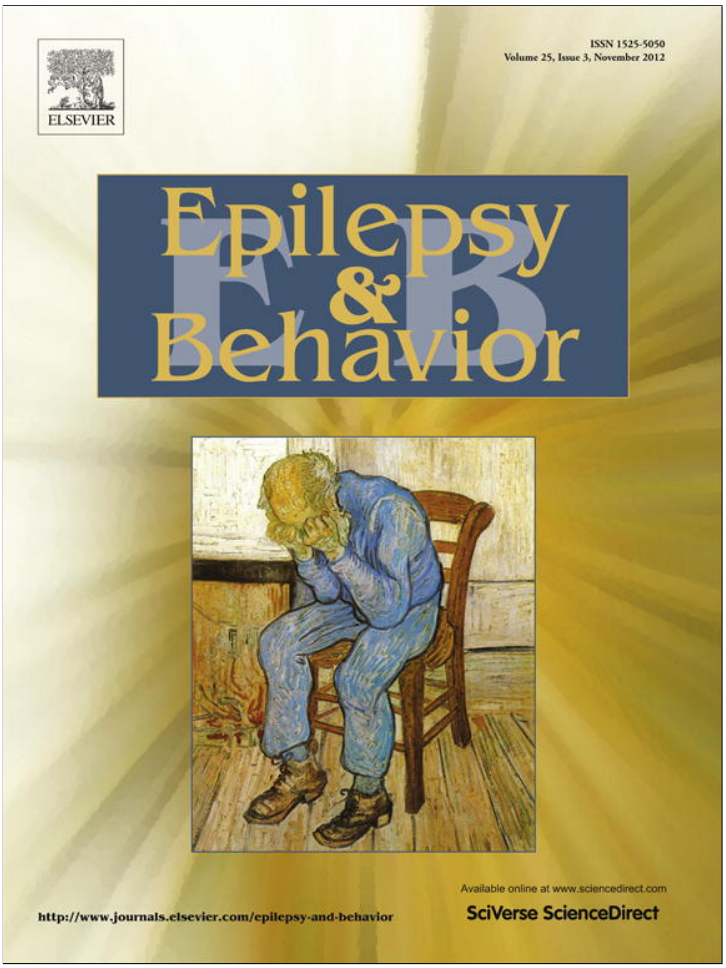

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Neurocognitive and neurobehavioral disabilities in Epilepsy with Electrical Status Epilepticus in slow sleep (ESES) and related syndromes

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ABSTRACT

Aim: The aims of this study were to assess the cognitive and behavioral problems of patients with Epilepsy with Electrical Status Epilepticus in slow sleep (ESES) and related syndromes and to review their EEG (electroencephalography) findings and treatment options.
Results: Fourteen patients with ESES were evaluated and treated in 2010. Nine children had continuous spike and wave during slow-wave sleep (CSWS)/ESES syndrome, 3 had Atypical BECTS (benign epilepsy with centrotemporal spikes), 1 had Opercular syndrome, and 1 had Landau–Kleffner syndrome. The duration of ESES ranged from 6 to 52 months. Eleven (91%) children had behavioral issues, most prominent being hyperactivity. Seven of the 13 children (53%) showed evidence of borderline to moderate cognitive impairment. A total of 28 EEG findings of ESES were analyzed for SWI (spike-wave index). Antiepileptic drugs received by the patients included valproate, clobazam, levetiracetam, and others. Eleven patients had been treated with oral steroids and it was found to be efficacious in seven (63%).
Conclusion: Disabilities caused by ESES affect multiple domains. Patients with an SWI > 50% should be followed up frequently with neuropsychological assessments. Steroids appear to be effective, although there is a need to standardize the dose and duration of treatment.

