

EPILEPTIC SEIZURES & SYNDROMES

-Dr Surekha Rajadhyaksha

Introduction

Convulsions are attacks of involuntary muscle contraction, either sustained, (tonic), or interrupted, (clonic,) e.g. - febrile convulsion, hypoglycemic convulsion.

Epilepsy - In the past term 'epilepsy' was loosely applied to any paroxysmal clinical events and was confused with hysterical fits, syncope, etc.

Epilepsy is a chronic condition characterised by the repeated occurrence of epileptic seizures.

Seizures are clinical events during which abnormal and excessive activity of a group of cerebral neurons occurs.

The excessive activity of cerebral neurons is demonstrated by the paroxysmal change in EEG activity which may be diffuse or localised.

Earlier epileptic seizures were simply described as big (grand mal) or small (petit mal) and later, terms such as psychomotor epilepsy and minor motor epilepsy were coined.

Confusion existed in the minds of physicians regarding the interpretation of these terms and in order to develop a uniform terminology the ILAE published the International Classification of Seizures in 1969.

This was the first time that the concept of relating a clinical event to ictal and inter-ictal EEG was introduced and a clear distinction was made between seizures that were generalised from the beginning to those that were partial or focal at onset and later became generalised.

This was followed by the era of intensive EEG and video monitoring which led to improved descriptive accuracy in the sequence of an individual seizure and a distinction could be made between simple partial (SP) and complex partial (CP) seizures. By repetitive replay of videotapes one was able to discern whether the patient's awareness and responsiveness was maintained or impaired. If a patient is aware and can recall the attack it is a simple seizure and if awareness and responsiveness is altered it is a complex partial seizure.

When we say partial seizures we mean that the initial epileptic discharge originates and remains confined to a restricted area of cerebral cortex. This is so in SP. If this discharge is in parts of limbic system and its projections-then it results in CP.

The epileptic discharge which was initially (?) may then spread Bilaterally to the entire cerebral cortex to trigger a generalised seizure. (P-GTC)

Simple partial (SP) - consciousness is preserved
Complex partial (CP) -consciousness is impaired

The seizure may progress from SP - CP - GTC
termed as P - GTC

In generalised epilepsy, relatively simultaneous bilaterally large parts of cerebral hemisphere are involved. The genesis of spike and wave activity occurs in cortical structures and spreads via a corticoreticular cortical loop.

Classification of seizures

1. Partial Seizures

Simple (consciousness not impaired)

- With motor, somatosensory, special sensory, autonomic or psychic symptoms.
- Complex (with impairment of consciousness)
- Beginning as simple partial seizures, progressing to complex seizures with or without automatisms.
- May progress to become secondarily generalised.

2. Generalised Seizures

- Absence seizures (typical or atypical)
- Myoclonic seizures
- Clonic, tonic or tonic clonic seizures
- Atonic seizures

3. Unclassified Seizures : Whether Partial or Generalised

Clinical Features

Typical absence : Blank stares, total amnesia, slight motor changes such as blinking, eye rolling or jerking. Sudden onset and cessation. Mostly associated as a part of a syndrome of idiopathic epilepsy.

Atypical absence : Blank stares, consciousness partly impaired, associated with tonic changes, automatisms and seizures are usually prolonged. Usually seen in patients with diffuse cerebral and mental retardation.

Myoclonic seizures : Brief, occur singly or in series. The intensity varies from slight movement to massive jerking leading to a fall. May be stimulus-sensitive; consciousness is not altered; occur as a part of various idiopathic and symptomatic syndromes.

Atonic seizure : Atonia with fall, astatic or drop attacks. Attacks may progress in step-wise fashion and are commonly seen in patients with diffuse brain damage.

Automatisms : Involuntary motor activity occurring during the state of clouding of consciousness either in the course of or after the epileptic seizure. They may be in the form of chewing, lip smacking, drooling, laughing, crying, anger, gestural, fiddling, tapping with fingers, rubbing, patting, undressing ambulatory , etc.

