

Utility of “International League against Epilepsy (ILAE)” Classification in Patients with Epilepsy

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Abstract

Context: Epilepsy should always be defined by its type. This helps in appropriate management and also guides the physician in prognosis. Management duration of epilepsy depends upon syndrome. For each epilepsy syndrome, the management varies. This is challenging to have precise diagnosis. Hence ILAE gave classification of epilepsy. **Aims:** Application of International League Against Epilepsy (ILAE) classification in patients with epilepsy. **Settings and design:** Hospital based cross sectional study was carried out at Osmania General Hospital and Niloufer Hospital for women and children. **Methods and Material:** The study material consisted of consecutive patients (both inpatients and outpatients) with all seizure disorder. Details about each patient's birth and development, age at epilepsy onset, number of seizures, precipitating factors, and seizure description, family history, and neurologic findings were documented. Computerized tomography (CT) scans were performed on most patients and MRI scans in some cases. EEGs were recorded in most patients. **Statistical analysis:** Proportions were used to analyze the data. **Results:** In this study majority of the cases were children less than 10 years of age. Of the study population, 52.61% were classified as localization-related, 26.3% patients with generalized epilepsies and epileptic syndromes, 11.69% cases were Undetermined whether focal or generalized and 9.37% were Special syndromes (Situation - related). Symptomatic category of epilepsies was seen in majority of the localization related epilepsy cases. Ring enhancing lesion and cerebral calcified lesion on neuro imaging are common etiologic factors for epilepsy in our study. **Conclusion:** The 1989 ILAE classification is a good method of classification of epilepsies and related syndromes.

Keywords: Epilepsy; Classification; Patients; Syndrome.

Introduction

There is high incidence as well as prevalence of seizures due to epilepsy and epilepsy syndromes. It can affect anyone irrespective of age, sex or race. Physician come across cases of epilepsy in their day to day practice. Epilepsy is a group of syndromes. Each syndrome has different cause and manifests differently. All such entities are well defined now a days and hence not difficult to diagnose. Epilepsy should always be defined by its type. This helps in appropriate management and also guides the

physician in prognosis. Management duration of epilepsy depends upon syndrome. For each epilepsy syndrome, the management varies. This is challenging to have precise diagnosis. Hence ILAE gave classification of epilepsy. Epileptic disorder is defined as a long term condition of neurological origin where patient experiences repeated episodes of seizures of epileptic in nature [1].

Epilepsy can be defined as a condition with 2 or more episodes of seizures of epileptic in nature which is not provoked and where cause may not be found [2,3].

Even though a patient may have multiple episodes of seizures in 24 hours, it is taken as single event. Active epilepsy can be defined as a patient of epileptic nature in the last five years irrespective of whether the same patient is taking treatment for epilepsy or not [2,3].

“Epilepsy in remission with treatment,” can be defined as, “a prevalent case of epilepsy with no

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seizures for ≤ 5 years and receiving AED treatment at the time of ascertainment [2,3]."

"ILAE" defined epilepsy as brain disorder where there is tendency to seizures and there are not only neurological and cognitive consequences but also social and psychological. As per this definition at least one episode of seizure of epileptic nature should occur with evidence of seizure activity in the brain [4].

Aim of the present study was application of "International League Against Epilepsy (ILAE)" classification in patients with epilepsy as seen in Osmania General Hospital and Niloufer Hospital for women and children.

Materials and Methods

The study material consisted of consecutive patients (both inpatients and outpatients) with all seizure disorder seen between August 2010 and January 2013 at Osmania General Hospital and Niloufer Hospital for women and children.

Details about each patient's birth and development, age at epilepsy onset, number of seizures, precipitating factors, and seizure description, family history, and neurologic findings were documented. Computerized tomography (CT) scans were performed on most patients and MRI scans in some cases. EEGs were recorded in most patients.

Because we had no facility for special investigations such as continuous EEG monitoring or video EEG analysis, classification was based on a detailed history, personal observation of seizures in some cases, thorough clinical examination, and a 30-min scalp EEG recording.

CT scan and MRI evidence of abnormal focal lesions was recorded. A local spike or "sharp wave discharge" or "persistent focal slowing" in EEG was taken as focal epilepsy.

From this database, 821 consecutive entries were selected for this study. Epilepsy was classified as per "International Classification of Epilepsies and Epileptic Syndromes (ICEES)" proposed by the "ILAE 1989".

