Research Article

Idiopathic epilepsies - A population based epidemiologic study

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ABSTRACT

Background: *There are only few epidemiological studies regarding epilepsy from this region. The classification of epilepsies has not been adequately stressed in these studies.*

Objective: *To assess the occurrence of idiopathic epilepsies in Punjab (A region situated in the Indo-Gagentic Plains), India.*

Methods: *A door to door, cross - sectional epidemiological survey covering an entire 103693 population residing in 74 contiguous villages situated outside the municipal limits of the District Headquarter town. Survey of all houses was done by general village level workers following which detailed case work up was done by postgraduate physicians. Final case identification was done by neurologists.*

Results: *Active epilepsy (n = 795) and inactive epilepsy (n = 128) comprised 74.78 % and 12.04 % of total number of seizure cases (n = 1063) and 86.13 % and 13.87 % of all epilepsy cases (n = 923) respectively. Non epileptic conditions comprised 11.74 % of total number of seizure cases (n = 1063). Single and febrile seizures were predominant among the non epileptic conditions being 6.2 % and 3.57 % of total number of seizure cases (n = 1063). Active epilepsy cases (n = 795) included electroclinical syndromes and constellations (n = 117, 14.72 %), symptomatic (n = 153, 19.24 %) and probably symptomatic cases (n = 513, 64.53 %) and cases with dual diagnosis (n = 12, 1.51 %). Idiopathic epilepsies (n = 112) consisted of idiopathic generalized epilepsies (n = 111) and benign childhood epilepsy (n = 1). Juvenile myoclonic epilepsy (n = 42) predominated idiopathic generalized epilepsy cases (n = 111).*

Conclusions: *Use of different terminology, definitions and criteria for diagnosis of various types of epilepsy precludes comparison between this study and other studies. This study showed significant variation in juvenile myoclonic epilepsy cases regarding sex distribution, diurnal variation and precipitating factors as compared to other studies. These differences need further confirmation.*

Key Words: *Epilepsy, population - based epidemiology, idiopathic epilepsy, active epilepsy, electroclinical syndromes, symptomatic epilepsy*

Introduction

It is estimated that >80% of individuals with epilepsy live in developing countries where the diseases remain largely untreated. They could live normal lives if properly treated, but the majority of patients do not receive any treatment at all. Global Burden of Diseases 2000 estimates the aggregate burden due to epilepsy at ~0.5% of the total disease burden making it clear that it results in a considerable burden. The suffering and the disability caused by the disease is physical and psychosocial, bringing a huge burden to people with epilepsy, their families and society at large. [1] Epilepsy may cause high levels of psychosocial difficulties for all family members, including stigmatization, stress,

psychiatric morbidity, marital problems, poor self esteem and restriction of social activities. The family environment may be an important intervening factor between the condition and the outcome for the family unit. People with epilepsy do not live in a vacuum: any negative consequences experienced are likely to extend to all family members. [2] Epidemiological surveys in several countries have shown high incidence and prevalence of epilepsy, with high associated disability and high rates of serious complications, including physical injury and premature mortality. Studies have also demonstrated that most people with epilepsy do not receive appropriate treatment. The "treatment gap" in less developed countries is an estimated 70% to 95%. Safe, effective and inexpensive medication exists that can control seizures in most people with epilepsy but a range of cultural, economic and other factors can prevent them from receiving the treatment they need. [3] Epilepsy is a common, largely untreated disorder in many resource poor regions. The basic medical principles of epilepsy care are the same in developed and developing regions, but there are unique challenges to providing good care in developing country settings and equally unique solutions may be considered. As with optimal epilepsy care anywhere, it is critical to consider the person with epilepsy in the context of their home, family, community and broader environment. [4]

This article is part of the series of articles based on findings of a door to door epidemiology survey covering an entire 103693 rural population of Punjab (India) situated in the geographic region called Indo-Gagentic Plains. Various reasons for undertaking this study have been elucidated in the introduction to the first article of this series titled 'Epilepsy in Punjab (India): A Population - Based Epidemiologic Study': Neuroepidemiology. (Under review) Briefly stated most of the studies from this region suffer from poor design or inadequate size of the population covered. Information about idiopathic epilepsy is important due to several reasons. Idiopathic epilepsy has its specific characteristics including precipitating factors and medications. These epilepsies, in particular, effect the younger population and hence counseling has an important place regarding career and child bearing issues. This is important for the planning of resources in the health sector.

In this article idiopathic epilepsies have been analyzed and compared with other studies.