Specific features of seizures arising in different anatomical area :

Temporal lobe : Auras are common, slow evolution of seizures, prominent motor arrest or motionless stares, post ictal confusion with automatisms such as lip smacking, chewing, swallowing, fumbling, repetitive motor activity and other motor automatisms.

Duration of seizure : 15 seconds - 8 minutes, may have secondary generalisation.

Etiology : Both organic and unknown insult. Is seen following status epilepticus or following prolonged unilateral febrile convulsions. Usual pathology is mesial temporal sclerosis, neuronal migration defects etc.

Frontal lobe : Frequent attacks, more at night, prominent ictal tonic posturing, in clusters, sudden onset and cessation with bizarre automatisms.

Central Region :

Simple partial onset, contralateral side jerking. The seizure may have a Jacksonian march to motor cortex. If supplementary motor area is affected adverse head and eye deviation may occur.

Parietal And Occipital Lobe :

Subjective sensory and visual disturbances. The 'limitation' of the above classification was that it was confined to an individual seizure type. A seizure is an event with which a patient come to a physician but the condition to which this seizure belongs is the epileptic syndrome and it is also the language in which two physicians can communicate.

What are syndromes ?

Syndromes are clusters of signs and symptoms customarily occurring together and include

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Lennox - Gastaut Syndrome

Incidence- 3% of childhood seizures.

Etiology : Many causes. 25% are Cryptogenic.

Cardinal features :

- Neurologically abnormal children
- Atypical absences
- Axial tonic seizures
- With sudden falls (atonic/Myoclonic)
- Progressive mental deterioration

Age 2-8 years (3-5 years), seen more in males with no family history
EEG: shows diffuse slow spike and wave (1-2.5 Hz), multifocal abnormalities during inter ictal period and abnormal background activity.

Seizures :

- Tonic seizures are a necessary component.
- Atypical absences commonly seen.
- Massive myoclonic jerks, myoclonic atonic seizures.
- Episodes of status epilepticus lasting for days.
- Recurrent loss of tone and falls forward with facial injuries.
- Myoclonic twitches of eyelid and mouth observed.

Resistant to treatment; the following treatment has been tried :
Sodium valproate, nitrazepam, clonazepam, vigabatrin, felbamate.
ACTH, IV gammaglobulin, TRH
Ketogenic diet
Corpus callosotomy.
Prognosis: For seizure control and mental development is poor.

Myoclonic Astatic

Incidence -1-2% of all epilepsies.

Age 2-5 years

Characterised by high genetic predisposition

Normal development and no neurological deficits before onset.

Seizure type - Primarily generalised myoclonic, myoclonic astatic. Short absences with myoclonic jerks. Generalised tonic clonic seizures in 75%. History of febrile seizures in 28%

EEG - Irregular spike and wave activity in sleep Bilateral synchronous irregular spike and wave activity. Paroxysms of irregular spike, wave and polyspikes Photosensitivity No multifocal abnormality

Prognosis related to seizure frequency. Spontaneous remission occasionally occurs 50% continue to have seizures.

Later childhood (5-10 years)

Typical absence epilepsy Benign partial epilepsy with rolandic (centro-temporal) spikes Benign partial epilepsy with occipital occipital spikes/ paroxysms Landau - Kleffner syndrome

Childhood Absence (Petit mal)

Onset : Before puberty (5-10years)

Previously normal children, more often in females.

Type of seizure : Absences are the initial presenting seizures which are short in duration with abrupt onset and termination and high frequency of seizures per day. There is impairment of consciousness with total amnesia of the event. This can be easily done at the bedside by asking the patient to hyperventilate for 2- 3 minutes and precipitate a seizure.

EEG : shows a bilateral, symmetrical, synchronous discharges of spike and wave at 3c/second, with a normal background activity and precipitated by hyperventilation, with an abrupt onset and termination on spike and wave complexes.

