Continuous spikes and waves during sleep (CSWS): A Descriptive Clinical-epidemiological Profile from India

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INTRODUCTION

Continuous spikes and waves during sleep (CSWS) also known as Electrical Status Epilepticus in Sleep (**ESES**): Characteristic EEG changes, pharmaco-resistant seizures and cognitive regression.

EEG shows potentiation of epileptiform activity in the transition from wakefulness to sleep leading to near-continuous bilateral (or occasionally lateralized) slow spikes and waves that occupy more than 50% of non-REM sleep which is believed to contribute to cerebral dysfunction in children.

Landau Kleffner Syndrome (LKS) :

Epileptic encephalopathy with mainly language regression.

0.2%-0.6% - hospital prevalence of children with CSWS.

(1) ***Dormant stage*** (birth to onset of epilepsy)

(2) ***Prodromal stage*** (epilepsy onset to neuroregression)

(3) ***Acute stage*** (neuroregression to seizure freedom)

(4) ***Residual stage***.

OBJECTIVES

Present the epidemiological, etiological, clinical, EEG and pharmacological data in children with CSWS from a tertiary care centre of a developing country.

MATERIAL and METHODS

A retrospective study was carried out at the Epilepsy Clinic of a tertiary care institution in North India.

The profiles of all patients diagnosed with CSWS from 2012 to 2021 were reviewed using medical records.

Information obtained included patient demographics including the age of onset, gender, aetiology of epilepsy, semiology of seizures, comorbidities and treatment modalities which were noted down.

Descriptive statistics were used to summarize the demographics and clinical characteristics of the patients.

RESULTS

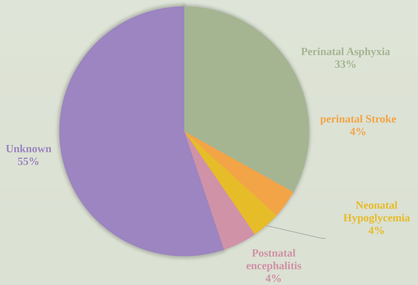
During the study period **24 children (3%)** were found to be diagnosed with CSWS at our centre after reviewing records of almost 850 epileptic children.

- Male preponderance (M: F = 5:1)
- Median age of onset : 3 (IQR 2.5-12) years
- Median age of presentation 6 (IQR 5-13) years

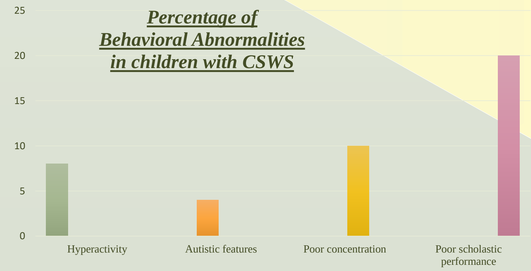
Seizure Characteristics

- Focal seizures with impaired awareness (50%, n=12)
- Nocturnal seizures (46%, n=11)
- Febrile episodes a/w initial seizures (33%, n=8)

Etiology of CSWS



Percentage of Behavioral Abnormalities in children with CSWS



All children (n = 24, 100%) received steroids with seizure control in all cases

- 66% (n =16) - Polytherapy combinations : valproate, clobazam, levetiracetam and topiramate
- 33% (n = 8) - high dose valproate monotherapy.

- Two children (8% each) each had progressed from Rolandic and Panayiotopoulos Epilepsy to develop CSWS
- Three children (12%) had classical regression of language milestones constituting the LKS spectrum of CSWS.

CONCLUSION

CSWS is a childhood epileptic-developmental encephalopathy that represents the most severe end of the seizure susceptibility syndrome requiring prompt diagnosis and aggressive treatment.

The aetiology of CSWS is unknown, but early developmental lesions involving the thalamus especially play a major role in around half of the cases.

Novel genetic mutations are being detected as a cause which may provide insight into its pathophysiology and subsequent treatment strategies.

High-dose benzodiazepines along with Polytherapy with combinations of valproate, ethosuximide, levetiracetam, sulthiame or lamotrigine, and corticosteroids is frequently used. Epilepsy surgery can be considered in a few selected patients.

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