Clinical Spectrum and Treatment Outcome of 95 Children with Continuous Spikes and Waves during Sleep (CSWS)
Eur J Paediatr Neurol. 2020

Background: Continuous spikes and waves during sleep (CSWS) is an age dependent epileptic encephalopathy characterized by generalized epileptiform activity during Non-REM sleep and neurocognitive dysfunction. EEG shows continuous slow spike waves with a frequency of 1.5-3 Hz during NREM sleep. Cognitive dysfunction include aphasia, attention, learning deficits and motor delay. Causes and outcome are diverse, and treatment is mainly empirical.

ACADEMIC P.E.A.R.L.S

Pediatric Evidence And Research Learning Snippet

Clinical Spectrum & Treatment Outcome of 95 Children with Continuous Spikes & Waves during Sleep (CSWS)

Methodology:

- Retrospective descriptive analysis conducted at University Hospital of Heidelberg, Germany, from January 1998 and December 2018. Children with confirmed diagnosis of CSWS were included.
- Cases were divided in two group: CSWS with a Spike Wave Index (SWI) >85% and Near-CSWS with SWI 40- 85%.

Results:

- Median age at diagnosis 5.6 years (range 2 months-13 years). 80% children had pre-existing developmental disorder. 63% had focal seizures, 63% had bilateral tonic-clonic seizures and 10% patients had no clinical apparent seizures
- Etiology- 43% structural and metabolic etiology, 17% genetic and 39% unknown
- EEG: 59% complete CSWS (SWI>85%), 30% near CSWS (SWI 60-85%), and 11% had SWI of 40-60%
- Treatment: 23 different modalities used, including ASM, steroids and epilepsy surgery.
- On an average individual patient received 4.8 different treatment regimens
- Valproate and levetiracetam lead to reduction in SWI in minority of subjects
- Phenobarbitone, vigabatrin and clobazam did not show any improvement in SWI in EEG
- In 70% children treated with steroids had improvement in SWI and cognition
- Epilepsy surgery in 5 cases lead to improvement in seizure frequency and cognition

TREATMENT	EEG IMPROVEMENT[A] (%, 95%CI)	SEIZURE IMPROVEMENT[B] (%, 95%CI)	COG. IMPROVEMENT (%, 95%CI)
TOTAL	63/345 (18.3, 6-41)	186/356 (52.2, 48-59)	100/333 (30.0, 25-35)
STEROIDS	**15/36 (41.7, 27-58)****	26/37 (70.3, 59-80)	24/36 (66.7, 50-80)
CAI	22/96 (22.9, 16-32)	56/98 (57.1, 44-69)	26/89 (29.2, 21-39)
NA-CB	6/55 (10.9, 5-22)	22/57 (38.6, 25-54)	13/53 (24.5, 15-38)
BENZO	0/19 (0, 0-17)	5/19 (26.3, 13-45)	1/17 (5.9, 1-27)
VPA	9/51 (17.6, 10-30)	26/51 (51.0, 38-64)	11/52 (21.2, 12-34)
CA-CB	4/23 (17.4, 7-37)	11/23 (47.8, 34-62)	6/22 (27.3, 12-32)
VGB	0/7 (0, 0-35)	3/7 (42.9, 29-58)	3/6 (50, 19-81)
LEV	4/45 (8.9, 4-21)	24/47 (51.1, 37-65)	10/45 (22.2, 12-36)
SURGERY	**3/5 (60, 23-88)***	4/5 (80, 70-87)	4/5 (80, 38-96)
PB	0/8 (0, 0-32)	9/12 (75, 64-83)	2/8 (25, 7-59)

[A]: including a SWI-reduction >25%, [B]: moderate-market reduction of seizure frequency
CAI: Carbonic anhydrase-Inhibitors, Na-CB: Na^+-Channel Blocker, Benzo: Benzodiazepine, VPA: Valproic acid, Ca-CB: Ca^{2+}-Channel Blockers, VGB: Vigabatrin, LEV: Levetiracetam, PB: Phenobarbital
*: $p < .05$, **: $p < .01$

Conclusion:

- Children with identifiable etiology had an earlier onset of CSWS. A significant increase in genetic etiologies of CSWS is seen during the last decade. Significant better treatment effects were found for the treatment with steroids or neurosurgery with regards to the reduction of SWI and seizure frequency

EXPERT COMMENT

"CSWS is an age dependent epileptic encephalopathy with characteristic EEG & cognitive dysfunction. Anti-seizure medications do not have effect on EEG spike wave index & cognition in children with CSWS. Children treated with steroids reported reduction in seizure frequency, spike wave index (SWI) & cognitive improvement. Prompt treatment with steroids and follow up EEG with SWI monitoring should be used to guide therapy in children with CSWS"

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Reference

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