Clinical profile and electroencephalogram characteristics of children with self-limited epilepsy with centro-temporal spikes attending paediatric neurology clinic, Teaching Hospital, Karapitiya, Sri Lanka

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Abstract

Introduction: Self-limited epilepsy with centro-temporal spikes (SLECTS) is the commonest type of epilepsy in childhood. Though considered benign in the past, there is growing evidence of its multiple neuropsychiatric involvement. Antiepileptic drugs (AEDs) were not recommended in children with SLECTS due to its benign nature.

Objectives: To describe the clinical profile and electroencephalogram (EEG) characteristics of children with SLECTS followed up at the paediatric neurology unit, Teaching Hospital Karapitiya.

Method: A descriptive cross-sectional study, using an interviewer-administered questionnaire, was carried out at the paediatric neurology clinic, THK from December 2021 to February 2022.

Results: A total of 113 children with SLECTS was included in the study. Of them, 61.1% were males. Mean age of sample was 10±2.8 years. Mean age of onset of seizures was 7±2.5 years; 87.5% were diagnosed after one seizure episode and 13.3% had a history of febrile seizures. Developmental delay was evident among 5 (4.4%) patients. Speech arrest (91.2%), hypersalivation (74.3%) and hemifacial sensory-motor seizures (56.6%) were the main manifestations; 87.6% had seizures at night. Abnormal EEGs were found in 110 (97.3%) patients and 51.3% had bilateral typical SLECTS changes in the EEG. Twenty-seven (23.9%) parents had concerns about their children's school performance. Behavioural problems were reported in 37 (32.7%). Child psychiatry referrals were done in 12 (10.2%). AEDs were used in 72.6% patients and 27.4% achieved remission. Sodium valproate was the most widely used AED (79.6%). Most patients were managed with monotherapy.

Conclusions: There was male predominance. Mean age of onset of seizures was 7 years; 87.5% were diagnosed after one seizure and 87.6% had seizures at night. Predominant EEG characteristic was bilateral typical centro-temporal spikes and waves. Speech arrest (91.2%), hypersalivation (74.3%) and hemifacial sensory-motor seizures (56.6%) were the main manifestations of the seizures.

(Key words: Self-limited epilepsy with centro-temporal spikes, Clinical profile, Sri Lanka)

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Introduction

Self-limited epilepsy with centro-temporal spikes (SLECTS), earlier known as benign epilepsy with centro-temporal spikes (BECTS), is the commonest type of childhood epilepsy[1]. Incidence of SLECTS is 10-20 per 100,000 children aged 3-15 years, and it accounts for 8-25% of all childhood epilepsy[1]. It is more common among males[1]. Though considered a benign condition in the past, there is growing evidence of its multiple neuropsychiatric involvement. SLECTS was first described in 1952 by Gastaut, but the name Rolandic was used after the pioneering work by Luigi Rolando about localising the anatomical location of the epilepsy origin[2].

Children with SLECTS classically present around 6-7 years of age, during sleep. Oropharyngeal symptoms such as grunting, gurgling, numbness and paraesthesia are the common manifestations of SLECTS seen in 50% of patients. Speech arrest and hypersalivation are seen in 40% and 30% of children with SLECTS, respectively. In addition, 30% of children with SLECTS have focal facial sensory-motor symptoms such as jaw numbness and tonic deviation of mouth[2,3]. Neuropsychological manifestations such as speech disorders, reading disorders and linguistic problems occur during an acute episode of SLECTS. However, a learning disability can be associated with SLECTS even in

the presence of longstanding seizure-free periods[4]. Atypical SLECTS tends to occur during the daytime and is associated with focal seizures with Todd's paralysis and status epilepticus. In addition, children with atypical SLECTS are more prone to develop neuropsychiatric manifestations such as Landau–Kleffner syndrome (LKS) and encephalopathy with status epilepticus in sleep (ESES). Conversely, 1.3–4.6% of children with typical SLECTS can develop these neuropsychiatric manifestations[4,5].

Objectives

The main objective of the study was to describe the clinical profile and EEG characteristics of children with SLECTS followed up in the paediatric neurology unit, Teaching Hospital Karapitiya (THK). Other objectives were to describe associated neurocognitive abnormalities among children with SLECTS and the pattern of antiepileptic usage.

