

Approach to a child with Epilepsy

-Dr K.N. Shah

Epilepsy is a very common disorder constituting 70% of all pediatric neurological disorders. Of all epilepsies 70% have their onset in the pediatric age group.

Table 1
Epilepsy in children : advances in recent years
<ol style="list-style-type: none">1. Classification of seizure types (1981)2. Classification of syndromes (1989)3. Neuroimaging (CT, MRI, SPECT, PET)4. Ambulatory EEG5. Long term video EEG monitoring6. Knowledge of neurotransmitters7. Pharmacokinetics of AEDs8. Pathophysiology of Epilepsy9. Newer digital EEG machines

1. History taking
2. Rule out non-epileptic paroxysmal events
3. Type of epileptic seizures
4. Type of epileptic syndromes
5. Etiology of syndromes
6. Rx of epilepsy

FIRST STEP

- History taking**
- Patient or eye witness
 - GTC easy to diagnose
 - Myoclonic, Partial, Complex Partial @ Observation - not always correct. Aura in a child difficult, frequency and duration of spell difficult to remember

SECOND STEP

Once certain about paroxysmal events @ rule out non-epileptic paroxysmal events

1. Febrile seizures are very common and are divided into typical and atypical. Atypical ones are more likely to develop future epilepsy. They disappear after 6 years of age, EEG is normal in the majority of cases.
2. Breath-holding spasms which is very often misdiagnosed as epilepsy proper history taking is very important and they disappear after 3-5 years.
3. Migraine - which is not uncommon in children and can mimic various aura of epilepsy and can have focal seizures.
4. Pseudo seizures can mimic frontal lobe seizures and simultaneous video EEG helps in its differentiation from true epilepsy.
5. Syncope (Reflex, cardiac). Syncope may have mild GTC.
6. Situation related seizures are common in children due to electrolyte disturbance, meningoencephalites, tumors, infarcts etc. By definition, seizures occur during acute illness within 7 days and if they persist, then such cases are diagnosed to be suffering from secondary, symptomatic epilepsy.
7. Recurrent abdominal pain and cyclical vomiting are also very common and come in the differential diagnosis of TLE.
8. Narcolepsy, cataplexy, spasms nutans, paroxymal vertigo, apnoea in new born, behavioural outbursts like temper tantrums, crying episodes etc.

A thorough history taking and EEG evaluation are very crucial in differentiating above conditions from epilepsy.

Hallmarks of epileptic seizures:

1. Recurrent
2. Spontaneous, unprovoked, sudden
3. Brief

Sensory, motor, behavioural and emotional functions can be recurrent, spontaneous and brief and epilepsy should be suspected in such circumstances.

Single, isolated but unprovoked, sudden, spontaneous, brief episode by definition is not considered to be epileptic and should not be treated although from the history it appears to be epilepsy.

In view of social and economic implications, as a rule epilepsy should not be diagnosed without unequivocal clinical evidence, supported by EEG and other investigations. Normal EEG does not rule out epilepsy and when in doubt, it is better to observe the child.

Once epilepsy is diagnosed :

THIRD STEP

What type of epileptic seizure is the child suffering from ?

I .Generalised seizures

1. Generalised tonic-clonic seizure
Generalised tonic seizure
Generalised clonic seizure
2. Myoclonic seizure
3. Atonic seizure
4. Absence seizure
5. Infantile spasms.

II. Partial seizures

1. Simple where consciousness is not lost and the child remains responsive to commands. This may be motor, somato-sensory, autonomic or psychic.
2. Complex partial seizures: Simple partial seizures may progress to complex partial seizures with loss of responsiveness or consciousness or consciousness is lost at the onset followed by seizures.
3. Secondary generalised seizures which occur when simple or complex partial seizures become generalised. Commonly into tonic-clonic seizures.
4. Unclassified: Few types of seizures cannot be classified into any definite category e.g. neonatal seizures

ILAE (1981) classified various seizure types based on clinical, routine scalp EEG (ictal and inter ictal) and in few cases video EEG studies. As video EEG studies are done at very few centers in India and EEG may be normal in some cases, classification depends on history and observation of episodes.

Symptoms according to localisation in partial seizures :

Partial seizures can arise from the frontal lobe, temporal lobe, occipital lobe and parietal lobe. Temporal and frontal lobe partial seizures are the commonest. It is not always easy to differentiate clinically because of overlap in semiology and rapid spread from one area to another which may ultimately lead to secondary generalisation. Scalp EEG also may or may not localise the site and may show multifocal neuronal hyperexcitability.

FOURTH STEP

What is the nature of an epileptic syndrome?

Epilepsy seizure type is one which patient presents with while epilepsy syndrome is decided by the clinician, based on age, semiology of seizures, etiology, anatomy, precipitating factors, severity, chronicity, diurnal variation, prognosis and response to treatment. It is a cluster of signs and symptoms occurring together. Seizure type does not take into account etiology, age of onset, precipitating factors, chronicity etc.

