www.jmscr.igmpublication.org
Impact Factor 5.84
Index Copernicus Value: 71.58
ISSN (e)-2347-176x ISSN (p) 2455-0450
crossref DOI: https://dx.doi.org/10.18535/jmscr/v5i11.07

IGM Publication
Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

Original Research Article

Study about Childhood Epilepsy & Epileptic Syndromes

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Abstract
Epilepsy is the most common neurological disorder of childhood. Most of the cases are idiopathic. Some are benign, needs no treatment and self-remits in adulthood.
Aims: *To study the clinical profile, aetiological factors, correlation between seizure semiology, EEG, brain imaging and prognosis of childhood epileptic syndromes.*
Material & Methods: *A total of 840 epilepsy patients seen over 1 year of study. Out of which 177 were between 1-14 years of age, but only 100 were included in the study according to inclusion criteria. Proper clinical history, surface EEG and brain imaging were performed.*
Results: *Childhood epilepsy accounted for 21% of all epilepsy patients. The incidence of idiopathic localisation related epilepsy was 6.2%, Idiopathic generalised epilepsy accounted for 18%.The commonest seizure type among localisation related epilepsies was partial seizures evolving to secondarily generalized.*
Conclusion: *Childhood epilepsies are a common neurological problems accounting for 21.1% of the total epilepsy patients. Benign epilepsy of childhood with centro temporal spike (BECTS) was the commonest idiopathic localization related epilepsy, whereas Juvenile myoclonic epilepsy (JME) was the commonest idiopathic generalized epilepsy. A family history of epilepsy was more commonly noted in JME. Therapeutic response were better observed in idiopathic epilepsies.*
Keywords: *Epilepsy, Seizure, Convulsion, Pediatric, EEG.*

Introduction

An epileptic seizure is a paroxysmal involuntary disturbance of brain function that may manifest as an impairment or loss of consciousness, abnormal motor activity, behavioral abnormalities, sensory disturbances, or autonomic dysfunction [1]. The prevalence of epilepsy in children varies between 2.5 to 11.5 averaging 4-6/1000 children[2]. It is higher in developing countries [3,4].70% of all epilepsies have their onset in the childhood. The incidence of epilepsy is greatest in first year of life, remains high up to 4 years of age, falls during childhood and declines slowly during adolescence and adult life and then rises sharply again after the age of 50 years [5].

Aims and objectives

1. To study the clinical profile of childhood epilepsy syndromes age 1-14 yrs.
2. To determine etiological factors in childhood epilepsy syndrome.
3. To determine clinicoradiological & clinicoectrographic correlation in childhood seizure disorder.

4. To study the correlation between International League Against Epilepsy (ILAE) 1989, seizure classification and semiological seizure classification and assess the usefulness with respect to ILAE epilepsysyndromic classification. To assess correlation between ILAE seizure type classification and semiological seizure classification.
5. To assess the short-term (three month) therapeutic response in the various childhood epilepsy syndromes.

Material and methods

The present study was undertaken to evaluate the clinical profile of childhood epilepsy syndromes in a tertiary care referral institute. Cases were evaluated in the epilepsy clinic between the age group of 1-14yrs for a period of 1 yr.

Inclusion criteria

(1) Children between 1-14 yrs of age
(2) Two or more unprovoked seizures 24 hrs apart
(3) Idiopathic and cryptogenic epilepsy
(4) Well-defined symptomatic epilepsy syndrome, irrespective of the underlying brain lesions.

Exclusion criteria

(1) Children < 1 yr or > 14 yrs of age
(2) Febrile seizures,
(3) Single or isolated seizures (i.e. one or more epileptic seizures occurring in 24 hrs period),
(4) Provoked seizures occurring in close temporal association with an acute systemic, metabolic or toxic insults or in association with an acute CNS disorder e.g. infection, Trauma, haemorrhage etc.
(5) Acquired symptomatic epilepsy like children with CNS granulomas, mass or any other obvious lesions like large infarct, cyst or gliosis etc.

Individual seizure types were classified according to the international league against epilepsy classification of epileptic seizures (ICES)[6] and the semiological seizure classification (SSC) proposed by Luders et.al.[7] in 1998.

Result and Discussion

Of the total 840 new epilepsy patient in the study period, 177(21.1%) were children (between 1 to14 yrs of age),of these 100 children with idiopathic, cryptogenic epilepsy and other rare epilepsy syndromes were included in the study. 61 were boy and 39 were girl. Cases were classified according to ILAE classification (Table-1). Age wise distribution of cases are shown in table-2. Localisation related epilepsy were 27.6%, generalized 27% and undetermined cases were 1.7%. History of febrile seizure was found in 6% of localization related epilepsy and 7% of generalized epilepsy. Family history was seen in 8.25% of localized and 6% in generalized epilepsy. Aura was noted in 32.3% of cryptogenic localization related epilepsy, whereas tonic-clonic seizure was noted in 87.7% of localization related epilepsy (Table-3). EEG abnormality was noted in 77.5% of localization related epilepsy and 75% of generalized epilepsy.