1. Introduction

Epilepsy with Electrical Status Epilepticus in slow sleep (ESES) was first described in 1971 [1]. It is an age-related, self-limited disorder and is characterized by the following features: epilepsy with different seizure types, neuropsychological impairment in the form of global or selective regression of cognitive functions, motor impairment, and typical EEG (electroencephalography) findings of continuous epileptic activity occupying ≥85% of non-rapid eye movement (REM) sleep [2]. The term CSWS (continuous spike and wave during slow-wave sleep) has been used synonymously with ESES. It is defined as a functional disorder of childhood with the following characteristics: severe EEG disturbance occupying at least 85% of sleep, seizures, behavioral deterioration, no demonstrable brain pathology and stabilization or improvement of behavior once the EEG abnormality resolves [3].

This retrospective analysis aimed to describe the clinical spectrum of patients with ESES with an emphasis on the associated cognitive impairments and behavioral issues. This study also analyzed the pattern of epileptic activity in these patients and the yield of various therapeutic options used for ESES.

2. Method

The patient records in the pediatric neurology clinic at P D Hinduja National Hospital were screened for ESES syndrome.

2.1. Inclusion and exclusion criteria

All newly diagnosed or follow-up patients with appearance of continuous or semi-continuous epileptic activity in non-REM slow-wave sleep who attended the pediatric neurology clinic during the year 2010 were included. 'Continuous' epileptic activity was considered to be synonymous with an SWI (spike-wave index) of >85% and 'semi-continuous' with an SWI between 50 and 85%. These patients also had documented evidence of functional regression associated with worsening EEG findings. Patients with other epileptic encephalopathies like Lennox–Gastaut syndrome were excluded.

All the recruited patients were called to the clinic for an evaluation to document their current status. A total of 14 children were included in the analysis. All of the children had documentation of any of the following: developmental arrest, new-onset cognitive impairment,

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new-onset behavioral disorders, motor regression, or oromotor dysfunction.

Clinical syndromes related to ESES were classified as the following [4]: *CSWS syndrome* — global cognitive decline, (atonic) seizures, and motor disturbances with an SWI of >85% or >50%; *Landau–Kleffner syndrome* (*LKS*) — receptive/mixed aphasia, verbal agnosia, infrequent seizures, and an SWI of any % (bitemporal/diffuse); *Atypical BECTS* (benign epilepsy with centrotemporal spikes) — mostly nocturnal (partial) seizures with cognitive and behavioral disturbances, and an SWI of >50% (local/diffuse); and *Opercular syndrome* — resembling BECTS, opercular features, and an SWI of >50%. This acquired epileptiform opercular syndrome is characterized by oro-facio-lingual dysfunction with dysarthria, weakness of facial and tongue muscles, rolandic seizures, and atypical absences [2].

2.2. EEG

All children had one pre-ESES EEG and 1–4 sleep EEGs with ESES/near ESES. Epilepsy with Electrical Status Epilepticus in slow sleep was defined as ≥85% of SWI in non-REM sleep. Near ESES was defined as 50–85% of SWI in non-REM sleep. All EEG findings were visually calculated for SWI. Since this was a retrospective study in patients with a clinical diagnosis of ESES, SWI was defined as percentage of seconds with ≥1 spike-wave complex in 30 min or more of non-REM sleep [4]. Electroencephalography findings were also analyzed for distribution and lateralization of discharges.

2.3. Therapies

Details of the various therapies tried in these patients were reviewed. Response to therapy was analyzed on the basis of clinical and EEG improvement.

2.4. Neurocognitive and behavioral evaluation

Thirteen children had neuropsychological assessments. Information from occupational and speech therapy assessment was also included. These children underwent an evaluation during ESES which included intelligence testing (Intelligence Quotient — IQ score) and/or evaluation of development (Development subscale scores), and/or social–adaptive functioning (Social Quotient — SQ score). One child was assessed only for social and adaptive functioning, and one child did not complete the evaluation.

Behavior was assessed on at least one out of the four standardized parent-rated behavior scales and on clinical interviews using the DSM IV (Diagnostic and Statistical Manual of Mental Disorders) criteria (see Appendix 1).