Patients were classified in category 1.2 when there was evidence of structural lesion on neuroimaging with clear prior history and having either generalized or isolated seizures.

Patients were classified as category 1.3 i.e. cryptogenic localization related epilepsy when patient gave history of recurrent seizures and there

was an evidence of focal onset on EEG but cause may not be found.

Patients were classified as category 2.3.1 i.e. "other symptomatic generalized epilepsies not defined" who were having seizure episodes more frequently with mental retardation with lack of defined syndromes.

Patients were classified as category 3.2 i.e. "undetermined epilepsies without unequivocal generalized or focal features" having normal EEG with clinical generalized seizures.

Results

Table 1 shows age and sex distribution of the study group. Of the 821 patients, 477 (58.09%) were males and 344 (41.9%) were females. There were 330 (40.19%) patients < 10 years of age. These included 119 (36.03%) children in the age group of < 2 years, 211 (63.93%) in the age group of 3-10 years.

Table 2 shows distribution of ILAE categories of epilepsies. Of the 821 patients, 432 (52.61%) were classified as localization-related, 216 (26.3%) patients with generalized epilepsies and epileptic syndromes, 96 (11.69%) cases were Undetermined whether focal or generalized and 77 (9.37%) were Special syndromes (Situation - related).

Table 3 shows localization related epilepsies and epileptic syndromes. Of the 821 patients, 432 (52.61%) were classified as localization-related (categories 1.1, 1.2 and 1.3), based on seizure semiology and/or findings at investigations. Idiopathic localization-related epileptic syndromes (category 1.1) formed 1.4% of the study population. Of the 12 patients, 10 had Benign epilepsy with centrotemporal spikes (BECTS), accounted for 0.6% of the study population, 2 patients had childhood epilepsy with occipital paroxysm. Benign epilepsy with centro-temporal spikes (BECT) was suspected based on electro clinical features, although not all had undergone sleep studies or neuroimaging. CT scan was performed on seven of them with no pathological findings.

Table 4 shows age and sex distribution of Symptomatic localization related epilepsies. Symptomatic localization related epilepsies (category 1.2) were found in 278 (33.86%) patients. Of these, 157 were males and 121 were females. In each patient so diagnosed, the risk factor was considered sufficient to provoke epilepsy. All patients had abnormal findings on neuro imaging.

Table 5 shows age and sex distribution of cryptogenic localization related epilepsies. Of the

Table 1: Age and sex distribution of the study group

Age groups (years)	No. of patients (821)		Total
	Male	Female	
0-10	199	131	330
11-20	123	96	219
21-30	57	61	118
31-40	43	30	73
41-50	33	16	49
51-60	17	7	24
61-70	4	2	6
> 70	1	1	2

Table 2: Distribution of ILAE categories of epilepsies

Category	No. of patients (821)	Total	Percentage
Localization related	240	432	52.6
Generalized	130	216	17.3
Undetermined: whether focal or generalized	54	96	11.7
Special syndromes	53	77	9.37

Table 3: Localization related epilepsies and epileptic syndromes

Localization related epilepsy category	No. of patients (432)		Total	%
	Male	Female		
Idiopathic (1.1)	8	4	12	2.7
Symptomatic (1.2)	157	121	278	64.3
Cryptogenic (1.3)	75	67	142	32.9

Table 4: Age and sex distribution of Symptomatic localization related epilepsies

Age group (years)	No. of patients (278)		Total
	Male	Female	
0-10	45	27	72
11-20	40	39	79
21-30	29	26	55
31-40	17	15	32
41-50	15	10	25
51-60	8	3	11
61-70	2	0	2
> 70	1	1	2

localization-related epilepsies, 142 (17.29%) patients were cryptogenic (category 1.3). Brain imaging was obtained in only 106 (74.64%) cases. Diagnoses of all other cases were based on seizure semiology, clinical data, and EEG findings. None of these patients had any antecedent event to suggest CNS injury.

Table 6 shows generalized epilepsies and epileptic syndromes: There were 216 (26.3%) patients with generalized epilepsies and epileptic syndromes: 69 (8.4%) cases of idiopathic generalized epilepsies (IGE) (category 2.1), 59 (7.18%) cases of cryptogenic or symptomatic generalized epilepsies (category 2.2), and 88 (10.17%) cases of symptomatic generalized epilepsies (category 2.3).