Material and Methods

Detailed information regarding study area and the population, study phases, instruments used in various phases, specific definitions of various diseases and syndromes, the classification scheme framed for this survey, general definitions and statistical methods have been covered in 'Epilepsy in Punjab (India): A Population - Based Epidemiologic Study': Neuroepidemiology. (Under review)

Two neurologists, post graduates (Internal medicine and Psychiatry), medical graduates and paramedical staff (postgraduate in nursing and a pharmacist) each were involved in the survey. Anganwadi Workers (AWs) and the Private Health Workers (PHWs) worked at the village level. Training workshops, devoted to cardinal features of seizures, classification schemes and methods of epilepsy epidemiology, were held during the preparatory phase for the participating doctors and the paramedical staff by the neurologists. Two proformas in the

vernacular language and a booklet for detailed evaluation of epilepsy cases were framed and tested in a pilot survey after validation by two senior neurologists. Radio and TV broadcasts, wall posters, leaflets, news items and meetings of community leaders were used to sensitize the people regarding the survey. Entire study area was divided into four clusters with each comprising 15 - 20 villages. The medical graduates and the paramedical staff carried out coordination, data handling and record keeping. Postgraduates held the training session for field workers which included mimicking of seizure episodes and mock filling up of performas covering one cluster at one time. Training of field workers, screening (door to door survey by Aganwari Workers to fill the first performa and filling up of second performa by Private Health Workers for positive cases), evaluation (examination and filling up of the detailed epilepsy booklets by the postgraduate doctors) and confirmation (verification by the neurologists) phases were undertaken in each cluster. The screening phase was supervised by the medical graduates and the paramedical staff. Regular visits were made by the medical graduates and the paramedical staff to supervise and coordinate the screening phase. Patients were examined by the postgraduate doctors and the neurologists collected at a suitable place. A suitable classification scheme acknowledging the limitations of field situations was evolved. Various terms including epileptic seizure, epilepsy, active epilepsy, epilepsy in remission (inactive epilepsy), undetermined (active or inactive) epilepsy**,** unclassified epilepsy, neonatal seizure, prevalence (point ,period and life time), incidence (or incidence number), incidence rate and cumulative incidence have been defined in the first article mentioned above.

Entire information from the master chart was transferred to Microsoft Excel Data Sheet. Standardizations were carried out based on appropriate standard populations. Definitions and criteria for diagnosing only the relevant idiopathic syndromes are given here.

Definitions

Idiopathic epilepsy syndrome: A syndrome that is only epilepsy, with no underlying structural brain lesion or other neurologic signs or symptoms. These are presumed to be genetic and are usually age dependent. [5] In this study Juvenile Myoclonic Epilepsy, Juvenile Absence Epilepsy, Febrile Seizures Plus, Benign Childhood Epilepsy, Idiopathic Generalized Epilepsies (unspecified) and cases with only impairment of sensorium as defined below are included.

Criteria for idiopathic epilepsy syndromes

Generalized Epilepsy with Febrile seizures plus [6]:

1. Age of onset >1 month but seizures continue beyond the age range (> 5 Years) of febrile seizures
2. Precipitated by fever
3. Heterogeneous clinical phenotypes including myoclonic and absence seizures
4. Clinically normal
5. Neuroimaging normal or not done
6. Electroencephalogram (EEG) normal or shows nonspecific focal or generalized discharges or is not done

Juvenile myoclonic epilepsy [6]:

1. Age of onset = 14-15 (6 - 25) years
2. Myoclonus, +/- Absences, +/- Photosensitivity
3. +/- Aura
3. +/- Generalized seizures or other types of motor seizures

4. Neurologically normal
5. EEG showing primary generalized epilepsy or photosensitivity or non specific focal or generalized abnormalities or normal EEG or EEG not done
6. Neuroimaging normal or shows abnormalities which cannot be clinicaly correlated or not done

Juvenile absence epilepsy [6]:
1. Age of onset = 9-13(5 - 20) years
2. Absences
3. Generalized seizures or other motor seizures
4. Neurologically normal
5. EEG showing primary generalized epilepsy or non specific focal or generalized abnormalities or EEG not done
6. Neuroimaging normal or not done

Benign Childhood Epilepsy:
1. Age of onset 4 – 10 years
2. Afebrile seizures. Remission (Inactive epilepsy at present i.e.; seizure free period of 5 Years) within 1 -3 years from onset
3. EEG normal or non specific abnormality or suggestive of benign childhood epilepsy or not done
4. Neuroimaging normal or not done
Ideopathic Generalized Epilepsies

Unspecified:
1. Absences or myoclonus are present but classification into any specific idiopathic epilepsy syndrome, as defined for the purpose of this study, is not possible because of age of onset or presence of both absences and myoclonus simultaneously
2. +/- Motor seizures
3. Neurologically normal
4. EEG abnormality consistent with primary generalized epilepsy must be present when both absences and myoclonus are absent otherwise it may be normal or shows on specific abnormality or is not done
5. Neuroimaging normal or not done