As seen in (Table-4) the prevalence of BECTS in different studies has varied considerably. This may be related to the different populations screened in the different studies. Results were compared with Eiji Oka.[8], Eriksson[9], Iceland[10], Murthy's[11] etc. Our results are similar to those by Eriksson[9], Shah[12] and Iceland[10] where age groups were almost similar. The higher percentage by Waaler[13] in his study can be explained by the age group screened which included only children between 6-12 years of age where this epilepsy syndrome is most common. The low prevalence in Murthy's[11] study is also explained on the different population group. The study by Eiji Oka et. al[8] was a retrospective study where past medical records were analyzed. Many cases in their study could not be classified due to inadequate clinical or EEG data hence the prevalence of this benign epilepsy may have been found to be lower than in the usual population. We have compared the characteristics of our patients with BECTS with those in the Iceland study[10]. The characteristics are similar as far the seizure pattern, sex distribution and age of onset are concerned.

Table 1: Distribution of Cases as Per ILAE Classification

Code	Syndrome type	No of cases	% of total
1	**Localization related epilepsy syndromes**	**49**	**27.6**
1.1.	Idiopathic	11	6.2
1/1.1	BECTS	10	5.6
1/1.2	CEOP	1	0.6
1/2.	Symptomatic	7	3.9
1/3.	Cryptogenic	31	17.5
2	**Generalised Epilepsy Syndromes**	**48**	**27**
2/1.	Idiopathic	32	18
2/1.1	Childhood Absence Epilepsy	4	2.3
2/1.2	Juvenile Absence Epilepsy (JAE)	2	1.1
2/1.3	Juvenile Myoclonic Epilepsy (JME)	9	5
2/1.4	Epilepsy with seizures on awakening	1	0.6
2/1.5	Others	16	9
2/2.	Idiopathic/ Symptomatic or both	10	5.6
2/2.1	West's Syndrome	3	1.7
2/2.2	Lennox Gastaut Synd. (LGS)	4	2.2
2/2.3	Epilepsy with myoclonic astatic sz.	3	1.7
2/3.	Other Symptomatic generalised epilepsy	6	3.4
3	**Epilepsies and syndromes undetermined whether focal or generalised**	**3**	**1.7**
3/1.	Landau Kleffner syndrome	1	0.6
3/2.	GTC in which clinical & EEG don't permit classification	2	1.1
Total cases		**100**	**out of 177**

It has also been proposed that BECTS and Landau Kleffner syndrome form the two ends of a syndromic spectrum with varying prognosis from excellent to poor[14,15,16]. Of the total cases of childhood epilepsy we had 17.5% cases, which were classified as cryptogenic localization related epilepsy.

Table 2: Age Wise Distribution of Cases

1	Localization related epilepsy synd.	**1 (2%)**	**7 (14.3%)**	**12 (24.5%)**	**29 (59.2%)**
1.1.	Idiopathic	0	4	3	4
1/2.	Symptomatic	1	0	1	5
1/3.	Cryptogenic	0	3	8	20
2	Generalised Epilepsy Syndromes	**4 (8.3%**	**6 (12.5%)**	**11 (22.9%)**	**27 (56.3%)**
2/1.	Idiopathic	0	3	7	22
2/2.	opathic/ Symptomatic or b	3	2	2	3
2/3.	Other Symptomatic generalised epilepsy	1	1	2	2
3	Epilepsies and synd. undetermined whether focal or generalised	**0**	**1**	**1**	**1**
	TOTAL	5	14	24	57

Table 3: Seizure Semiology in Childhoood Epileptic Syndromes

Seizure semiology	Localization related epilepsy	generalized epileptic syndromes	Undetermined whether focal or generalized	TOTAL
No of cases	**49**	**48**	**3**	**100**
Aura	**10**	**0**	**0**	**10**
Abdominal	1	0	0	1
Autonomic	1	0	0	1
Visual	2	0	0	2
Unclassifiable	6	0	0	6
Dialeptic seizures	**6**	**10**	**0**	**16**
Motor seizures	**55**	**49**	**4**	**108**
Myoclonic	0	15	0	15
Tonic	1	4	1	6
Clonic	1	0	0	1
Tonic clonic	43	26	2	71
Versive	2	0	1	3
Epileptic spasms	0	3	0	3
Automotor	8	1	0	9
Special Seizures	**0**	**9**	**0**	**9**
Atonic	0	3	0	3
Astatic	0	4	0	4
Hypomotor	0	2	0	2
Total	**71**	**68**	**4**	**143**