Each test/scale was scored and classified as per the standardized guidelines for that test; then, for the purpose of uniformity, the scores across tests and scales were re-classified on a *performance-based scale* as normal = 1, borderline/mild = 2, and moderate/severe = 3. Thus, for example, in the intelligence test and the social–adaptive functioning scale, the IQ and SQ scores were derived respectively and categorized as per the DSM IV classification as normal (>84), borderline (71–84), or challenged (<71) which was equivalent to the scores 1, 2, and 3, respectively, on the performance-based scale. For the development screening, test scores were interpreted on the basis of 'risk categorization' as per the scale manual guidelines wherein items were marked as pass/fail, and number of items in which the child's scores were below the expected age determined whether the child was classified as within normal, suspect/questionable, or delayed range.

For the behavioral scales, interpretation of the scores was as per the cutoff scores provided in the test technical manuals. Thus, for example, in the Conners' Behavior Rating Scale the T score of <60 is in the normal range, 61–70 = mildly atypical range, and >70 = markedly atypical range which was equivalent to 1, 2, and 3 on the performance-based scale. Children whose behaviors were rated as 3 (moderate to severe issues) on the performance-based scale were further evaluated using the DSM IV criteria for classification into the various clinical conditions such as attention deficit hyperactivity disorder or pervasive developmental disorders.

Academic failure was defined as securing marks or grades below the passing criteria set by the school in the annual exams or dropping out of school due to inability to cope with the academic demands and/or failures.

3. Results

Fourteen children (8 males and 6 females) were included in the analysis. They were either newly diagnosed or follow-up patients seen in the pediatric neurology clinic in 2010 and were under treatment from 2002 to 2010.

3.1. Seizures and ESES

All 14 children had seizure onset before the development of ESES with a mean age of seizure onset of 3 years and 6 months (range 8 months–9 years). The duration of follow-up varied from 6 months to 8 years. Mean age of documented ESES in EEG was 6 years and 10 months and duration of ESES was 17.5 ± 13.7 months (range 6–52 months). The average time lag between the onset of epilepsy and ESES was 3 years and 6 months.

Nocturnal focal motor seizure was the most frequent seizure type in these patients (Table 1).

All 14 patients had an EEG of the pre-ESES period, and there was presence of focal interictal epileptic discharges in 11 (78%) patients during either the awake or sleep state. Clinically, during the pre-ESES period, all patients were seeking treatment for infrequent seizures; 9 of them had focal seizures. History of speech delay was present in 3 patients, while motor as well as speech delay was noted in 3.

These 14 patients had a total of 28 EEGs which fulfilled the criteria for ESES/near ESES. Twenty-one (75%) of them had an SWI of more than 85%. Distribution of the discharges did not definitely correlate with the clinical ESES syndrome. The syndromes and the corresponding analysis of the EEG findings are shown in Table 2.

3.2. Clinical syndrome related to ESES

Based on the diagnostic criteria, 9 patients were classified as CSWS, 3 as Atypical BECTS, one as LKS, and one patient as Opercular syndrome (Table 1).

3.3. Cognitive impairments and behavioral problems

On the basis of the performance-based classification of 1 = normal, 2 = borderline/mild, and 3 = moderate/severe, seven (53%) out of 13 children showed evidence of mild to moderate cognitive challenges/developmental delay; their score on the intelligence test/social–adaptive scale/development scale was below the normal range (Table 3 and Fig. 1). Eight (61%) had academic failures or were school dropouts. Eleven (91%) out of 12 had behavioral issues with mild or markedly atypical behaviors, most prominent being hyperactivity and poor social interaction (Table 3 and Fig. 1). Behavioral problems rated in the moderate–severe range, which fulfilled the DSM IV diagnostic criteria warranting diagnosis of a comorbid disorder, were present in eleven; six (50%) had ADHD (attention deficit hyperactivity disorder), two (16%) had ADD (attention deficit disorder), two (16%) had PDD (pervasive developmental disorder), and three (25%) had LD (learning disability) (Table 3 and Fig. 2). Two patients who did not undergo a complete evaluation were not attending school.

