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Infantile Epileptic Spasms Syndrome: Clinical profile and outcomes at a Tertiary Hospital in Kenya

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ABSTRACT

Background: Infantile epileptic spasms syndrome (IESS) is characterised by onset of epileptic spasms between 3 and 12 months, with or without typical hypsarrhythmia, and developmental stagnation or regression. There are limited data on the clinical profile and treatment outcomes of IESS in sub-Saharan Africa. **Objective:** To describe the clinical profile and treatment outcomes of infants diagnosed with and managed for IESS at Aga Khan University Hospital, Nairobi. **Methods:** Records of children aged 3–24 months diagnosed with IESS between January 2012 and December 2021 were identified from the hospital information system using the terms “infantile spasm”, “epileptic spasms”, and “West syndrome”. Medical history, investigations, treatment, and outcomes were extracted using a structured tool. Outcomes at six weeks were categorised as complete response (spasm-free), partial response (any reduction in spasms), or no response. **Results:** Forty-two children with IESS were identified. The median age at onset was 5 months (IQR 3.5–8.5). The male-to-female ratio was 1:2.8. Age at diagnosis and treatment initiation was 6.5 months (IQR 4.0–10.5). Flexor spasms were most frequent (24/42; 57.1%). The commonest aetiology was structural (26/42; 61.9%), with unknown causes in 14/42 (33.3%). Most patients had received non–first-line anti-seizure medicines before referral. At the study site, patients received vigabatrin plus prednisolone irrespective of aetiology. At six weeks, 24/42 (57.1%) were spasm-free, 17/42 (40.5%) had a partial response, and 1/42 (2.4%) was lost to follow-up. By two years of age, 3/37 (8.1%) assessed children had normal development, while 34/37 (91.9%) had delays in multiple domains. Ongoing seizures were reported in 32/42 (76%) patients, with a mean duration of 7.04 weeks. **Conclusion:** Despite more than half of patients achieving spasm cessation at six weeks, few children attained typical developmental milestones at follow-up, likely reflecting delays to diagnosis and treatment. Standardised referral pathways are urgently needed to enable timely management of IESS and improve outcomes. A prospective, multicentre study in the region is warranted.

Keywords: Epileptic spasms, hypsarrhythmia, vigabatrin, infantile epileptic spasm syndrome

INTRODUCTION

Infantile epileptic spasm syndrome (IESS) is a severe early-onset epileptic disorder presenting between 1 and 24 months of age. It is characterized by epileptic spasms, accompanied by one of the following: an EEG pattern of hypsarrhythmia and/or developmental delay (1–4). The hallmark seizure type is the epileptic spasm, most often flexor, though extensor and mixed types occur. Spasms typically cluster, may be symmetric or asymmetric, and are frequently

accompanied by developmental arrest or regression (2,6,7).

The International League Against Epilepsy (ILAE) proposed the term IESS to encompass both West Syndrome as well as infants presenting with epileptic spasms who do not fulfill all the criteria for West Syndrome (4). West Syndrome classically referred to the triad of epileptic spasms, hypsarrhythmia, and developmental regression.

IESS occurs worldwide, affecting infants across all ethnic groups with no sex predilection (3). Etiologies are diverse, including hypoxic–ischemic encephalopathy, congenital brain malformations, and tuberous sclerosis complex, though a substantial proportion remains of unknown cause (8). Neuroimaging is abnormal in up to two-thirds of patients (9), while genetic and metabolic investigations are recommended when structural causes are not identified (8). First-line therapies include hormonal treatment (ACTH or prednisolone) and vigabatrin (10–12). Evidence from clinical trials remains mixed, with vigabatrin favored in tuberous sclerosis (10), International Collaborative Infantile Spasms Study (ICISS) reporting benefit of combined vigabatrin and hormonal therapy (14), and a Sri Lankan trial finding prednisolone superior, likely reflecting different etiological profiles (15).

Although IESS has been well described in high-income settings, there is limited data from Sub-Saharan Africa. A South African study reported IESS in 0.4% of children admitted with seizures (5), but information on clinical features, etiology, and treatment outcomes in the region remains sparse. This study, therefore, aimed to describe the clinical profile and treatment outcomes of infants with IESS managed at a tertiary hospital in Sub-Saharan Africa.