Photosensitive Seizures:
1. Seizures
2. Neurologically normal
3. Either clinical or EEG or both indicate relation to photosensitivity
4. EEG shows photosensitivity or normal or non specific abnormality or not done
5. Neuroimaging normal or not done

Results

Number of cases of various epileptic syndromes of Idiopathic Generalized Epilepsies excluding the cases with dual diagnosis (N=111) expressed as percent of total number of cases of Idiopathic Generalized Epilepsies were Juvenile myoclonic epilepsy (n=42, 37.84%), Idiopathic generalized epilepsies unspecified(n=29, 26.12%), photo sensitive epilepsy (n=14, 12.61%), Febrile seizures plus (n=10, 9.01%), Impairment of sensorium only (n=9, 8.11 %) and Juvenile absence epilepsy (n=7, 6.31 %).

The age of patients of juvenile myoclonic epilepsy at the time of survey is given in Figure 1. Majority of cases were seen from the 2nd to 5th decade with peaks in the 2nd and the 4th decade of life.

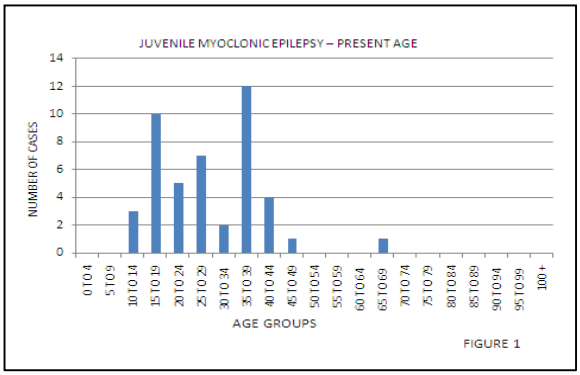

Fig.1 Present age of juvenile myoclonic epilepsy cases

Juvenile myoclonic epilepsy was more common in females (n = 30) than males (n = 15). The age of patients of Juvenile myoclonic epilepsy at the onset of epilepsy is given in Figure 2. Age of onset was in the first three decades of life with a peak in the 2nd decade.

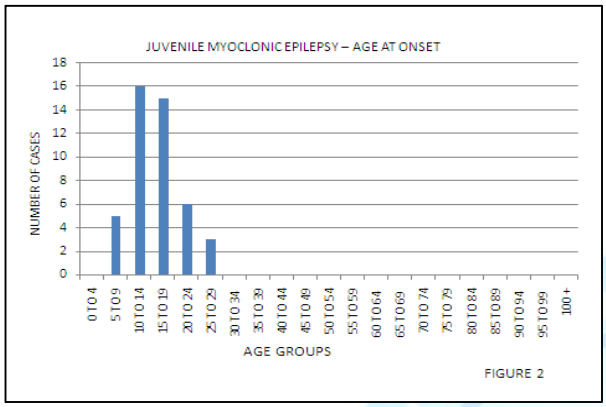

Fig. 2 Age of onset of juvenile myoclonic epilepsy cases

Number of cases of juvenile myoclonic epilepsy with diurnal variation is shown in Figure 3. Majority of patients did not show any diurnal variation. Only a few patients had seizures on awakening or in the morning hours.

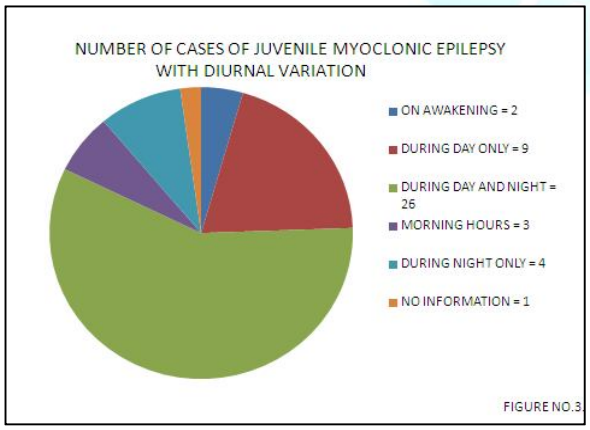

Fig. 3 Number of cases of juvenile myoclonic epilepsy with diurnal variation

Number of cases of juvenile myoclonic epilepsy with various precipitating factors is depicted in Figure 4. Majority of patients did not report any precipitating factors.

