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Epileptic spasms at Muhimbili National Hospital Tanzania, a retrospective study

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Abstract

Background: Epileptic spasms (ES) is an epileptic encephalopathy occurring during infancy and early childhood. Early recognition and management are important to prevent severe neurological impairment. This study aimed at describing the clinical presentation, management, and outcome of patients with Epileptic Spasms attending Muhimbili National Hospital (MNH) in Dar Es Salaam, Tanzania **Methods:** A retrospective cross-sectional study of all patients diagnosed with epileptic spasms was conducted at MNH from July 2016 to January 2021. **Results:** A total of 73 patients diagnosed with epileptic spasms were retrieved with a male to female ratio of 3:2. In this study, 37 (50.7%) patients had a documented history of perinatal insult. The median age of onset of spasms was five months (IQR 1-12 months). Thirty (88%) out of 34 patients whose electroencephalography (EEG) findings were retrieved had abnormal EEG findings showing generalized epileptiform discharges in 11(32.4%), generalized slowing in 5(14.7%), hypsarrhythmia in 7(20.6%), focal epileptiform discharges in 3 (8.8%), and suppression burst pattern in 4(11.8%). Fifty- six (77%) received prednisolone, with a median time of spasms cessation of 1 month with a range of 10 days to 3 months in about 80% of them. **Conclusion:** The median age of onset of epileptic spasms at MNH is five months, with the most common cause being a perinatal insult in more than 50% of the patients. High dose prednisolone showed a good response in patients with epileptic spasms at MNH.

Keywords: Epileptic spasms, hypsarrhythmia, prednisolone.

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Introduction

Epileptic spasms (ES), formerly called infantile spasms, is a sudden flexion, extension, or mixed extension–flexion of predominantly proximal and truncal muscles lasting about 2-3 seconds, that is usually more sustained than a myoclonic event but not as sustained as a tonic seizure. Other manifestations which may occur during an episode of epileptic spasms include Grimacing, head nodding, and subtle eye movements. Epileptic spasms frequently occur in clusters predominantly upon awakening. Epileptic spasms commonly occur during infancy. However, these events have been reported to occur in children who are older than one year [1]. It is one of the commonest epileptic encephalopathies causing developmental regression and intellectual disability. The incidence of epileptic spasms has been estimated to range from 2–5/10,000 newborns globally. In most cases, ES is due to perinatal hypoxia, with some cases being a result of different post-natal factors. The most common etiologies of ES reported are hypoxic-ischemic encephalopathy, chromosomal abnormalities, brain malformations, stroke, tuberous sclerosis complex, and periventricular leukomalacia or hemorrhage [2, 3, 4]. Poor antenatal, natal, and post-natal care stands out as the main underlying problem in developing countries.

This study was conducted to describe the clinical presentation, etiology, management, and outcome of patients with Epileptic Spasms attending Muhimbili National Hospital in Dar Es Salaam, Tanzania.

Methods

A retrospective cross-sectional study of all patients diagnosed with epileptic spasms was conducted at MNH. Paper based case notes and an electronic database of all patients with neurological conditions under the age of five years for both inpatients and outpatients from July 2016 to January 2021 were retrieved and thoroughly reviewed. Patients who met the diagnostic criteria for ES as stipulated by the International League Against Epilepsies (ILAE) 2017 edition were recruited into the study [1]. A structured questionnaire specifically designed for this study was used to collect data from the paper-based case notes and electronic database of the hospital. The data collected included social demographic characteristics, clinical presentations, electroencephalography (EEG) findings, Magnetic Resonance Imaging (MRI) features, medications used, and the outcome. The outcome assessed in this study was the duration of cessation of the spasms.

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Results

Paper based case notes and an electronic database for 2740 patients were reviewed, and 73 met the criteria for ES. Most of the patients were males, constituting about 60% of the studied patients. More than 80% (60 patients) included in the study were below three years of age, with the rest being 4 to 5 years old.

In this study, most patients with epileptic spasms had a history of a neurological insult that may have occurred during the prenatal, natal, or post-natal period. Among 37 patients whose data on etiology were retrieved, 22 (59.5%) had birth asphyxia, 2(5.4%) had neonatal hyperbilirubinemia, and 1(2.7%) had a history of preeclampsia. Moreover, intrauterine infections, congenital brain formation, and prematurity occurred in 4(10.8%) each.

