

Classifications of seizures

It is important to classify the type of seizure, why?
1. May provide clue to the cause of the seizure disorder.
2. Allow to make the prognosis.
3. Allow to choose the appropriate treatment.

International classification of epileptic seizures according to the International League Against Epilepsy (ILAE):

1. FOCAL (PARTIAL) SEIZURES:
A. Focal seizure - awareness retained (focal aware seizure).

B. Focal seizures – awareness altered (focal seizures with impaired awareness).

C. Focal to bilateral tonic-clonic.

2. GENERALIZED SEIZURES
Absences
Generalized tonic-clonic
Tonic
Clonic
Myoclonic
Atonic

3. UNKNOWN SEIZURES
Epileptic spasms
4. **UNCLASSIFIED SEIZURES**

Focal seizures

Account for about 40% of childhood seizures, may be classified into:
1. Focal seizure with preserved awareness (Focal aware seizures).
2. Focal Seizures with impaired awareness.
3. Focal to bilateral tonic-clonic seizures.

Focal seizure with preserved awareness (Focal aware seizure)
1. Brief motor seizures are the most common manifestations and include focal tonic, clonic, or atonic seizure.

2. Versive seizures (head turning and conjugate eye movement) are common.
3. Automatism does not occur.
4. Some patients complain of aura (chest discomfort, headache).

5. The average seizure persists for 10-20 - sec

Focal Seizures with impaired awareness
- Are often preceded by an aura such as rising abdominal feeling and sense of fear.

- Subsequent manifestations consist of decreased responsiveness, staring, looking around seemingly purposelessly, and automatisms.

- Automatisms are common features in children:
The automatic behavior observed in infants is characterized by alimentary automatisms (lip smacking, chewing, swallowing, and excessive salivation), automatic behavior in older children may consist of fidgeting or washing movements of the hand, running or walking.
- The average duration is 1-2 min (longer than focal aware seizure).

Treatment:
Focal seizures can be treated with carbamazepine, oxcarbazepine, valproate, or levetiracetam.

GENERALIZED SEIZURES

Absence seizures:
Typical absence seizures of childhood characterized by:
1. Sudden cessation of motor activity or speech with a blank facial expression and flickering of the eyelids, absence seizure can have simple automatisms like lip-smacking and the head can minimally falls forward.
2. Never associated with aura.
3. Rarely persists longer than 30 sec.
4. Uncommon before age 5 yr.
5. More prevalent in girls.
6. Patients may develop countless (10-100+ times) seizures daily.
7. No postictal impairment with immediate resumption of what the patient was doing before the seizure.
8. Hyperventilation for 3-4 min routinely produces absence seizures.
9. The EEG shows a typical 3/sec spike and wave discharge.

Treatment:
1. Ethosuximide (first choice).
2. Alternative drugs (valproate, lamotrigine).

Generalized tonic-clonic seizures:
It may follow Focal seizure (second generalization) or occur de novo. They may be associated with an aura.
Patients suddenly lose consciousness, their eyes roll back, the entire body musculature undergoes tonic contractions, and they rapidly become cyanotic in association with apnea.
The clonic phase started by rhythmic clonic contractions alternating with relaxation of all muscle groups.
During the seizures the patients may bite their tongue, loss of sphincter control, particularly the bladder, is common during a generalized tonic-clonic seizure.
The attack usually persists for few minutes.
Postictally, children are initially semi comatose and typically remain in a deep sleep from 30 min to 2 hr. the postictal phase is often associated with vomiting and intense bifrontal headache.

Infantile epileptic spasms
West syndrome: is a triad of infantile epileptic spasms, developmental regression, and a typical EEG picture of hysarrhythmia.

Usually begin between the ages of 2-12 mos and are characterized by brief symmetric contractions of the neck, trunk, and extremities.
There are 3 types of infantile spasms:
1. Flexor spasms: occur in clusters and consist of sudden flexion of the neck, arms, and legs onto the trunk.
2. Extensor spasms: produce extension of the trunk and extremities (least common type).
3. Mixed infantile spasms: consisting of flexion in some clusters and extension in others (the most common type).

A cry may precede or follow an infantile spasm, accounting for the confusion with colic in a few cases.
The spasms occur during sleep or arousal but have a tendency to develop while patients are drowsy or immediately on awakening.
EEG findings: is called hypsarrhythmia.

Etiology:
Classified into 2 groups:
1. cryptogenic (idiopathic) account for 10-20% of cases , characterized by uneventful pregnancy and birth history as well as normal developmental milestones before the onset of seizures. CT and MRI scan of the head are normal and there are no associated risk factors.

2. Symptomatic: due to:
A. several prenatal and perinatal factors like hypoxic-ischemic encephalopathy, congenital infections, inborn errors of metabolism, neurocutaneous syndromes such as tuberous sclerosis, congenital malformations of the brain and prematurity.
B. postnatal conditions include CNS infections, head trauma and Hypoxic-ischemic encephalopathy.

Prognosis:
Cryptogenic infantile spasms have good prognosis, whereas those with the symptomatic type have an 80-90% risk of mental retardation. The underlying CNS disorder has a major role in the neurological outcome.

Pathogenesis of infantile spasms:
There are several theories; one of these theories implicates corticotrophin-releasing hormone (CRH) overproduction, CRH act as a neurotransmitter resulting in neuronal hyper excitability and seizures.
Treatment:
West syndrome, especially in cases of unknown etiology *is a medical emergency* because diagnosis delayed for 3 wk or longer can affect long-term prognosis.

1. ACTH is the best treatment.
2. Vigabatrin.
3. Other alternative medications: valproate, clonazepam, lamotrigine, pyridoxine, and IVIG

USE OF THE EEG TO DIAGNOSE EPILEPSY

Demonstration of paroxysmal discharges on the EEG during a clinical seizure is diagnostic of epilepsy.
A normal EEG does not exclude the diagnosis of epilepsy, because the interictal recording is normal in about 40% of patients.
Activation procedures such as hyperventilation, eye closure, photic stimulation and sleep deprivation may increase the positive yield.

TREATMENT OF EPILEPSY

Steps in the management of epilepsy:
1. Ensure that the patient has a seizure disorder and not a condition that mimics epilepsy.
2. Antiepileptic should not be used in a previously healthy child with the first afebrile convulsion if there is a negative family history, normal results of physical examination and EEG, and a cooperative and compliant family, because of low recurrence risk (20% risk). Approximately 70% of these children will not experience another convulsion.
If the patient has an abnormal EEG, MRI, development status, and/or neurologic exam and/or has a positive family history of epilepsy the risk is higher, and often treatment is started.

3. A recurrent seizure, particularly if it occurs in close proximity to the 1st seizure, is an indication to begin an anticonvulsant.
4. Choosing the appropriate anticonvulsant depends on the classification of the seizure, determined by the history and EEG findings.
5. The goal for every patient should be the use of only one drug with the fewest possible side effects for the control of seizures .The drug is increased slowly until seizure control is accomplished or until undesirable side effects develop.
6. If complete seizure control is achieved by an anticonvulsant, a minimum of two seizure-free years is an adequate and safe period of treatment.
7. The chance of recurrence is 20-25%, particularly in the 1st 6 mo after discontinuation of the anticonvulsant. When decision is made to discontinue the drug, the weaning process should occur over 3-6 mo, because abrupt withdrawal may cause status epilepticus.
Factors associated with a higher risk of seizure relapse after antiepileptic drug withdrawals are:
1. Older age of epilepsy onset.
2. Longer duration of epilepsy.
3. Presence of multiple seizure types.
4. Need to use > 1 antiepileptic drug.