Syndromes are divided into localisation related or generalised

Localisation related syndromes are further divided into

1. Idiopathic
2. Symptomatic, and
3. Cryptogenic

Generalised are further divided into

1. Idiopathic
2. Cryptogenic or secondary
3. Specific syndromes
4. Some of the syndromes remain undetermined

Approach to a child with Epilepsy

-Dr K.N. Shah

FIFTH STEP

What is the etiology of the syndrome?

1. Symptomatic - known CNS disorder
2. Idiopathic - no known cause
3. Cryptogenic - presumed to be symptomatic but no known cause.

In our experience, symptomatic constitutes 48% and idiopathic 52%. Incidence of symptomatic is higher as compared to figures from developed countries because of perinatal insult and CNS infections which are very common. Other causes of symptomatic epilepsy are tumors, intracranial granuloma, (ICG), A.V. malformations, trauma, vascular strokes, congenital malformations inborn error of metabolism etc. Since the advent of CT scan, ICG are more commonly diagnosed.

To diagnose a syndrome,

1. Thorough history of age of onset, developmental milestones, antenatal, perinatal events, family history of febrile convulsion and epilepsy, precipitating factors, trauma, intracranial infections, history of febrile and neonatal seizures, should be taken.

2. Detailed physical examinations :

CNS disorders :

- Anthropometry
- Size of the head (micro/macro)
- Dysmorphic features
- Congenital malformations
- Chromosomal disorders
- Skin (Sturge Weber, tuberous sclerosis, neurofibromatosis)
- Intracranial brui transillumination, cerebral palsy, abnormal movements.

Non neurological disorders :

- Congenital cyanotic heart disease
- Renal and liver disease
- Hypoglycemia
- Hypertension
- Hypocalcemic states
- Organomegaly - storage diseases
- Eyes, fundus (for cherry red spots and storage disorders)

History of dreamy states

- Learning disorders
- Attention deficits
- Cognitive impairment due to
 - *Seizures
 - *Previous CNS pathology
 - *AEDs
- Family problems
- School problems (Pseudo seizures)
- Determine IQ and DQ

Hyperventilation for 3-5 minutes is a very simple bedside test to rule out absence because it precipitates seizures in majority of patients.

Investigations :

1. **Scalp EEG** - is a very important non invasive gold standard in the investigation of epilepsy. First record is positive in 40-50% of cases and with repeated recordings it is positive in a majority. It is a supportive evidence and should always be correlated with the clinical picture.

It helps in classifying seizure types and diagnosis of various syndromes. EEG patterns in children are different from adults and newborn records are different from other older age group children. It requires a lot of experience to interpret children's EEGs. Sometimes the EEG is grossly abnormal without clinical seizures in some children with mental retardation. Such cases should not be treated with AEDs. EEG is also useful in structural lesions which shows focal delta bur neuroimaging is far superior in structural lesions.

Simultaneous video EEG recording is extremely useful in determining type of seizures, classification of syndromes, to rule out pseudoseizures and presurgical evaluation of patients. It can be normal in 2-3% of epilepsy particularly simple partial type.

Intracranial EEG recording is done for those cases which are likely candidates for surgery.

2. **Skull x-ray** - does not help
3. **Neuroimaging** - is very useful to rule out structural causes. MRI has replaced CT scan in epilepsy because focal cortical dysplasia, small angiomas, mesio temporal sclerosis, white matter disorders are better appreciated in MRI. Volumetric MRI is still better than conventional MRI. MRI may not diagnose calcified lesions where CT is indicated. Neuroimaging should be done in cases with focal epilepsy, focal CNS signs, focal EEG abnormality, mental retardation, intractable epilepsy etc.

In our country, intracranial granuloma is so common that neuroimaging is a must in children presenting with focal seizures who are otherwise asymptomatic.

Radioisotope studies

SPECT studies can be done at few centres in India. Ictal SPECT is more useful than inter ictal. There is hyper metabolism during seizures and hypometabolism during inter ictal period.

PET is not done in India.

Are other investigations indicated routinely?

Skull x-ray, CBC, ESR urine, LP, electrolytes, urea, creatinine, serum calciums etc, have a very low yield and conditions are clinically obvious and should be done only when indicated. Various tests for inborn errors of metabolism and work up for mentally retarded children with epilepsy should be done when indicated.

SIXTH STEP

Treatment

After deciding the type of epileptic seizures and syndromes and ruling out pseudoseizures, the patient should be started with the first line of drugs e.g. CMZ, VPA, PHT or PB. Monotherapy is the rule and the minimum dose of the drug that is indicated is initiated and slowly increased till seizures are controlled. If seizures are not controlled then the drug level is estimated and the dose is increased till the therapeutic level is reached or side effects occur. If there is no control of seizures, a second drug is added and first drug is slowly tapered off. When first line drugs fail to control seizures, add-on drugs like acetazolamide, clonazepam, clobazam, pyridoxine are tried or newer AEDs like VGB, LTG, OTBP etc are added, If epilepsy becomes intractable, ketogenic diet is considered. When everything fails, patients should be referred for pre-surgical evaluation and epilepsy surgery.