Simple absences seen in 10%

Complex with mild motor component seen in 45%. There may be increased postural tone, decreased postural tone or if prolonged then associated with automatisms.

Prognosis : There are no recognisable focal deficits and seizures do not persist beyond adolescence. Later may develop generalised tonic clonic seizures in 40%.

The response to ethosuximide, valproate or clonazepam is complete.

Benign Partial Epilepsy with Centrotemporal Spikes (BECT-Rolandic)

Age of occurrence : 3-10 years

History of previous febrile seizures seen in 7-9%

Family history of epilepsy or abnormal EEG in 40%

Neurologically : Normal children

Seizures: are many, brief, in clusters, mainly focal and involving face and oropharynx with clonic jerking of face and mouth with speech arrest, guttural sounds and drooling, somatosensory or may progress to generalised tonic clonic seizures. Two thirds occur in sleep.

EEG : Large, diphasic, high voltage centrotemporal spikes, followed by slow waves, seen in clusters. Unilateral in 60% and bilateral in 40% more prominent in sleep with a normal background activity.

Prognosis is excellent and recovery is the rule without recurrence after discontinuation of medication.

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Benign Epilepsy with Occipital Paroxysms (BEOP)

Age of onset: 15 months -17 years (mean age 7.5 years)

Neurologically - Normal children

Seizures which include visual symptoms amaurosis, hemianopia, visual hallucinations and illusions, followed by motor or psychomotor manifestation and at times terminates with a migrainous headache.

EEG : Normal background, spike and waves occurring in distinct paroxysms over the occipital and posterior temporal region in bursts or in trains with prompt disappearance on eye opening. Reappearance on eye closure after a latent period.

Prognosis is usually good but not as good as BECT. Seizures cease in adult life.

Landau Kleffner : The Acquired Aphasia with Epilepsy Syndrome

Age of onset: 3 years -9 years

Abrupt or gradual onset.

Defect in language function and verbal auditory with an inability to comprehend language and reduction in spontaneous oral expression. Behavioural and intellectual difficulties are present. Seizures are partial or generalised seen in two thirds of cases and not very severe. They remit by 15 years.

EEG-Multifocal spikes and spike and wave discharges.

Long term recovery of language function is not very good especially in those with early age of onset before age of 5 years.

Early diagnosis is essential as treatment with ACTH or steroids can reverse or arrest the disorder. Valproate or carbamazepine are used for seizures.

Adolescence

Juvenile absence epilepsy.

Juvenile myoclonic epilepsy (Janz syndrome)

Grand mal seizures on awakening

Juvenile Absence

Age of occurrence is around puberty with equal male - female rating.

Seizures: As opposed to childhood absence the seizures are infrequent, longer duration and less impairment of consciousness.

Patients may have generalised tonic clonic seizures on awakening or myoclonic seizures.

EEG shows generalised spike and wave discharges with frontal predominance which are more than 3 c/sec. Precipitated by hyper ventilation and sleep deprivation.

Response to treatment is seen not as readily as childhood absence.

Juvenile Myoclonic Epilepsy of Janz. (Impulsive petit mal)

Incidence 5-10% of epilepsy

Age of occurrence : 8-26 years (more between 12-18 years), more in females 2:1 ratio

Seizures : three types of seizures are seen, typical absence seen first, followed by myoclonic jerks and later by generalised tonic clonic seizures. Myoclonic jerks are usually bilateral, arrhythmic, irregular, rapid, more pronounced in the upper limbs. No disturbance of consciousness. The severity of myoclonic seizures is variable and may cause the patient to fall. The seizures are precipitated by sleep deprivation and alcohol. 50% of JME are photosensitive. Myoclonic and generalised tonic clonic seizures occur more often on awakening and patients experience difficulty brushing teeth or carrying a cup in the morning.

Patients have normal intellect and development and no neurological deficits.

EEG: 4-6 Hz polyspike and slow wave generalised discharges with normal background activity and are also found inter ictally. No Close correlation between EEG spikes and jerks.