Table 1
Seizure type, frequency, and clinical syndrome in 14 children with Epilepsy with Electrical Status Epilepticus in slow sleep (ESES).

Patient serial number	Seizure types	Seizure frequency	Clinical syndrome of ESES/diagnosis of epilepsy
1	GTC	Only twice	CSWS
	Focal brief in sleep	1–2/year	Cryptogenic focal epilepsy
2	Nocturnal focal motor	In clusters	CSWS
	Head drops (negative myoclonus)	Many per day with seizure-free periods	Cryptogenic focal epilepsy
3	GTC	1–2/year	LKS
	Nocturnal focal perioral	4–5/month since 6 months	
4	GTC	1/year	CSWS
	Negative myoclonus (slow drops)	Occasional	Cryptogenic focal epilepsy
	Prolonged absence	2–3/year	To rule out progressive disorder (consanguinity present)
5	CPS with oromotor automatisms	1/month earlier. Seizure free since 2 years	CSWS
			Remote symptomatic epilepsy (PVL)
6	GTC	4–30/year	CSWS
			Remote symptomatic epilepsy (right hemi cortical dysplasia)
7	Nocturnal focal motor	Only once	Atypical evolution of BECTS
	Absence	4–5/month for 3 months	
8	Nocturnal focal motor	1–2/year	CSWS
	Negative myoclonus/atypical absence	Many per day since 1.5 years	Cryptogenic focal epilepsy
9	Nocturnal focal motor	1/3 months	CSWS
	Absence	4–5/months	Cryptogenic focal epilepsy
10	Focal motor (left) in sleep	4–5/year	Atypical evolution of BCECTS
11	Focal motor with occasional secondary generalization	1/month	Opercular syndrome
			Cryptogenic focal epilepsy
12	Nocturnal focal motor	1/2 months	CSWS
			Cryptogenic focal epilepsy
13	CPS (left focal)	2–3/month	CSWS
	Negative myoclonus (slow drops)	In clusters	Remote symptomatic epilepsy (NHBI)
14	Focal motor	Only once	Atypical evolution of BECTS

CSWS: continuous spike and wave during slow-wave sleep.
GTC: generalized tonic-clonic seizures.
CPS: complex partial seizures.
LKS: Landau–Kleffner syndrome.
PVL: periventricular leukomalacia.
NHBI: neonatal hypoglycemic brain injury.
BECTS: benign epilepsy with centrotemporal spikes.

3.4. Other problems

Other problems included the following: motor regression in three (25%) out of fourteen patients and severe oromotor dysfunction requiring repeated Ryle's tube feeding in one; there was presence of mild dysfunction like drooling, impaired chewing, and tongue movements in three others.

Table 2
Analysis of electroencephalography (EEG) findings in Epilepsy with Electrical Status Epilepticus in slow sleep (ESES). A total of 28 EEGs were analyzed for 14 children with the diagnosis of ESES.

	LKS	Atypical evolution of BECTS	Opercular syndrome	CSWS
Number of EEGs	3	5	2	18
SWI ≥ 85%	3	4	1	13
SWI 50–85%	0	1	1	5
Lateralization	Left (3/3)	Right (5/5)	Right (1/2)	Right (4), left (6)
Distribution	Centroparietal/ temporoparietal	Unclear	Unclear	Frontal, frontocentral, centroparietal, temporoparietal, temporooccipital

LKS = Landau–Kleffner syndrome.
CSWS = continuous spike and wave during slow-wave sleep.
BECTS = benign epilepsy with centrotemporal spikes.
EEG = electroencephalography.
SWI = spike-wave index.

3.5. Therapies

3.5.1. AEDs (antiepileptic drugs)

Antiepileptic drugs that were tried in these patients included valproic acid — 14 (100%), clobazam — 8 (57%), levetiracetam — 9 (64%), and others including topiramate and lamotrigine.

