Practice patterns in electrical status epilepticus in sleep: A survey study

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Background and Aims

- Definitions for "electrical status epilepticus in sleep" (ESES) and "continuous spike and wave in slow wave sleep" (CSWS) are variable and treatments lack agreement[1,2]
- The scope of current practice in ESES has not been recently evaluated
- For this project, our definitions:
  - ESES = electroencephalographic pattern
  - CSWS = electroclinical syndrome with ESES on EEG and regression/stagnation
- Aims: Amongst practicing child neurology and pediatric epilepsy providers:
  - Assess variability and clinical practice in the diagnosis and treatment of sleep potentiated discharges, ESES, CSWS, etc.
  - Evaluate provider definitions of these terms

Methods

- 22-item survey designed by members of the ESES special interest group of the Pediatric Epilepsy Research Consortium (PERC)
  - Assessing definitions, diagnosis, and management of children with increased activation of spikes in sleep
- Distributed electronically between July 2020 and May 2021 to members of PERC, Child Neurology Society and American Epilepsy Society

Results

Table 1: Terminology, definitions, diagnosis

Are ESES and CSWS synonyms?		**N=252, n (%)**
Yes		78 (31%)
No		154 (61%)
Unsure		20 (8%)
Which are required to diagnose: ESES? CSWS?		**N=225, n (%)**
	ESES	**CSWS**
Excessive spike waves in sleep	221 (98%)	216 (96%)
Behavior/cognitive impairment	96 (43%)	140 (62%)
Imaging abnormalities	7 (3%)	17 (7.5%)
Genetic abnormalities	15 (7%)	18 (8%)
Minimum spikes to diagnose ESES or CSWS?		**N=228, n (%)**
I do not have a minimum cut-off value		15 (6.5%)
Spike waves in at least 50% of sleep		75 (33%)
Spike waves in at least 85% of sleep		127 (56%)
Other		11 (4.5%)
Metric used to diagnose ESES or CSWS?		**N=226, n (%)**
Spike wave index (SWI): Seconds w/ Spikes / Total Seconds		138 (61%)
Spike counts: # Spikes/Specific Time		26 (12%)
Visual recognition ("I know it when I see it")		39 (17%)
Other		23 (10%)

Figure 1: Practice focus

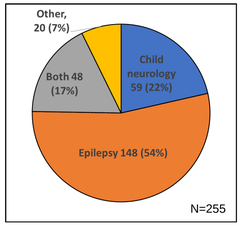

Figure 2: Standardized care within a practice

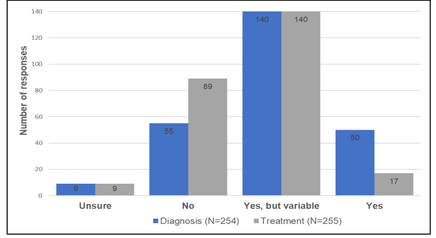

Table 2: Clinical vignettes and responses

Each case involves a 7-year-old girl with frequent spike waves in sleep:	**Would you treat and when (degree activation)?**		**How would you treat?**	
• New-onset language difficulty • No longer recognizes familiar sounds • Does not respond to parents' voices		N=207, n (%)		N=207, n (%)
	Never	0	Benzodiazepines	108 (52%)
	10-25%	17 (8%)	ASM	45 (22%)
	25-50%	25 (12%)	Steroids	44 (21%)
	50-85%	91 (44%)	Ketogenic diet	0
	>85%	20 (10%)	Surgery	0
	Always	54 (26%)	Other	10 (5%)
• History of hypoxic-ischemic encephalopathy and cerebral palsy with developmental stagnation • Undergoes a sleep study which includes EEG		N=205, n (%)		N=202, n (%)
	Never	15 (7%)	Benzodiazepines	96 (47.5%)
	10-25%	4 (2%)	ASM	81 (40%)
	25-50%	18 (9%)	Steroids	16 (8%)
	50-85%	72 (35%)	Ketogenic diet	0
	>85%	83 (40%)	Surgery	0
	Always	13 (6%)	Other	9 (4.5%)
• Long-standing diagnosis of autism without regression • Prolonged EEG for hand-flapping episodes		N=206, n (%)		N=187, n (%)
	Never	44 (21%)	Benzodiazepines	94 (50%)
	10-25%	6 (3%)	ASM	65 (35%)
	25-50%	9 (4%)	Steroids	14 (7.5%)
	50-85%	61 (30%)	Ketogenic diet	0
	>85%	83 (40%)	Surgery	0
	Always	3 (1.5%)	Other	14 (7.5%)
• Developmentally normal girl with ADHD • Undergoes a sleep study for snoring		N=207, n (%)		N=160, n (%)
	Never	90 (44%)	Benzodiazepines	68 (42.5%)
	10-25%	5 (2%)	ASM	65 (40.5%)
	25-50%	3 (1.5%)	Steroids	11 (7%)
	50-85%	61 (18%)	Ketogenic diet	0
	>85%	70 (34%)	Surgery	0
	Always	1 (0.5%)	Other	16 (10%)

Discussion

- Nearly all (96-98%) agreed that excessive spikes in sleep are required for a diagnosis of both ESES and CSWS
- All would treat a child with regression and nearly all (93%) a child with developmental stagnation in the setting of increased activation of spikes in sleep
- Benzodiazepines were the preferred treatment followed by ASMs, in contrast to prior studies[3]
- Steroids were considered primarily in the scenario with regression

Conclusions

- Variability in diagnosis and treatment of ESES-related syndromes remains high
- Ongoing multicenter collaboration, prospective trials, and expert consensus are needed for standardized classification, diagnosis, and treatment for ESES-related disorders

References

1. Sanchez-Fernandez A, Chapman KE, Peters JM, et al. The tower of Babel: Survey on concepts and terminology in electrical status epilepticus in sleep and continuous spikes and waves during sleep in North America. Epilepsia. 2013 Apr;54(4):741-50.
2. Sanchez-Fernandez A, Chapman KE, Peters JM, et al. Treatment for continuous spikes and waves during sleep (CSWS): Survey on treatment choices in North America. Epilepsia. 2014 Jul;55(7):1099-108.
3. van den Munckhof B, van Dee V, Sagi L, Caraballo RH, et al. Treatment of electrical status epilepticus in sleep: A pooled analysis of 575 cases. Epilepsia. 2015 Nov;56(11):1738-46.