Method

Study setting, design and period: The study was conducted at the Paediatric Neurology Unit in THK, Galle, the only paediatric neurology centre in the entire southern province. The study group included 128 children diagnosed with SLECTS for a minimum duration of three months, who were followed up at the paediatric neurology clinic and the general paediatric clinics at THK.

***Data collection and analysis*:** A convenient sample of consecutive patients with SLECTS attending the paediatric neurology clinic, THK, was enrolled. A pre-tested interviewer-administered questionnaire, consisting of three components, was used. In the first part, basic socio-demographic details of the child and the family were obtained. Disease-related data, such as the onset of the condition, semiology of seizures, precipitating factors, seizure frequency and associated neuro-behavioural concerns were recorded in the second part. Screening questions were included in the third part of the questionnaire to identify children with neurocognitive deficiencies. The third part of the questionnaire was completed after going through the patients' medical notes. Data analysis was carried out using SPSS version 20.

Ethical issues: Approval for the study was obtained from the Ethical Review Committee of the Sri Lanka College of Paediatricians (No. SLCP/ ERC/ 2021/ 31). Patients were enrolled in the study during their routine clinic visits and they were given an information sheet before enrollment. Informed written consent was obtained from the caregiver accompanying the child.

Results

A total of 128 children with SLECTS was enrolled in the study but only 113 were taken for the analysis due to missing data. Of them, 61.1% were males. Mean age of the sample was 10 ± 2.8 years. Mean age of the onset of seizures was 7 ± 2.5 years. Seven (6%) children were born to consanguineous parents. The basic sociodemographic features are depicted in the Table 1.

Table 1: ***Basic sociodemographic features***

Sociodemographic characteristic	Number (%)
Sex	
Male	69 (61.2)
Female	44 (39.8)
Age (years)	
<8	18 (15.9)
8-12	62 (54.8)
>12	33 (29.2)
Ethnicity	
Sinhala	110 (97.3)
Muslim	03 (02.7)
Mother's educational level	
Primary	03 (02.7)
Secondary	93 (82.3)
Graduate	13 (11.5)
Postgraduate	04 (03.5)
Father's educational level	
Primary	04 (03.5)
Secondary	96 (84.9)
Graduate	10 (08.8)
Postgraduate	03 (02.7)
Number of siblings	
None	16 (14.2)
1	49 (43.4)
2	38 (33.6)
>2	10 (08.8)

Ninety-nine (87.5%) were diagnosed after one seizure episode and 15 (13.3%) had a history of febrile seizures. Twenty-two patients gave a history of childhood epilepsy among first degree relatives. In 87.6% cases, seizures occurred during the night-time sleep. There were 5 (4.4%) children who had developmental delay. The predominant EEG characteristic was bilateral typical centro-temporal spikes and waves. Around 11% had atypical EEG features and one patient had features of electrical status epilepticus in sleep (ESES). Disease related information is given in Table 2. Seizure semiology is illustrated in Figure 1. Speech arrest (91.2%), hyper salivation (74.3%) and hemifacial sensory-motor seizures (56.6%) were the main manifestations.

Table 2: ***Disease related information***

Disease related information	Number (%)
Age of onset (years)	
<6	28 (24.8)
6-8	46 (40.7)
9-11	34 (30.0)
>11	05 (04.4)
Number of identified seizures before the diagnosis	
1	99 (87.6)
2 or more	14 (12.4)
Number of seizures during the previous 12 months	
None	24 (21.2)
1	64 (56.6)
2 or more	25 (22.0)
History of febrile seizures	
Yes	15 (13.3)
No	98 (86.7)
Family history of childhood epilepsy	
Yes	22 (19.5)
No	91 (80.5)
Currently on antiepileptics	
Yes	85 (75.2)
No	28 (24.8)
Timing of seizures	
Only during sleep	99 (87.6)
Only when awake	05 (04.4)
Both during sleep and when awake	09 (07.9)
Neuroimaging	
Not done	72 (63.7)
Normal	33 (29.2)
Abnormal	08 (07.1)
Electrocardiogram characteristics	
Normal	04 (03.5)
Typical unilateral SLECTS	38 (33.6)
Typical bilateral SLECTS	58 (51.3)
Atypical SLECTS	12 (10.6)
ESES	01 (0.9)