3.5.2. Steroids

Patients were started on 2 mg/kg/day of oral prednisolone for a period of 4–6 weeks. If no improvement was reported after this period, then steroids were tapered off, and patient was considered to be unresponsive to steroid. If some clinical improvement was noted, then steroids were tapered to a low dose (0.5 mg/kg/day alternate day dosing), and this was continued for a longer duration (up to 3–6 months). Eleven patients were treated with oral steroids and it was found to be efficacious in seven (63%). Clinical improvement was <25% in two patients, 25–50% in two and >50% in three patients.

Clinical improvement meant significant reduction in seizure frequency along with functional improvement noted by parents. Parents provided a subjective measure of improvement in the form of definite change in speech, gait, opercular dysfunctions like drooling, ability to perform ADL (activities of daily living), or better class performance notified by teacher. Electroencephalographic improvement with reduction of SWI to <50% was noted in five. There was definite evidence of recurrence of clinical and EEG findings in five of the seven patients whenever steroids were tapered.

3.5.3. IVIG (intravenous immunoglobulin)

Intravenous immunoglobulin therapy was tried in 4 (28%) patients.

Table 3
Cognition scale, behavior profiles, and academic performance of 14 children fulfilling the diagnosis of Epilepsy with Electrical Status Epilepticus in slow sleep (ESES).

Patient serial number	Cognition	Behavioral issues			Comorbid disorders				Academic performance
	FSIQ/SQ/DSS	Hyperactivity	Conduct disorder	Social/peer problems	ADD	ADHD	PDD	LD	
1	2	3	2	3	0	1	0	0	5
2	2	3	3	3	0	1	0	0	5
3	2	2	1	2	0	1	0	0	3
4	-*	-*	-*	–	-*	-*	-*	-*	-*
5	3	1	2	3	0	0	1	0	5
6	2	3	3	3	0	1	0	0	5
7	1	2	2	1	0	1	0	0	3
8	1	2	1	1	0	0	0	0	5
9	1	2	1	2	0	0	0	1	3
10	1	3	1	1	0	1	0	1	3
11	3	2	1	2	0	1	1	0	4
12	1	3	1	3	0	1	0	0	4
13	3	-*	-*	-*	-*	-*	-*	-*	5
14	1	2	1	1	1	0	0	1	2

-*Information not available.
Cognitive/developmental assessment: IQ/SQ/DSS.
(IQ = Intelligence Quotient, SQ = Social Quotient, DSS = Development subscale score).
1 = normal range, 2 = borderline/mildly challenged, 3 = moderately/severely challenged range.
Behavioral assessment: Parent-rated behavior scores.
1 = normal range, 2 = borderline range, 3 = clinical range.
Comorbid disorders: ADD = attention deficit disorder, ADHD = attention deficit hyperactivity disorder, PDD = pervasive developmental disorder, LD = learning disability, and presence/absence of these comorbid childhood disorders was noted as 0 = absent, 1 = present.
Academic performance assessment: school report scores.
1 = average, 2 = low average, 3 = borderline, 4 = failures, 5 = school dropout due to inability to cope.

3.5.4. Ketogenic diet

Ketogenic diet was tried in 4 (28%) patients.

4. Discussion

ESES is an electrographic pattern associated with focal and/or generalized seizures and neuropsychological, cognitive, and/or motor impairment [2]. Electroencephalography findings of spike and wave discharges in slow-wave sleep could be diffuse, hemispheric, or focal and may be continuous or semi-continuous [2].

The clinical syndrome may vary with presence of atonic seizures, motor and cognitive regression in CSWS/ESES syndrome, and oro-facio-lingual dysfunction in acquired Opercular syndrome [2,4].

In this study, we have analyzed the impact of ESES on cognition, behavior, and academic performances. We found that the majority of the children with ESES had academic failures or were school dropouts. Behavioral issues including hyperactivity and conduct disorders like aggressiveness and poor social relations were present in many, and most of these children had marked problems that fulfilled the criteria for diagnosis of a comorbid disorder. Also, significant cognitive and developmental disabilities were present thereby adding to suboptimal levels of functioning with nine out of 14 (65%) children requiring assistance for performing activities of daily living (ADLs).