The genetic component of epileptic spasms could not be elucidated in this study as it is not available in our setting. The median age of onset of patients with Epileptic spasms was five months (IQR 1-12 months). The nature of spasms demonstrated in a large number of patients with epileptic spasms was flexor type 68 (93%), with the remainder having both flexor and extensor type (mixed type). Most patients in this study had a variable documented number of frequencies of spasms per day, with most of them, about 20 (30%), having 2 to 5 clusters per day and 3 (4%) having more than 10 clusters per day. Thirty (88%) out of 34 patients whose EEG reports were retrieved had abnormal EEG findings showing hypsarrhythmia 7(20.6%), generalized epileptiform discharges 11(32.4%), generalized slowing 5(14.7%), focal epileptiform discharges 3 (8.8%) and suppression burst pattern 4(11.8%). Twelve (80%) out of 15 patients whose magnetic resonance imaging (MRI) results were found had documented abnormal MRI findings, with most of them showing features suggestive of hypoxic-ischemic encephalopathy (HIE), old subdural hematoma, megalencephaly, and bilateral hippocampal atrophy, severe brain atrophy, multicystic encephalomalacia, congenital hydrocephalus, and dandy walker malformations. MRI costs stand out to be the limiting factor as many parents cannot afford them.

The majority of the patients, 65 (89%) in this study, went through a course of different medications singly or in combination before the symptoms subsided. Before the diagnosis of epileptic spasms was made, many patients were receiving the following medications: sodium valproate 61(84%) and phenobarbitone 18 (25%). Other medications given were carbamazepine, clonazepam and Baclofen. After the diagnosis of epileptic spasms was made, 56 (77%) patients were documented to receive prednisolone, and 4 (6%) received Vigabatrin.

Thirty-three (80%) out of 41 documented patients treated for epileptic spasms with prednisolone, their symptoms subsided within the first month of treatment with an interquartile range from 10 days to 3 months.

Table 1. Etiology, EEG, MRI findings, and medications received by patients with epileptic spasms.

Etiology of ES (N=37)	**N (%)**
Birth asphyxia	22 (59.5%)
Intrauterine infections	4(10.8%)
Prematurity	4(10.8%)
Preeclampsia	1(2.7%)
Hyperbilirubinemia	2(5.4%)
Brain malformations	4 (10.8%)
EEG Findings (N=34)	
Generalized epileptiform discharges	11 (32.4%)
Generalized slowing	5 (14.7%)
Hypsarrhythmia	7 (20.6%)
Abnormal focal epileptiform discharges	3 (8.8%)
Suppression burst	4 (11.8%)
Normal finding	4 (11.8%)
MRI Findings (N=15)	
Abnormal	12 (80.0%)
Normal	
Medications received before the diagnosis (N= 73)	
Sodium Valproate	61 (83.6%)
Phenobarbitone	18 (24.7%)

Discussion

The study aimed at describing the clinical presentation, etiology, management, and outcome of patients with Epileptic Spasms seen at Muhimbili National Hospital. The median age at onset was five months, and most patients showed a good response to high-dose prednisolone.

The study showed most of the affected children had their onset of epileptic spasms before the first year of life, with a peak incidence in almost 60% being between 2 to 6 months and affecting slightly more males than females. Similar findings were reported in a multicenter randomized, double-blind clinical trial to assess the efficacy of Vigabatrin, and ACTH conducted in the United States by Pellock et al., where the peak onset was 3 to 7 months which occurred in 50-70% of the patients [5]. The pathogenesis of epileptic spasms is postulated to result from disruption of neuronal network function as a result of a defect either at the cellular, receptor, or molecular level, leading to abnormal interaction between cortical and subcortical structures [6, 7, 8]. Significant and rapid brain development occurs during the first year of life involving myelination, synaptogenesis, and pruning; subsequently, if any of these processes is severely affected, epileptic spasms and/or other seizure types are likely to arise during this period.

Patients in this study demonstrated two clinical types of spasms, with the majority of patients, 68 (93%), showing flexor type, and 7% had mixed flexor-extensor type involving the muscles of the neck, trunk, and extremities. This is different from

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the study done by Kellaway et al. in Texas, U.S.A using time-synchronized video EEG recording in 24 infants who showed most infants tend to have more than one type of spasms as 42% had a mixed type, 34% flexor, and 23% extensor type [9]. The differences in results are attributed to the variations in study designs where the retrospective method carried out in this study could have missed some important detailed semiology of the spasms as parents/caretakers are usually frightened by the event, and some forget the details of the semiology of the events which were easily picked up in the prospective study with video recording done in Texas, U.S.A.