SLECTS: Self-limited epilepsy with centro-temporal spikes, ESES: Electrical status epilepticus during slow-wave sleep

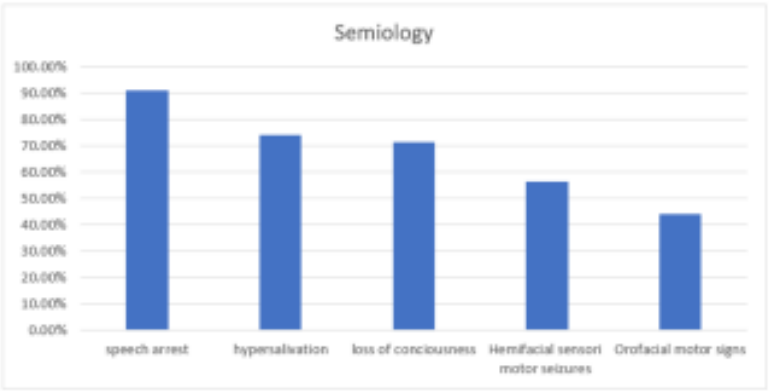

Figure 1: ***Seizure semiology***

Abnormal EEGs were found in 110 (97.3%), and 51.3% had bilateral typical SLECTS changes in the EEG. Electrical status epilepticus during slow-wave sleep (ESES) was seen in one child. Neuroimaging (either CT or MRI brain) was done in 41 (36.3%) patients, and abnormalities were detected in eight children. Behavioural problems were reported in 37 (32.7%) cases. Child psychiatry referrals were done on 12 (10.2%) occasions. Some form of developmental delay prior to the onset of seizures was noted in 4.4% of patients. Table 3 illustrates the neurocognitive concerns of children with SLECTS.

Table 3: ***Neurocognitive concerns of children with SLECTS***

Neurocognitive concern	Number (%)
History of developmental delay	05 (04.4)
Attending special school	04 (03.5)
Maternal concerns about scholastic skills	27 (23.9)
Teachers' concerns about school performance	23 (20.4)
Maternal concerns about behaviour	37 (32.7)
School refusal	17 (15.0)
Under child psychiatrist follow up	12 (10.6)

For the analysis, patients with normal EEG and EEG features with typical centrotemporal spikes were amalgamated as one group (typical SLECTS). Those who had atypical EEG features and ESES were taken as one group and considered as an atypical group. There was no significant difference in seizure semiology between the typical and the atypical groups. However, concerns regarding academic skills (p=0.00), school refusal (p=0.01) and behavioural problems (p=0.01) were more in the atypical group (Table 4).

Table 4: ***Comparison between the typical and atypical SLECTS***

Characteristic	Typical SLECTS *(n=100)* *n (%)*	Atypical SLECTS *(n=13)* *n (%)*	Chi-square	p- value
Male	60 (60)	09 (69.2)	0.63	0.42
Past history of febrile seizures	14 (14)	01 (07.7)	0.39	0.52
Hemi facial sensory motor seizures	57 (57)	07 (53.8)	0.04	0.83
Oropharyngeal ictal manifestations	44 (44)	06 (46.1)	0.02	0.88
Speech arrest	91 (91)	12 (92.3)	0.02	0.87
Hyper salivation	76 (76)	08 (61.5)	1.2	0.26
Poor school performance	20 (20)	07 (53.8)	7.2	**0.00**
Behavioural concerns	29 (29)	08 (61.5)	5.1	**0.01**
School refusal	12 (12)	05 (38.4)	6.3	**0.01**
Aggressive behaviours	28 (28)	07 (53.8)	3.5	0.05
Child psychiatry referral done	05 (05)	07 (53.8)	28.8	**0.00**
Abnormal neuroimaging	05 (05)	03 (23.1)	5.7	0,01

Eighty-five (75.2%) were on AEDs at the time of recruiting into the study and 72 (63.7%) were on monotherapy. However, 92 (81.4%) patients were on AEDs at some point of the illness. Sodium valproate remained the most common monotherapy (79.6%) followed by oxcarbazepine (16.8%) and levetiracetam (4.4%). Clobazam was the most common add-on AED.