Table 7 shows age and sex distribution of symptomatic epilepsies (nonspecific etiology). Of the 88 cases of symptomatic epilepsies, 74 patients (45 were males and 29 were females) with static encephalopathy with mental retardation had severe generalized seizures, they could not be classified as West syndrome or Lennox-Gastaut syndrome on clinical and EEG findings, so were classified under the 2.3.1 category of "nonspecific etiology." The most frequent seizure type was generalized tonic clonic in 39 (52.7%) of cases. Generalized tonic seizures were seen in 8 (10.8%), mixed seizure type in 22 (29.7%), myoclonic seizures in four and absence seizure in one patient in this subcategory. Generalized epileptic discharges in EEG were established in 61%, background slowing in 28.6% of cases. Neuro-radiological investigations

were performed in 53 patients, showed abnormality in 27 patients.

Table 8 shows age and sex distribution of epilepsies and syndromes undetermined whether focal or generalized. In 96 (11.69%) cases (54 were males and 42 were females), clinical and EEG findings were not sufficient to classify the epilepsy as either focal or generalized; therefore, these patients were categorized as having epilepsies with undetermined focal or generalized seizures

(category 3.2). Seven patients had generalized seizures clinically with focus on EEG. Most of the patients were generalized seizures clinically and with normal EEG. CT scan was done in 82 cases, abnormality found in seven cases. There were no cases under the ILAE category 3.1.

Table 9 shows distribution as per etiology. Ring enhancing lesions on imaging accounted for 75 (20.89%) of cases. Cerebral calcified lesion with focus on EEG accounted for 67 (18.6%) of cases.

Table 5: Age and sex distribution of cryptogenic localization related epilepsies

Age group (years)	No. of patients (278) Cryptogenic localization related epilepsies		
	Male	Female	Total
0-10	23	19	42
11-20	29	20	49
21-30	7	16	23
31-40	4	7	11
41-50	6	1	7
51-60	5	3	8
61-70	1	1	2
> 70	0	0	0

Table 6: Generalized epilepsies and epileptic syndromes

Generalized epilepsy category	No. of patients (432)		Total No.	Total %
	Male	Female		
Idiopathic (2.1)	36	33	69	31.9
Symptomatic or cryptogenic (2.2)	39	20	59	27.3
Symptomatic (2.3)	55	33	88	40.7

Table 7: Age and sex distribution of symptomatic epilepsies (nonspecific etiology)

Age group (years)	No. of patients (278) Symptomatic epilepsies (non specific etiology)		
	Male	Female	Total
0-10	27	23	50
11-20	12	2	14
21-30	4	3	7
31-40	2	1	3
41-50	0	0	0
51-60	0	0	0
61-70	0	0	0
> 70	0	0	0

Table 8: Age and sex distribution of epilepsies and syndromes undetermined whether focal or generalized

Age group (years)	No. of patients (278) Epilepsies and syndromes undetermined whether focal or generalized		
	Male	Female	Total
0-10	15	11	26
11-20	19	11	30
21-30	6	9	15
31-40	9	6	15
41-50	1	5	6
51-60	4	0	4
61-70	0	0	0
> 70	0	0	0

Table 9: Distribution as per etiology

Etiology	No. of patients (432)		Total No.	Total %
	Male	Female		
Ring enhancing lesion	40	35	75	20.9
Birth asphyxia	48	25	73	20.3
Calcified lesion with focus on EEG	35	32	67	18.6
Cerebrovascular disease	25	7	32	8.9
Intracranial infection	14	16	30	8.3
Hippocampal sclerosis	5	9	14	3.9
Head injury	6	4	10	2.7
Intra cranial tumor	3	6	9	2.5
Others	34	15	49	13.6

Combined ring enhancing lesion and cerebral calcified lesion accounted for 142 (51%) of symptomatic localization-related epilepsies. Birth asphyxia was noted as an etiologic factor in 73 (20.3%) of patients. Of these 48 (65.7%) were males and 25 (34.2%) were females. Intracranial infections accounted for 30 (8.3%) of cases. Cerebral tumors accounted for nine (2.5%) of cases. Cerebrovascular disease was the antecedent event in 32 (8.9%) of cases, particularly in the patients aged >40 years (53%). Head injury was an antecedent event in 10 (2.7%) of cases.