Saltik et al. have reported presence of mild to severe disturbances in cognitive and/or behavioral performances such as deterioration in language, hyperactivity, and attention deficit in varying degrees in nearly all cases of ESES [5].

Some studies have shown that cognitive and neuropsychological impairments in ESES spectrum disorders are most resistant to treatment, and the duration of ESES longer than 18 months is associated with definite residual deficit [5,6]. Since the average duration of ESES was 17.5 months in our patients, the significant cognitive and behavioral issues may be secondary to prolonged duration.

It has been proposed that sleep reduces total synaptic weight to an appropriate baseline level, thus, achieving synaptic homeostasis. This has benefits in terms of energy requirement, space requirement,

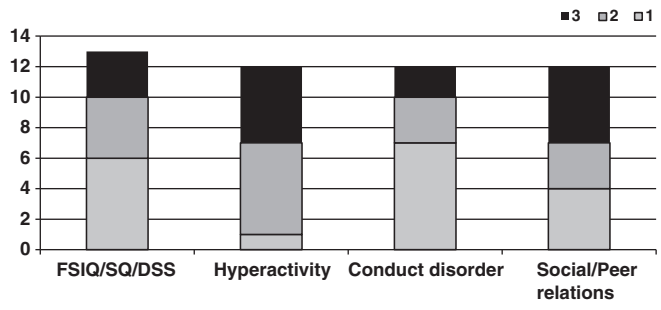

Fig. 1. Column chart representing cognitive and behavioral issues in children with Epilepsy with Electrical Status Epilepticus in slow sleep (ESES). Y axis = number of patients. FSIQ = Full Scale Intelligence Quotient, SQ = Social Quotient, DSS = Development subscale scores (DSS): 1 = normal, 2 = borderline–mild, and 3 = moderately–severely challenged. Parent-rated behavior scores for hyperactivity, conduct disorder and social/peer relations: 1 = normal range, 2 = borderline–mild range, and 3 = moderate–severe range.

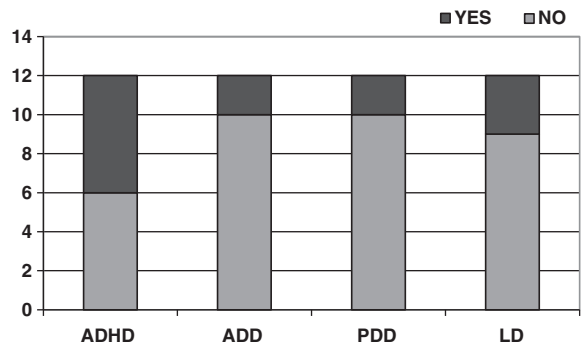

Fig. 2. Column chart demonstrating presence of comorbid disorders in 14 children with a diagnosis of Epilepsy with Electrical Status Epilepticus in slow sleep (ESES). Attention deficit disorder (ADD), attention deficit hyperactivity disorder (ADHD), pervasive developmental disorder (PDD), and learning disability (LD). X axis = comorbidities; y axis = number of patients.

learning, and memory [7]. Prolonged focal epileptic activity during sleep, as occurs in ESES, interferes with local normal slow-wave activity of sleep. Studies have shown that this impairs neural processes and possibly local plastic changes associated with learning and other cognitive functions [8,9].

With respect to treatment, although antiepileptic drugs like valproate, clobazam, and levetiracetam have been said to be 'probably effective' for this epileptic syndrome [10], no large case series is available to support this statement, and in our study, they were not found to be effective in reversing the cognitive impairments acquired during ESES.

Prolonged therapy with high dose oral steroids, for up to or more than one year, was found to be effective in 77% patients, although with high relapse rates of 47% in a case series published by Buzatu et al. [11]. Another study by Sinclair et al. showed improvement with prolonged steroid therapy (prednisolone 1 mg/kg/day for 6 months, 1 year and yearly) in 9 out of 10 patients (of which 8 had LKS) when followed up for 1–10 years [12]. In our study, some or definite clinical improvement was seen in seven out of the 11 patients treated with steroids, although with relapse of symptoms in five. Two or more courses of 8 weeks of oral steroids were tried in five of our patients.