This prevalence is similar to other studies[9,10,12,,13,]. A higher prevalence was seen in Eiji Oka's[8] study but that may have been due to the difficulties in classifying cases retrospectively. In the generalized epilepsy group 32 cases (18%) had idiopathic generalized epilepsy (Table-5). The prevalence of IGE was similar to that found by Eriksson[9], Shah[12] and Genton[18]. Waaler[13] and Murthy[11] had a lower prevalence of IGE. This may be related to the different study groups in both the studies as compared to ours. The prevalence of IGE was higher in Eiji Oka's[8] study. This may have been due to the lower age group in this study where idiopathic epilepsy is more common. We found a comparatively higher percentage of patients with isolated myoclonic jerks in our study as compared to the other studies in patients of all ages. As our study was limited to children it is possible that other seizure types may manifest in the further course of the illness.JME was the most common idiopathic epilepsy syndrome in our study (Table-5 & 6).

We had a lower prevalence of West syndrome than other studies by Eriksson[9] and Shah[12] (Table-7). The difference in the age groups included may be responsible for this Shah[12] & Eriksson[9] both have included infant below first year of life. Ours being a study of childhood syndromes we excluded the infant population where West Syndrome is more common .As Waaler[13] included children above 6 yrs of age their prevalence was even lower. Comparing the characteristics of patient of LGS (Table-8) with those found in the Atlanta study[21] they are found to be similar.

Table 4: Prevalence of Idiopathic Localization Related Epilepsy in Children

Study	Year	N	Age gp (yr)	BECTS (%)	Epilepsy occipital paroxysms (%)	Total (%)
Eiji Oka[8]	1995	1871	<10	0.2	0	0.2
Eriksson[9]	1997	329	<15	8	0	8
Iceland[10]	1998		3-15.	9	NA	9
Murthy[11]	1998	2531	all ages	0.6	0.1	0.7
Shah[12]	1999	1742	<15	6.4	0.7	7.1
Waaler[13]	2000	198	6-12.	16.7	0	16.7
Present	2003	177	1-14.	5.6	0.6	6.2

Table 5: Prevalence of Idiopathic Generalized Epilepsies in Children

Study	Year	N	Age gp (yr)	CAE (%)	JAE (%)	JME (%)	EGMA (%)	Other IGE (%)	Total IGE (%)
Janz[17]	1969	6500		7.8	3	4.3	0		
Genton[18]	1991	1486		4.9	1.9	4	2		18
Eiji Oka[8]	1995	1871	<10	1.5	0.2	0	0	24.5	26.2
Eriksson[9]	1997	329	<15	6	2	2	1	10	21
Murthy[11]	1998	2531	all ages	0.4	0.1	4.9	0.03	1	6.4
Shah[12]	1999	1742	<15	0.8	0.18	0.24	0.08	13.1	14.4
Waaler[13]	2000	198	6-12.	6.1	1	5.1	0	0	12.2
Present	**2003**	**177**	**1-14.**	**2.3**	**1.1**	**5**	**0.6**	**9**	**18**

Table 6: Comparison of Characteristics of JME Cases

Study	Year	N	Age gp (yr)	Prevalence of JME (%)	Family history	Isolated myoclonic jerks	Epilepti-form activity
Mani, Rangam[19]	1990		all ages	5.9	44		
Murthy[11]	1998	2531	all ages	4.4	24	12	
AIIMS[20]	1999	3442	all ages	10	35	6	85
Present	**2003**	**177**	**1-14.**	**5**	**33**	**33**	**88.9**

Table 7: Prevalence of Idiopathic/Symptomatic Generalized Epilepsy in Children

udy	Year	N	Age gp (yr)	West synd. (%)	LGS (%)	Myoclonic astatic (%)	Total Idiopathic/symptomatic (%)
Eiji Oka[8]	1995	1871	<10	2.1	4.4	0	6.5
Eriksson[9]	1997	329	<15	8	2	4	14
Murthy[10]	1998	2531	all ages	0.9	1.6	0	2.5
Shah[19]	1999	1742	<15	8.5	0.5	0.1	9.1
Waaler[16]	2000	198	6-12.	0.5	4	1	11.6
Present	**2003**	**177**	**1-14.**	**1.7**	**2.2**	**1.7**	**5.6**

Table 8: Comparison Of Characteristics of Lennox Gastaut Syndrome Cases

	Atlanta study[21]	Present study
Total patients	23	4
Percentage of childhood epilepsy	4	2.25
Male:Female (%)	74/26	75/25
Cryptogenic %	44	25
Mental retardation (IQ < 70) %	91.3	100
Cerebral palsy %	52.3	50
Atonic Seizures %	40	75