METHODS

Study design: This was a retrospective observational study conducted at Aga Khan University Hospital, Nairobi (AKUHN).

Study Setting: This study involved a review of clinical records of patients on follow-up at the neurology department in Aga Khan University Hospital, Nairobi (AKUHN). AKUHN is a private not-for-profit tertiary teaching and referral Hospital in Nairobi, Kenya. A multidisciplinary team led by two pediatric neurologists provides specialized neurological care.

Inclusion Criteria: We identified 50 children aged 3–24 months with a diagnosis of infantile epileptic spasm syndrome (IESS), based on a history of epileptic spasms, accompanied by an EEG pattern of hypsarrhythmia and/or developmental delay, and managed between January 2012 and December 2021.

Exclusion Criteria: We excluded 8 patients due to incomplete medical records.

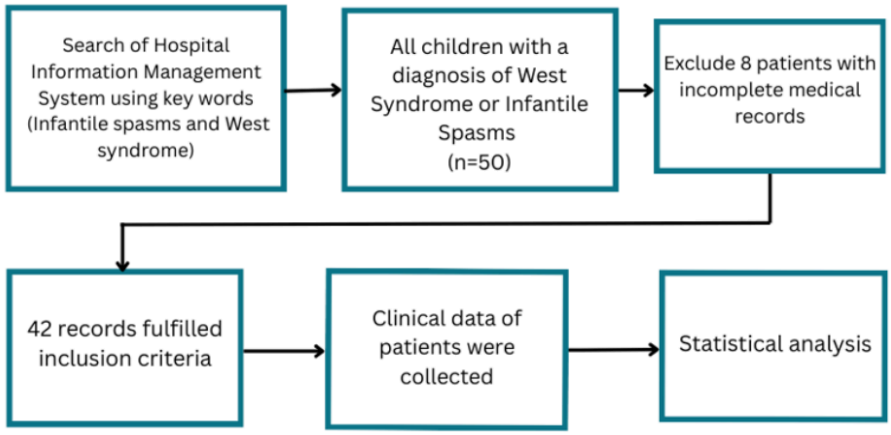
Study Procedures

After obtaining approval from the Aga Khan University- Kenya; Institutional Scientific and Ethics Review Committee (Ref:2022/ISERC-82), we took a census approach and identified 50 cases from the hospital information management system using the search terms “infantile spasm” “epileptic spasms”, and “West Syndrome”. Eight patients were excluded due to incomplete medical records. The study flow chart is shown in Figure 1. Demographic and clinical data were extracted from the patients’ files and electronic medical records using a structured data collection tool. Etiology was classified as known or unknown based on history, examination, and available relevant investigations such as brain MRI and metabolic work-up. Outcome was assessed at six weeks after the initiation of treatment, and categorized as *complete response* (no spasms), *partial response* (reduced frequency), *no response* (unchanged), or *loss to follow-up* (no documentation at six weeks). The children were reviewed for developmental outcome at 2 years of age or at the last encounter if done before the age of 2 years using parental milestones reports and clinical evaluation by the attending neurologist.

Statistical analysis

De-identified data were verified, completed, and entered into an Excel data form by a single reviewer. This was followed by validation checks by an independent data clerk. The data were analyzed using Statistical Package for the Social Sciences (SPSS) version 22.0, with descriptive statistics. The background, demographic, and clinical profile were summarized and presented in tables. Continuous variables like age were expressed as mean. The proportion of children with resolution of epileptic spasms at six weeks as well as gender were calculated as a percentage of the total number of children included in the study.

Figure 1: Study Flow Chart



RESULTS

Age at Onset and Diagnosis

Age of onset of IESS was 4.7 months (IQR 3.3-7.3), with diagnosis and treatment initiation delayed until 6.5 months (IQR 4-10.5). The demographic data is shown in Table 1.