IV immunoglobulin therapy has been found to be effective occasionally in patients with Landau–Kleffner syndrome [13]. In our series, one patient with LKS had some improvement in speech and auditory agnosia following IVIG therapy, while no benefit was noted in 3 patients. Ketogenic diet has been used for the treatment of ESES in isolated cases, and consequently, no conclusions can be drawn about efficacy. In our study, 1 patient had definite improvement in the form of reversal of opercular dysfunction; 2 reported some benefit of which one experienced reduced frequency of repeated atonic head drops and the other patient's parents reported significant benefit in behavior. Both of these patients could not continue on the diet.

Multiple subpial transections (MST) have been tried in patients with LKS with severe aphasia and lateralization of discharges. Morrell et al. reported 50% recovery in speech in a case series including 14 children [14]. A review on surgical treatment in LKS suggested that though two-thirds of the cases appear to benefit, full recovery of function is unlikely following MST [15].

Inutsuka et al. proposed a treatment strategy for ESES that included high-dose valproate therapy (maintaining levels > 100 μg/ml); valproate with ethosuximide; short cycles of high-dose diazepam; and intramuscular adrenocorticotrophin hormone (ACTH) [16]. This study showed definite and permanent remission of ESES in 67% (10 out of 15) of the patients with either valproate monotherapy or a combination therapy with valproate and ethosuximide. This treatment regime was not tried in our patients.

A study by M Van Hirtum-Das et al. suggests that the success of treatment and prognosis is better in idiopathic LKS than that in CSWS occurring in patients with possible symptomatic epilepsy [17]. We did not find any differences in the rate of remission between the various subgroups of ESES.

Limitations of this study include the inability to know the precise timing as to when exactly the ESES had its inception, although a pre-ESES EEG was available in all 14 patients. Another limitation is that the treatments are not readily comparable since a standard treatment algorithm was not followed.

5. Conclusion

The impact of ESES on the development and long-term functional ability of children is significant. The disabilities affect multiple domains of functioning. Patients with an SWI of > 50% should be followed up frequently along with neuropsychological assessments at regular intervals. Steroids appear to be efficacious in some of these patients, though there is a need to standardize the dose and regime. For effective management, it is vital to have a standard algorithm for treating these children based on their phenotypic variability.

Appendix 1. Cognitive and behavioral assessment

Domains	Tests/rating scales
General intelligence	Malin's Intelligence Scale for Indian Children (MISIC) (Indian adaptation of Wechsler Intelligence Scale — WISC)
Development functioning	Denver Developmental Screening Test (DDST)
Social and adaptive functioning	Vineland Social Maturity Scale (VSMS) (Indian adaptation)
Behavior	Child Behaviour Checklist (CBCL) Conners' Parent Rating Scale Strengths and Difficulties Questionnaire (SDQ) Childhood Autism Rating Scale (CARS) DSM IV (Diagnostic and Statistical Manual of Mental Disorders)

References

A. Malin AJ. Malin's Intelligence Scale for Children (MISIC). Indian J Men Retard 1969; 4: 15–25.
B. Malin AJ. Vineland Social Maturity Scale and Manual. Indian adaptation. Mysore: Swayam Sidha Prakashan, 1992.
C. Frankenburg, William K, Dobbs JB. The Denver Developmental Screening Test. J Pediatr 1967; 71 (2): 181–191.
D. Achenbach TM, Rescoria LA. Manual for the ASEBA School-Age Forms and Profiles. Burlington (Vermont): University of Vermont Research Center for Children, Youth and Families, 2001.
E. Conners CK. The Conners Rating Scales, Austin, TX: PRO-ED, 1985.
F. Conners CK. Symptom patterns in hyperkinetic, neurotic and normal children. Child Dev 1970; 41: 667–82.
G. Schopler E, Reichler RJ, DeVillis RF, Daly K. Towards objective classification of childhood autism: Childhood Autism Rating Scale (CARS). J Autism Dev Disorders 1980; 10 (1): 91–103.
H. Goodman R. The Strengths and difficulties Questionnaire: A Research Note. J Child Psychol Psychiatry 1997; 38: 581–586.