Table 9: Sex wise Prevalence of Childhood Epilepsies

Study	Year	N	Male (%)	Female (%)
Eriksson[9]	1997	329	53	47
Estonia[24]	1999	560	56	44
Waaler[13]	2000	198	61	39
Present	**2003**	**100**	**61**	**39**

Table 10: Comparison of All ILAE Seizure types

Code	Seizure type	Waaler[13] (%)	Present (%)
I A	Simple partial	8.2	2.7
I B	Complex partial	18.8	3.4
I D	Sec. generalized	8.9	37.8
II A $_1$	Typical absence	5.9	4.3
II A $_2$	Atypical absence	15.4	4.3
II B	Myoclonic	9.5	11.2
II C	Clonic	0.9	0
II D	Tonic	7.5	6.8
II E	Tonic clonic	21.6	23.4
II F	Atonic	3.3	6.1
	Total	**461**	**115**

Table 11: Comparison of epileptiform activity on the EEG in different studies

Study	Year	N	Percentage with epileptiform activity (%)	EEG protocol
Blom[32]	1978		81	First record
			62	Follow up at 3 years
Tsuboi[33]	1986	378	58.5	First record
Waaler[13]	2000	198	85.8	Repeated on several occasions
Present	**2003**	**100**	**68**	**First record**

Atonic seizures were the most predominant seizure type. As with other studies[22,23] the short-term seizures control was not achieved in any of our cases predicting poor long term prognosis.

There was a preponderance of epilepsy in boys in the study (Table-9). This has been found consistency in other studies where similar results were found. Certain epileptic syndromes like LGS, Landau Kleffner syndrome and myoclonic astatic epilepsy (Doose's syndrome) have a significant male preponderance. Moreover in India due to bias against the girl child it is also possible that many girls with epilepsy may not be reaching the medical care facilities. Of the 6 children with absence epilepsy 2 children (33%) had febrile seizure preceding onset of absence seizures. A high incidence of febrile seizures preceding absence epilepsy has also been reported by others [25,26,27,28]. Considering the high percentage of cases with febrile seizures in our study the risk of patients with febrile seizures going on to develop epilepsy seems to be outwardly high. Risk is highest (2.4% at 25yrs of age) for those where onset of febrile seizures occurred below 12 months of age with multiple febrile seizures [30].

We classified seizure types according to the ILAE classification. Comparing our results with those of Waaler et. al.[13] (Table-10)we found similar comparable results in the generalized seizure groups. There was a marked discrepancy in the group with partial seizures.This is mostly due to the exclusion of acquired symptomatic localization related epilepsy patients in our study. Hence the results in the partial seizure group are not comparable. However in several instances classification based on semiology was difficult. We did not witness the seizure pattern in most cases and whenever we did observations were recorded by a single observer only. Semiology in most cases hence was obtained from an eyewitness history. Video recording of seizure which is found to have a high inter observer reliability [31] was not possible in our cases and remained one of the drawbacks in our study. In our study EEG was abnormal in 77% of the children. Epileptiform activity in form of spike/sharp waves was present in 68% of the patients. As seen in table-11, the percentage of epileptiform activity in the studies by Waaler[13], Blom[32] were higher than our study. However, Waaler et. al.[13] have repeated the EEG on several occasions responsible for a higher prevalence of epileptiform activity in their study. The overall short-term therapeutic response in the study showed at 77% had no seizures when followed up over a minimum three month period. Eriksson et. al.[9] have also found a similar result with remission rate of 70-80%.

Conclusions

Childhood epilepsies are a common neurological problems accounting for 21.1% of the total

epilepsy patients. BECTS is a common childhood epilepsy syndrome forming 5.6% of the total childhood epilepsy patients. Cryptogenic localization related epilepsies were found in 17.5% of our children.9.7% of these patients in our study had a positive family history. Idiopathic generalized epilepsies formed 18% of the cases in our study. Subtle absences or myoclonic jerks in previous years are often missed, and lack of video EEG may have been responsible for the higher prevalence of this group. JME was the commonest generalized idiopathic epilepsy syndrome in our study. Eliciting history of myoclonic jerks on direct questioning is very important for the diagnosis of JME, which generally has an excellent therapeutic response though requires long term therapy. LGS accounted for 2.2% of the childhood epilepsy syndromes. Generalized tonic clonic seizures and partial seizures with secondary generalization remained the most common seizures types in our study. Auras were seen in 32% of cryptogenic localization related epilepsies. EEG showed epileptiform activity in 68% of our patients with childhood epilepsy syndromes. Short term therapeutic response was found in 77% of the patient with epilepsy. The results were best in patients with Idiopathic epilepsies while the patients with idiopathic/ symptomatic localization related epilepsies fared the worst.

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