Clinical and EEG findings

The birth and perinatal course of children diagnosed with IESS is shown in Table 2. Most of the patients (n=23, 57.1%) had comorbidities. Visual impairment was the most common comorbidity in 13(31.0%) cases, followed by recurrent chest infections in 6(14.3%). Less frequent presentations included combined hearing and visual impairment 2(4.8%) and combined visual impairment with recurrent chest infections 2(4.8%). Electroencephalogram (EEG) findings showed modified hypsarrhythmia in 21(50.0%) patients. With respect to spasm type, flexor spasms were the most frequent 24(57.1%), followed by extensor 3(7.1%) and mixed spasms 2(4.8%), while 13 (31.0%) were unclassified.

Etiology of IESS

The most prevalent aetiology was structural abnormalities, in 26(61.9%) patients. For 14(33.3%) children, no etiology was established (Table 3).

Initial and Definitive treatments

Before referral to our institution, most patients had been treated with non-first-line antiseizure medications at other facilities. The most common regimen was phenobarbitone monotherapy 7(17.0%). Notably, 5(12.2%) of the children had not received any antiseizure medication prior to referral.

All study participants received vigabatrin in combination with prednisone. Vigabatrin was issued at a mean dose of 120.5 ± 20.7 mg/kg/day for a mean duration of 6.0 ± 1.64 months. Prednisone was administered orally once daily at a mean dose of 6.06 ± 1.56 mg/kg/day for a mean duration of 4.7 ± 1.30 weeks.

Short-term Treatment Outcomes and Adverse Reactions

At six weeks, 24(57.1%) children were spasm-free. A further 17(40.5%) children showed a partial response, defined as a reduction in spasm frequency. Only 1(2.4%) patient was lost to follow-up after the first review.

Prednisone was well tolerated in most patients, with 39 children (83.3%) experiencing no adverse effects. Among the 6 participants with prednisolone-associated side effects, 4 presented with cushingoid facies only, 1 had cushingoid facies with hirsutism, while 1 was noted to have hirsutism in isolation. Parents of one child reported somnolence following initiation of vigabatrin at 94mg/kg/day.

Long-term developmental and seizure outcomes

At two years of age, or at the last available follow-up, most children demonstrated developmental impairment (Table 4). Delays in gross motor, speech, and social skills were observed in 16(43.2%) of patients, while only 3(8.1%) achieved normal development (Table 4).

Persistence of epileptic spasms beyond six weeks of treatment occurred in 8(21.6%) of the children. During follow-up 10(23.8%) remained seizure-free. Among those with ongoing seizures, the majority 27(73%), did not have sufficient information for a clear classification and were therefore unknown (whether focal or generalized).

Follow-up EEG Findings

Repeat EEG at weeks was performed for most patients. Ten (32.3%) had a follow-up EEG carried out between one and ten weeks after the initial EEG (Table 5). Nine of the patients had no repeat EEG. Notably, 5(11.9%) of the patients had a normal repeat EEG, which correlated with clinical spasm resolution. Others showed interictal or ictal focal and generalized abnormalities (Table 6). Four EEG tracings were not available for review.

Reported Deaths

There were 2(4.8%) deaths, both unrelated to epilepsy. One child died at 7 months of age at home with respiratory symptoms prior to his demise. The second child died in a hospital at 14 months of age from respiratory failure while on treatment for presumed pneumonia. Neither child was on steroids at the time of their illness.

Table 1: Demographic Data

Parameter	Percentage(n=42)
Gender	
Male	31(73.8)
Female	11(26.2)
Dysmorphic Features	
None	35(83.3)
Narrow nasal bridge, epicanthal folds, and overriding second toe on both feet	1(2.4)
Non-specific features	2(4.8)
Single palmar crease	1(2.4)
Low set ears	1(2.4)
Flat nasal bridge, epicanthic folds	1(2.4)
Micropthalmia, flat nasal bridge, high arched palate, corneal opacities	1(2.4)
Median Age at Diagnosis (Months)	6.5 (IQR 4-10.5)
Median Age at Treatment Initiation (Months)	6.5 (IQR 4-10)
EEG Initial findings	
Modified hypsarrhythmia	21 (50%)
Hypsarrhythmia	11 (26.2%)
Other	5 (11.9%)
Missing data	5 (11.9%)