Hippocampal sclerosis on MRI was seen in 14 (3.89%) of cases. Other structural lesions on neuro imaging was identified in 49 (13.6%) of cases.

Discussion

In this study 52.6% of the cases could be classified under localization related epilepsy. The dominance of localization related epilepsy is in keeping with the high proportion of partial seizures. Earlier studies from India also recorded a high frequency of partial epilepsies: 54.5% [5], and 80% [6]. Data in other developing countries and recent study from India were similar. ⁷The Okayana study [8] showed a rather different distribution of epilepsies (localization related epilepsy 22.4% and generalized 21.6%), presumably because of the large proportion of febrile convulsions (43.1%). Also, some hospital based studies of childhood epilepsies [9] and several studies including all ages [10] have revealed the clear predominance of localization-related cases.

The incidence of idiopathic partial syndrome (BECTS) in our series was 10 (1.2%) of 821 total syndromes. The low incidence in this study was similar to earlier observations from India [11] and also 1.3% reported by Eslava-Cobos and Narino [9],

but it was much lower than the 15.7% reported Heijbel et al. [12] as the study groups contain children. Those studies concerning all age groups, these syndromes have a relatively lower percentage.

We had 278 patients with symptomatic localization syndromes secondary to a variety of causes such as CNS infections, birth asphyxia and vascular pathology, who had simple partial, complex partial, or partial seizures secondarily generalized. CT scans were performed in all cases, showed various types of structural changes.

The incidence of symptomatic partial epileptic syndromes is obviously higher in developing countries because of the higher frequency of CNS infections such as tuberculosis, bacterial, and viral meningoencephalitis and perinatal insults [13]. Estimated symptomatic cases in various studies from developing countries have been variable: 39.4% in India [14], 17.5% in Libya [15]. This wide variation was probably the result of limited availability of neuroimaging.

Ring enhancing lesion, and cerebral calcified lesion are common etiologic factors for epilepsy in this part of the world; in our study, these two lesions combined accounted for 39.5% of cases in which an etiology was established by imaging and 51% of cases of symptomatic localization-related epilepsies.

In the present study, cryptogenic localization syndromes were diagnosed in 17.29% of cases. The frequency of this syndrome is seen similar in other studies: 27.4% [13], 22.8% [14]. Neuro imaging was done in 72% of cases (mainly CT scan compared to MRI). If such investigations done in all cases, would probably have increased the proportion of symptomatic cases, but many would probably still be classified under cryptogenic.

Among the patients with IGE, juvenile myoclonic epilepsy (JME) was reported in 2.4% of the total population. JME accounted for 4.9% [14], 0.8% [13].

Our data showed "other idiopathic generalized epilepsy not defined above" seems to be more common form of idiopathic generalized epilepsy, accounted for 5.1% of total study population and 61% of idiopathic generalized epilepsy. This is accounted for 19.9% [14], 15.5% [13] of epilepsy in various studies.

In the present study, West syndrome and Lennox-Gastaut syndrome were diagnosed in 7.18%. Other studies have reported variable frequency of this type which ranged from 0-6% [9,10,14]. This variability between the different studies is due to varying age groups studied. This incidence is higher in studies concerned with children only [9] but much lower in studies where the age group of patients was taken from childhood to adulthood [10,16].

The number of such patients with symptomatic generalized seizures was 74 (9%) in the present study. The proportion of this subcategory varies and is in the range of 1.0% to 8.8% in different surveys [14,16].

Epilepsies with specific syndromes (category 2.3.2) represented 1.7% of cases in the present study. In all other epidemiological studies this estimate is variable, ranging from 0.2% to 1.9% [9,14,16].

The category of either undetermined focal or generalized epilepsies included 11.69% of the cases. This particular epileptic syndrome is seen in 19.9% [14], 15.5% [13], and 23.3% [16] in other studies and requires careful electro-clinical study.

The category of special syndromes (situation – related seizures) represented in 9.37% of cases. In all other epidemiology studies this estimate is variable, ranging from 0.7% to 37.6% [9,13, 14,16].

Conclusions

Ring enhancing lesion and cerebral calcified lesion on neuro imaging are common etiologic factors for epilepsy in our study. The 1989 ILAE classification is a good method of classification of epilepsies and related syndromes.

Key messages

Early diagnosis of epileptic syndromes using tools like ILAE is important to benefit the patients.

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