Inheritance : Gene responsible for the syndrome has been localised to chromosome 6.

Treatment : With appropriate medication (valproate) patients are seizure free. There is high relapse rate if medication is withdrawn even if seizure free for several years.

Epilepsy with Generalised Tonic Clonic Seizures on Awakening

Incidence : 16-52% of all epilepsies

Age : 9-25 years (peak at puberty)

Seizures : Generalised tonic clonic

- Seizures within two hours of waking from sleep, (whatever time of day.)
- Second seizure peak : evening period of relaxation.

Absences in 50%

Myoclonic seizures in 30%

Neurological examination is unremarkable.

EEG shows 2.5 -4 Hz spike wave activity enhanced by hyperventilation in some, photosensitive in about 13%.

Prognosis : Controlled with medication in 65% but high relapse rate if medication is tapered.

Miscellaneous syndromes

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Chronic Progressive Epilepsia Partialis Continua (EPC) Rasmussen's Syndrome

Age of onset : Mean age 6.8 years, 85% before age of 10 years. Neurological and psychological deterioration in previously normal children Cytomegalovirus genomic material may be associated.

Clinical course in three stages :

First stage : Simple partial seizures, complex partial, EPC can occur, but no motor deficit seen, seizures are unresponsive to antiepileptic drugs and gradually increase.

Second stage : Neuropsychological deterioration, progressive hemiparesis, hemianopia and hemisensory loss and if dominant hemisphere involved then results in dysphasia. Epilepsia partialis continua are spontaneous regular or irregular clonic twitching of cerebral cortical origin, sometimes aggravated by action or sensory stimuli confined to one part of body, and continues for hours, days or weeks. It is seen in about 56% cases.

Third stage: Arrest of neuropsychic deterioration and seizures decrease in frequency. The arrest can occur 2 months to 10 years after onset.

EEG : shows progressive brain atrophy and areas of increase T2 weighted signal intensity in the affected hemisphere.

Brain biopsy shows perivascular cuffing, glial nodules, microcystic degeneration with neuronal loss.

Treatment :

Anti epileptic drugs not effective

High dose steroids

IV gamma

Interferon

? Gancyclovir

Functional hemispherectomy

Photosensitive Epilepsy

Photosensitive disorder

Age of onset : 12-14 years, more common in females.

40% are pure photosensitive, 60% have spontaneous along with precipitation by photic stimulation.

Seizures are tonic clonic in 84%, absence in 6%, partial in 2.5%

Flickering lights, TV, video games tend to precipitate seizures.

Treatment is to view TV under bright light from a distance of polaroid lens, dark glasses and sodium valproate.

EEG : Intermittent photic stimulation at 15-18 Hz flash frequency elicit photoconvulsive response.

Isolated Seizure

- Recurrence after first seizure: 51-55%
- After second seizure; 90%

- Recurrence more often in:
 1. Neurologically abnormal
 2. EEG showing focal activity
 3. Complex partial seizure
 4. With history of neonatal seizures
- Withhold treatment pending second seizure in :
 1. Neurologically and mentally normal children
 2. EEG normal / showing generalised neuronal hyperexcitability.
 3. History and EEG suggestive of Rolandic epilepsy

Continuous Spike and Wave During Slow Sleep (CSWS), or Electrical Status Epilepticus During Slow Wave Sleep (ESES).

Essential component is that spike and waves recorded on the EEG occupy 85% of time of slow wave sleep. Long-lasting syndrome and ESES is present for months to years but eventually disappears spontaneously or following therapy but leaves behind cognitive deficits.

Seizure semiology varies from infrequent nocturnal seizures, motor seizures, absences and frequent falls of any type.

Intellectual deterioration is a constant feature after onset of CSWS.

Electrical status occurs only during sleep, day after day.

CSWS may appear in diverse clinical setting either in previously normal delayed children.

20-30% have identifiable brain pathology.

Half have normal mental development before status.