Table 2: Birth and Perinatal Course

Mode of Delivery	Percentage (n=41)
Spontaneous vaginal delivery (SVD)	19 (45.2%)
Emergency caesarean section	13(31.0%)
Elective cesarean section	8 (19.0%)
Assisted vaginal delivery	1 (2.4%)

Perinatal Course	Percentage (n=41)
Uneventful	16(39.02)
Neonatal seizures	5(12.20%)
Jaundice requiring phototherapy/exchange transfusion	1 (2.44%)
Delayed cry, Neonatal seizures	5 (12.20%)
Premature rupture of membranes	2 (4.88%)
Delayed cry	6(14.63%)
Low birth weight	1 (2.44%)
Neonatal sepsis	1 (2.44%)
Meconium-stained liquor, Neonatal seizures	1 (2.44%)
Newborn unit admission	1 (2.44%)
Low birth weight, polyhydramnios	1 (2.44%)
Neonatal seizures, Neonatal sepsis	1 (2.44%)
	41 (100)

Table 3: Aetiology of IESS

Aetiology	*Count*	*Percentage*
Unknown	14	33.3
Structural	26	61.9
Infectious	2	4.8

Table 4: Developmental Outcomes

Developmental Outcome	*Count*	*Percentage*
Delayed gross motor skills, speech, and social skills	16	43.2
Delayed gross motor skills	11	29.7
Normal	3	8.1
Delayed speech	4	10.8
Delayed gross motor skills and speech	4	10.8
Total	**37**	**100**

Table 5: Time to Follow-up EEG (weeks)

Cluster	*Count*	*Percentage*
1- 10	10	28.6
11-20	4	4.8
21-30	3	2.4
31-40	3	4.8
51-60	1	3.2
Above 60	1	3.2
Not done	9	29.0
Total	**31**	**100**

Table 6: Follow-up EEG Findings

	Count	*Percentage*
Generalized poly spikes and wave	1	2.4
Bilateral independent poly spike and wave and spike and wave with generalization	2	4.8
Left sided spike and wave activities	1	2.4
Focal seizure disorder involving the right frontal central and parietal area	1	2.4
Seizure disorder prominent over the left side	1	2.4
Bilateral independent spikes and poly spikes prominent over the left temporal area	1	2.4
Sharp and wave discharges over the left frontal and temporal region	1	2.4
Posterior sharp and wave activities with a diffuse slow background	1	2.4
Multifocal spike and wave activities noted more over the left side	1	2.4
Bi-frontal sharp waves prominent over the left frontal area. Independent right sided sharply contoured activities.	1	2.4
Left temporal sharply contoured slowing activities	1	2.4
Right temporal and bi-posterior spike and wave	1	2.4
Normal	5	11.9
Missing	4	9.5
Total	**22**	**100**

DISCUSSION

Early identification and prompt treatment of infantile spasms are essential to achieving the best possible outcomes for affected young children. However, several significant challenges hinder this goal, including frequent misdiagnosis or failure to recognize the seizures, limited treatment response, and the risk of relapse, hence the need to evaluate the clinical description of infantile spasms in this setting.

In this study, the average time from spasm onset to diagnosis and treatment was 2 months, which is shorter than the 3.1 months reported in the South African study (5). This delay could be attributed to inefficient referral to the neurology service and is consistent with findings by Abath et al., who emphasized the importance of early referrals to improve outcomes (1). We believe that the longer lead times to treatment contributed to the poor developmental and seizure outcomes, as reported

in other studies (5, 15, 17). Paprocka et al. emphasized the need for randomized, controlled clinical trials with extended follow-up periods to understand treatment outcomes better (10).

Visual inattention was commonly reported at presentation, aligning with prior observations (2). Most patients experienced flexor spasms, while fewer presented extensor or mixed spasms. This pattern resonates with other findings, which identified flexor spasms as the most prevalent type (6). Various perinatal courses, including neonatal seizures and delayed cry, were observed, which are also discussed in a separate study (4). Initial EEG findings predominantly showed modified hypsarrhythmia and hypsarrhythmia, reinforcing the diverse electrographic findings in epileptic spasm syndrome as noted in the literature (6, 7).