Perinatal insults accounted for 50% of the etiologies of ES in this study compared to 10%, which were found in a multicenter study done in Canada, France, and the United States of America. Inadequate perinatal care in our setting, as compared to developed countries, leads to an increased number of children who experience perinatal insults leading to a higher proportional contribution to ES. Most patients with ES in this study had Cerebral Palsy. The risk factors for cerebral palsy found in this study were birth asphyxia, intrauterine infections, neonatal hyperbilirubinemia, preeclampsia, and prolonged labor, which are all preventable with improved antenatal care. Other etiologies found in that multicenter study included genetic (8%), Tuberous Sclerosis (7%), stroke (8%), cerebral malformations including porencephaly (8%), and periventricular leukomalacia (5%) [2].

In this study, 30 (88%) out of 34 patients whose EEG reports were retrieved had abnormal EEG findings. The typical EEG finding in ES of hypsarrhythmia was only found in 7(20.6%) compared to 75%, which has been reported in other studies [10]. This could be attributed to late diagnosis in most of our patients, as well as EEG being done late during the course of illness after receiving several medications, which could have altered the EEG patterns. Poor awareness of the condition among health care workers and family financial constraints are key in limiting early access to EEG. Other EEG findings reported in this study were generalized epileptiform discharges 11 (32.4%), generalized slowing 5(14.7%), focal epileptiform discharges 3 (8.8%), or suppression burst 4(11.8%).

In this study, 12 (80%) out of 15 patients whose MRI results were found had documented abnormal features, with most of them showing features suggestive of hypoxic-ischemic encephalopathy (HIE) due to perinatal insults. The abnormal MRI findings included old subdural hematoma, megalencephaly, bilateral hippocampal atrophy, severe brain atrophy, multicystic encephalomalacia, congenital hydrocephalus, and dandy walker malformations. In a retrospective study done by Khatami et al. in Canada on Brain MRI findings in patients with ES where a total of 26 patients were included, 19 (73%) had abnormal MRI findings, which were features of HIE sequalae, tuberous sclerosis (including one with megalencephaly), Lissencephaly, infarcts secondary to meningitis and frontal heterotopia. This shows structural causes of ES are quite similar between Africa and the west [11].

The majority of the patients, 65 (89%) in this study, went through a course of different medications before the spasms subsided. Before the diagnosis of epileptic spasms was made, many patients received sodium valproate 61(84%) and phenobarbitone 18 (25%). Other medications given were carbamazepine, clonazepam, and baclofen. After the diagnosis of epileptic spasms was made, 56 (77%) patients received prednisolone. Thirty-three (80%) out of 41 patients who received prednisolone, spasms subsided within the first month of treatment with an interquartile range of 10 days to 3 months, similar to a randomized clinical trial by Lux et al. in which 70% responded with a median duration of 14 days after being given high dose prednisolone for 14 days. This study emphasizes that high-dose prednisolone is effective in the cessation of spasms even in resource-limited settings where the diagnosis is often made late [12]. The prednisolone dose given was 4mg/kg/day during the first two weeks in divided dosages, then tapered down for the next four weeks.

Conclusion

The median age of onset of epileptic spasms at MNH is five months. The most common cause of epileptic spasms in children at MNH is perinatal insults occurring in more than 50%. Despite late presentation, most patients showed a good response to high-dose prednisolone.

Abbreviations

EEG	Electroencephalogram
ES	Epileptic spasm
HIE	Hypoxic ischemic encephalopathy
IQR	Inter quartile range
MRI	Magnetic resonance imaging
MUHAS	Muhimbili University of Health and Allied Sciences
MNH	Muhimbili National Hospital
SPSS	Statistical package for the social sciences
ILAE	International League Against Epilepsy

Ethical issues

Permission for data collection was obtained from the administration of Muhimbili National Hospital (MNH) and MUHAS with letter Ref.No.DA.282/298/01.C, and the data were kept as confidential information.

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Competing interests

The authors declare that they have no competing interests.

Author contributions

JM and EK assisted with study design, identifying patients for study inclusion, data collection, and results analysis. JM drafted the manuscript. EK contributed to the manuscript content and revised the final manuscript.

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