Discussion

This is the first study conducted in Sri Lanka to assess the clinical profile of children with SLECTS. However, a few studies were done elsewhere. The male: female ratio in SLECTS is 3:2, and in this study, there were 61% males, which is compatible with the standard ratio[5,6]. The mean age of this cohort was around ten years, whereas the mean age was 8.8 years, 7.7 years and 7.2 years in studies by Liu M, *et al*[5], Trave TD, *et al*[6] and Sable S, *et al*[7], respectively. The positive family history and SLECTS association range between 3.5-59%[8,9]. The current study showed a 19.5% positive family history, which is on par with previous studies. The most common semiology found in this study were speech arrest (91%), hypersalivation (74%), loss of consciousness (71%), hemifacial sensory-motor seizures (57%) and oropharyngeal ictal manifestations (44%). A study by Sable S, *et al*[7] showed that 81% of patients had hemifacial seizures, and 42% had hypersalivation.

This study's most common (51%) EEG finding was bilateral discharges in the Rolandic region, followed by typical unilateral changes in 38% of patients. However, a study by Liu M, *et al*[5] revealed that 71% and 29% of patients had unilateral and bilateral EEG features, respectively. Similarly, studies by Bedoin N, *et al*[10], Riva D, *et al*[11] and Zhao X, *et al*[12] showed unilateral EEG changes on 60%, 66.7% and 75% occasions respectively. According to the study by Liu M, *et al*[5], ESES was found in 2% of patients, but in our study, only one patient (0.9%) had EEG compatible with ESES.

In the past AEDs were not recommended in children with SLECTS due to their benign nature. However, with the current understanding of the disease entity, a significant number of children with SLECTS are on AEDs. Liu M, *et al*[5] revealed that around 75% of patients were on some form of AEDs, and 10.6% were on multidrug therapy. Similarly, in our study, 75% of children were on AEDs at the time of the study, and 81% had received AEDs at some stage of the disease. In contrast, two studies by Sable S, *et al*[7] and Kessi M, *et al*[13] have revealed that all children were on at least one AED. In our study, sodium valproate remained the most commonly used drug. However, different studies showed different drugs as the most prevalent medication. According to Liu M, *et al*[5], oxcarbazepine (37%) was the most widely used drug, whereas, levetiracetam was the most prevalent treatment method based on the study results of Kessi M, *et al*[13]. The studies' availability, cost, and timing would have contributed to the drug of choice in patients with SLECTS. Since our research was conducted in a paediatric neurology clinic in a referral center, a higher percentage of children would have been on AEDs.

Neuropsychiatric manifestations are known to be associated with SLECTs, and this study assessed only parental perceptions about the behaviour and cognitive functions of their children. Around one-third of parents had concerns regarding the conduct of their children. Moreover, 24% of parents were not happy about their children's school performances. A study by Ozgen Y, *et al*[14] showed that 29% of children with SLECTS had attention deficit hyperactive disorder, and 5% had oppositional defiant disorder. However, the current study did not aim to diagnose underlying neuropsychiatric disorders using a diagnostic tool, but only the parental perception regarding the child's cognitive behavioural aspects was taken. Furthermore, children with atypical SLECTS had a more frequent occurrence of school refusal, behavioural concerns, poor school performance and child psychiatry involvement. Similar findings were observed in a number of previous studies[5,6,7,13].

The study had some limitations. It did not assess the neurocognitive aspects using a standard tool and the IQ assessments were not done. The study did not consider the neurocognitive differences between those who were on AEDs and those who were not. More children with SLECTS were on AEDs, and neurocognitive concerns were more common in atypical SLECTS. Therefore, it is important to conduct objective assessments of the neurocognitive outcome of children with SLECTS, preferably using a case-control study.

Conclusions

There was a male predominance. Mean age of onset of seizures was around 7 years; 87.5% were diagnosed after one seizure episode and 87.6% had seizures at night. The predominant EEG characteristic was bilateral typical centro-temporal spikes and waves. Speech arrest (91.2%), hypersalivation (74.3%) and hemifacial sensory-motor seizures (56.6%) were the main manifestations of the seizure.

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References

1. Pavlou E, Gkampeta A, Evangeliou A, Athanasiadou-Piperopoulou F. Benign epilepsy with centro-temporal spikes (SLECTS): Relationship between unilateral or bilateral localization of interictal stereotyped focal spikes on EEG and the effectiveness of anti-epileptic medication. *Hippokratia* 2012; **16**(3): 221-4.