The etiology of IESS remains multifaceted. Among our cohort, a third of the cases had an unknown aetiology; limited access to genetic studies may have inflated this figure. Hypoxic ischemic encephalopathy, congenital brain malformations, and Tuberous sclerosis complex were identified as contributing factors in varied proportions. These findings align with another study, which also reported associations between infantile spasms and hypoxic-ischemic insults, tuberous sclerosis, and cortical malformations (8, 9).

First-line treatments for infantile spasms include hormonal therapy or vigabatrin. The ICISS study suggested that a combination of ACTH/hydrocortisone and vigabatrin is more effective at stopping infantile spasms (11). In this study center, vigabatrin in combination with prednisolone was the standard of care for all patients, irrespective of underlying etiology. Many patients in our study were initially treated with non-first-line therapy at other facilities before being referred to the neurology clinic. This practice is consistent with other studies, in which most patients received ineffective antiseizure medication before referral (1, 5). This indicates a lack of awareness among clinicians and a lack of clear national guidelines for diagnosing and managing IESS. Furthermore, Mao et al. demonstrated that early administration of standard therapies is superior to non-standard options (12).

In this study, treatment response at six weeks was complete in slightly over half of the patients and partial in less than half. This aligns with findings from Kulsoom et al., who similarly categorized treatment outcomes as complete or partial response based on seizure reduction (17). There was no perceived effect of etiology on the rate of cessation of spasms in keeping with the United Kingdom Infantile Spasm Study (UKISS) (19). Excessive sleepiness was reported after initiating vigabatrin at 94mg/kg/day; the symptoms resolved after a dose reduction followed by gradual increments. We recommend starting at a lower dose and increasing it based on response. High-dose prednisone was well tolerated with few instances of short-term adverse effects. No patients required treatment withdrawal, as reported in another study (11).

Limited resources likely impacted the acquisition of follow-up EEGs for some patients unlike in the ICISS study where pre-treatment and post-treatment EEGs were obtained between day 14 and day 21 (11). Our follow-up EEG findings showed normal results in slightly more than a quarter of the patients, contrasting with another study that reported no change in 75% of cases on ACTH (10). All patients with normalized EEGs at 2 weeks had accompanied clinical resolution of spasms. A 2022 paper on EEG biomarkers in infantile spasms notes that a short lag time between onset and treatment is a favourable prognostic factor, underlining that prompt therapy improves clinical outcomes for patients with IESS (20). There were two deaths that were not linked to the diagnosis or treatment of IESS, both patients had developed respiratory symptoms prior to their demise. Harini et al. reported similar causes of death in infantile spasms, with respiratory failure and cardiac arrhythmia being common causes unrelated to epilepsy (18).

CONCLUSION

Longer lead times to treatment were associated with developmental delay. Standardized referral pathways are needed to ensure prompt diagnosis and management of IESS. Vigabatrin in combination with prednisolone was more effective than non-first-line anti-seizure medication in achieving epileptic spasm freedom. Clear national guidelines for the management of IESS are needed in this setting.

Limitations

The study had limitations, including restricted access to genetic studies. Accessible and affordable genetic testing could identify the genetic causes of IESS in Kenya and support precision therapy for individual patients. Only a few patients had a repeat EEG at 2 weeks of treatment. There is a need for expanded, subsidized EEG services for children who need this investigation. This was a single-center study of a rare epilepsy syndrome,

which accounted for the small cohort recruited. This limits the generalizability of the results from this study. There was also no routine monitoring for visual field defects associated with vigabatrin. However, visual field defects are typically associated with prolonged high-dose vigabatrin administration. In this study, vigabatrin was tapered and stopped after 6 months of treatment. Moreover, there was no standardized developmental assessment tool; instead, developmental progress was based on parental reports of milestones achieved and clinical evaluations by the attending neurologist, which also limits the reproducibility of this data. A multicenter, prospective study is recommended further to elucidate the developmental outcomes of IESS in the region.

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Conflict Of Interest: None

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