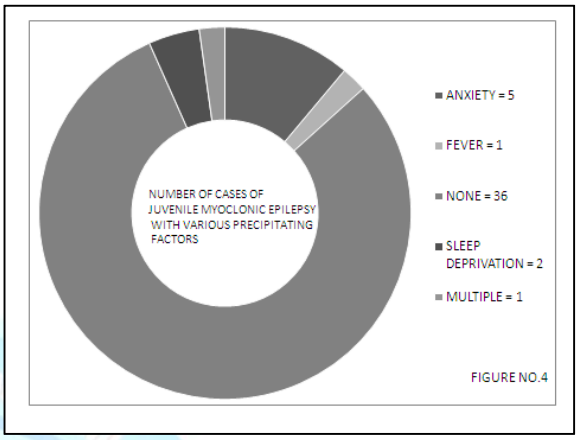

Fig.4 Number of cases of juvenile myoclonic epilepsy with various precipitating factors

Discussion

Idiopathic epilepsies

The electroclinical syndromes which are age dependent and are presumed to be genetic have been included in this category. The diagnosis of these conditions was made according to the criteria (vide supra) laid down for the purpose of this study. In the present study idiopathic generalized epilepsies and benign childhood epilepsy cases have been included in this category.

Idiopathic Generalized Epilepsies (n = 111) were found to be 13.96 % and benign childhood epilepsy (n = 1) to be 0.13 % of the active cases (n = 795) in the present study. Earlier studies did not give the criteria for syndromic diagnosis. Generally cases with myoclonus and absences are included in various studies. Idiopathic generalized epilepsies in various studies are given in Table No.1

Table: 1 Idiopathic generalised epilepsies in various studies

Study	Idiopathic generalised epilepsies (percent)	Total number of active epilepsy cases	standard error of difference in comparison to present study(1.96*SE)	Actual difference in comparison to present study	statistically significant difference present (actual difference > 2*SE)
***C* Present study**	13.96	795			
***C* Jammu & Kashmir, India** [7]	3.19	157	1.86 (3.66)	10.77	YES
***C* Pakistan** [8]	6.6	241	2.02 (3.95)	7.36	YES
***C* Turkey** [8]	6.1	81	2.93 (5.74)	7.86	YES
***A* US** [9]	13	383	2.11 (4.14)	0.96	NO
***C* Saudi Arabia** [10]	18.9	148	3.46 (6.78)	4.94	NO
A* = age adjusted to 1980 US population** ***C = crude					

The figures of primary generalized epilepsy in this study agree with the USA study (11), [9]are marginally lower than Saudi Arabian study (12)[10] and are higher than the study from Pakistan and Turkey. [8] In cases of juvenile myoclonic epilepsy this study showed significant variation regarding sex distribution, diurnal variation and precipitating factors as compared to other studies. [6] These differences need further confirmation.

Generalised Epilepsy

The term "Generalized Epilepsy" is not well defined in various studies. Criteria of the International League Against Epilepsy state that generalized seizures are those in which the first clinical changes indicate initial involvement of both hemispheres. Three studies[9,12,13] given below have applied the criteria of the International League Against Epilepsy (Commission, 1981) [11] to define this term. The study from US (11) US [9]has also applied the revised ILAE(1989) classification.[15] Another study[16] has applied the criteria given by ILAE Commission on Epidemiology and Prognosis[14] and revised ILAE (1989) classification. [15] The prevalence of generalized epilepsy is given in Table No.2

Table: 2 Generalized Epilepsy in various studies

STUDY	Generalized epilepsy cases (Percent)	Total number of active epilepsy cases	standard error of difference in comparison to present study(1.96*SE)	Actual difference in comparison to present study	Statistically significant difference present (actual difference > 1.96*se)
***Present study, Punjab , India**	34.46	795			
***USA[9]**	40	383	3.02 (5.91)	5.54	NO
***North Africa [12]**	97	141	2.21 (4.34)	62.54	YES
****East Africa[13]**	58	207	3.82 (7.49)	23.54	YES
***South Indian Study [16]**	58.8	1175	2.21 (4.34)	24.34	YES
****Bengal, India[17]**	55.1	38	8.24 (16.16)	20.64	YES
***Pakistan[18)**	55.6% (39.4 % definite + 16.2 % probably generalized cases)	241	3.62 (7.09)	21.14	YES
***=active epilepsy **= incidence cases only ***= active , inactive and some cases of febrile convulsions**					

In the present study, applying the above criteria, for purpose of comparison, both primary generalized epilepsies and generalized epilepsies have been put in this category. These account for 34.46 % of total active cases. This figure matches only with a study from USA [9] and is significantly lower than all other studies given above. Various studies [12,17,18] suggest that the high proportion of generalized convulsive epilepsy may be explained by the misclassification of partial seizures secondarily generalized as generalized seizures. Most studies that show a preponderance of partial over generalized seizures involve urban, semi urban, or hospital populations, not rural populations.[13] In this context it may be

mentioned that present study area lies just outside the municipal limits of the District Headquarter town hence resembles semi urban area. The study from USA [9] also involves urban area and population in a developed country may be more sensitized to focal features as compared to illiterate rural population of a developing country.

Use of different terminology, definitions and criteria for diagnosis of various types of epilepsy precludes comparison between this study and other studies.

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