2. Parakh M, Katewa V. A review of the not so benign-benign childhood epilepsy with centrotemporal spikes. *Journal of Neurology and Neurophysiology* 2015; **6**(4): 1-4.

3. Jun YH, Eom TH, Kim YH, Chung SY, Lee IG, Kim JM. Changes in background electroencephalographic activity in benign childhood epilepsy with centrotemporal spikes after oxcarbazepine treatment: a standardized low-resolution brain electromagnetic tomography (sLORETA) study. *BMC Neurology* 2019; **19**(1): 1-8. https://doi.org/10.1186/s12883-018-1228-8
PMid: 30606133 PMCid: PMC6317234

4. Dryżałowski P, Jóźwiak S, Franckiewicz M, Strzelecka J. Benign epilepsy with centrotemporal spikes–Current concepts of diagnosis and treatment. *Neurologia i Neurochirurgia Polska* 2018; **52**(6): 677-89. https://doi.org/10.1016/j.pjnns.2018.08.010
PMid: 30219586

5. Liu M, Su X, Shi X, Wu G, Zhang Y, Gao L, *et al*. Clinical features of benign epilepsy of childhood with centrotemporal spikes in

Chinese children. *Medicine (Baltimore)* 2017; **96**(4): e5623. https://doi.org/10.1097/MD.0000000000005623 PMid: 28121917 PMCid: PMC5287941

6. Travé TD, Yoldi-Petri ME, Gallinas-Victoriano F, de Gurtuba IG. Rolandic epilepsy: Epidemiological, clinical and evolutionary characteristics. *Anales de Pediatría* 2008; **68**(5): 466-73. https://doi.org/10.1157/13120044 PMid: 18447991

7. Sable S, Sable R, Tamhankar P, Tamhankar V. Clinical profile of patients with Rolandic epilepsy at a clinic in rural Maharashtra. *Journal of Family Medicine and Primary Care* 2021; **10**(3):1263-6. https://doi.org/10.4103/jfmpc.jfmpc_1355_20 PMid: 34041163 PMCid: PMC8140235

8. Bouma P, Bovenkerk AC, Westendorp R, Brouwer O. The course of benign partial epilepsy of childhood with centrotemporal spikes: A meta-analysis. *Neurology* 1997; **48**(2): 430-7. https://doi.org/10.1212/WNL.48.2.430 PMid: 9040734

9. Xiong W, Zhou D. Progress in unraveling the genetic etiology of Rolandic epilepsy. *Seizure* 2017; **47**: 99-104. https://doi.org/10.1016/j.seizure.2017.02.012 PMid: 28351718

10. Bedoin N, Herbillon V, Lamoury I, Arthaud-Garde P, Ostrowsky K, De Bellescize J, *et al*. Hemispheric lateralization of cognitive functions in children with centrotemporal spikes. *Epilepsy and Behavior* 2006; **9**(2): 268-74. https://doi.org/10.1016/j.yebeh.2006.06.002 PMid: 16875880

11. Riva D, Vago C, Franceschetti S, Pantaleoni C, D'Arrigo S, Granata T, *et al.* Intellectual and language findings and their relationship to EEG characteristics in benign childhood epilepsy with centrotemporal spikes. *Epilepsy and Behavior* 2007; **10**(2): 278-85. https://doi.org/10.1016/j.yebeh.2006.12.003 PMid: 17267289

12. Zhao X, Chi Z, Chi L, Shang W, Liu X. Clinical and EEG characteristics of benign Rolandic epilepsy in Chinese patients. *Brain and Development* 2007; **29**(1): 13-8. https://doi.org/10.1016/j.braindev.2006.05.006 PMid: 16806777

13. Kessi M, Yan F, Pan L, Chen B, Olatoutou E, Li D, *et al.* Treatment for the benign childhood epilepsy with centrotemporal spikes: a monocentric study. *Frontiers in Neurology* 2021; **12**: 233744998. https://doi.org/10.3389/fneur.2021.670958 PMid: 34025572 PMCid: PMC8134665

14. Özgen Y, Güngör M, Kutlu M, Kara B. Clinical and electrophysiological predictors of behavioral disorders in patients with benign childhood epilepsy with centrotemporal spikes. *Epilepsy and Behavior*. 2021; **121**: 108037. https://doi.org/10.1016/j.yebeh.2021.108037 PMid: 34058495