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Grading of Improvement in Hypsarrhythmia with Standard Epileptic Spasms Treatment at a Large Pediatric Tertiary Care Center

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Background

- Epileptic spasm often entails the clinical spasms and a characteristic electroencephalogram (EEG) abnormality often called hypsarrhythmia or its variants.
- The main goal of epileptic spasm treatment with standard therapy is to suppress clinical spasms and abolish the hypsarrhythmia and its variant EEG pattern.
- This interictal EEG pattern frequently heralds developmental regression. The elimination of hypsarrhythmia is a principal goal of therapy and a key outcome measure in clinical trials.
- There have been several studies in the interpretation or grading of hypsarrhythmia,[1,2] but none in the grading of electrographic improvement with the standard treatment and its effects on the outcome, mainly in terms of remission versus relapse.

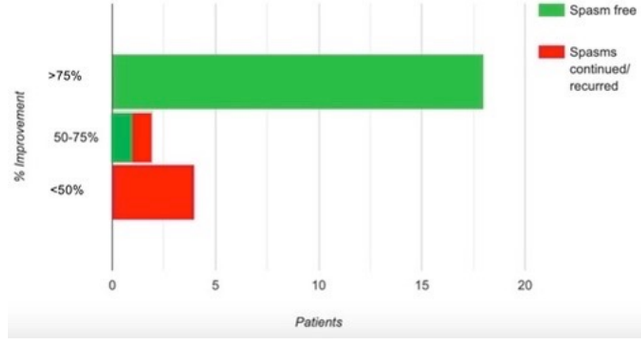
Methods

- This is a retrospective chart review examining several EEGs before and after treatment using a standard scoring system [3] for a follow up period of at least 6 months to 1 year.
- We graded EEGs based on a standard scoring system on pretreatment EEG (at the time of diagnosis) and post-treatment EEG changes (1-2wks, 2-3 months, or 6-12 months) in patients with epileptic spasms who received standard hormonal (ACTH or prednisone) or Vigabatrin therapy.
- We selected patients with common etiologies (Cryptogenic, Down Syndrome, and Tuberous Sclerosis) and determined the EEG features predicting remission or relapse of epileptic spasms.

Kramer Scale

A. Disorganization	
Good gradient and synchrony (normal for age)	0
Partially formed gradient with some synchrony	1
No gradient, some synchrony of background	2
Chaos, no synchrony, no gradient	3
B. Diffuse delta activity	
<50%	0
≥50% but <75%	1
≥75% but <100%	2
100%	3
C. Voltage	
<120 μV	0
120–200 μV	1
200–500 μV	2
>500 μV	3
D. Spikes and sharp waves	
No spikes or sharp waves	0
Spikes at a frequency of ≤1/5 sec	1
Spikes at a frequency of 1/5 sec–1/sec	2
Spikes at a frequency of ≥1/sec	3
E. Other items	
Electrodecremental discharges	1
Burst suppression in sleep	1
Absence of normal sleep pattern	1
Relative normalization	1

Results Data



Results

- Of 24 patients with epileptic spasms, 18 had EEG improvement of over 75% in the Kramer Scale score with resolution of spasms. Four had less than 50% improvement with refractory or recurrent spasms. Two patients had improvement between 50 and 75%. One had resolution of spasms, while the other had recurrent spasms.
- All the patients were treated with different treatment options as 10 received only ACTH, 9 received both ACTH & Vigabatrin, 4 received only Vigabatrin, and 1 received only prednisone and Vigabatrin.

Conclusions

- Patients with sustained improvement in serial EEGs over 3-6 months of about >75% compared to pretreatment EEG were associated with spasms remission.
- Patients in whom the improvements were less than 50% continued to have ongoing spasms or spasm recurrence. These results highlight the need for the level of aggressiveness and close follow-up depending on the degree of EEG improvements with standard therapy.

References

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