References

[1] Patry G, Lyagoubi S, Tassinari CA. Subclinical electrical status epilepticus induced by sleep in children. Arch Neurol 1971;24:242-52.
[2] Tassinari CA, Rubboli G, Volpi L, et al. Electrical status epilepticus during slow sleep (ESES or CSWS) including acquired epileptic aphasia (Landau–Kleffner syndrome). Epileptic syndromes in infancy, childhood and adolescence. 4th edition. London: John Libbey; 2005. p. 295-314.
[3] Smith MC, Polkey CE. Landau Kleffner syndrome and CSWS. In: Engel Jr J, Pedley TA, editors. Epilepsy: A comprehensive textbook (2nd edition). Lippincott Williams and Wilkins: Philadelphia; 2008. p. 2429-38.
[4] Scheltens-deBoer Marjan. Guidelines for EEG in encephalopathy related to ESES/CSWS in children. Epilepsia 2009;50(Suppl. 7):13-7.
[5] Saltik S, Uluduz D, Cokar O, Demirbilek V, Dervent A. A clinical and EEG study on idiopathic partial epilepsies with evolution into ESES spectrum disorders. Epilepsia 2005;46(4):524-33.
[6] Kramer U, Sagi L, Goldberg-Stern H, Zelnik N, Nissenkorn A, Ben-Zeev B. Clinical spectrum and medical treatment of children with electrical status epilepticus in sleep (ESES). Epilepsia 2009;50(6):1517-24.
[7] Tononi G, Cirelli C. Sleep and synaptic homeostasis. Sleep Med Rev 2006;10:49-62.
[8] Tassinari CA, Rubboli G. Cognition and paroxysmal EEG activities: from a single spike to electrical status epilepticus during sleep. Epilepsia 2006;47(Suppl. 2):40-3.
[9] Tassinari CA, Cantalupo G, Rios-Pohl L, Giustina ED, Rubboli G. Encephalopathy with status epilepticus during sleep: "the Penelope syndrome". Epilepsia 2009;50(Suppl. 7):4-8.
[10] Lagae Lieven. Rational treatment options with AEDs and ketogenic diet in Landau–Kleffner syndrome: still waiting after all these years. Epilepsia 2009;50(Suppl. 7): 59-62.
[11] Buzatu M, Bulteau C, Altuzarra C, Dulac O, Van Bogaert P. Corticosteroids as treatment of epileptic syndromes with continuous spike–wave during slow-wave sleep. Epilepsia 2009;50(Suppl. 7):68-72.
[12] Sinclair DB, Synder TJ. Corticosteroids for the treatment of Landau–Kleffner syndrome and continuous spike wave discharge during sleep. Pediatr Neurol 2005;32:300-6.
[13] Arts FMW, Aersen F. Landau–Kleffner syndrome and CSWS syndrome: treatment with intravenous immunoglobulin. Epilepsia 2009;50(Suppl. 7):55-8.
[14] Morrell F, Whisler WW, Smith MC, et al. Landau–Kleffner syndrome. Treatment with subpial intracortical transaction. Brain 1995;118:1529-46.
[15] Cross HJ, Neville B. The surgical treatment of Landau–Kleffner syndrome. Epilepsia 2009;50(Suppl. 7):63-7.
[16] Inutsuka M, Kobayashi K, Oka M, Hattori J, Ohtsuka Y. Treatment of epilepsy with electrical status epilepticus during slow sleep and its related disorders. Brain Dev 2006;28:281-6.
[17] Hirtum-Das MV, Licht EA. Children with ESES: variability in the syndrome. Epilepsy Res 2006;70s:S248-58.