



# Effect of Long-term ACTH therapy on refractory infantile epileptic spasms syndrome

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## Introduction

ACTH therapy is recognized as most effective treatment for infantile epileptic spasms syndrome (IESS). Ordinarily ACTH therapy (short-term ACTH; ST-ACTH) treats IESS by injecting ACTH over a short period of 2 to 4 weeks. However, it is known that some cases repeatedly relapse after the treatment and become refractory. Long-term ACTH therapy (LT-ACTH) has been attempted for such refractory cases, but the reported number of cases is small with limited information. In this study, we reported cases in which LT-ACTH was performed for IESS that repeatedly recurred after ST-ACTH.

## Criteria for administration of LT-ACTH therapy

- The subjects for LT-ACTH are cases that meet the following criteria;
1. IESS developed during infancy,
  2. Epileptic spasm (ES) is temporarily suppressed with ST-ACTH, followed by rapid recurrence,
  3. ES are not suppressed by poly-antiseizure medications,
  4. ES restricts daily life.
- The LT-ACTH protocol consisted of two weeks of daily injections followed by weekly injections.
- Two cases were matched to this study.

## Results

Case 1 is an 8-year-old girl, who had status epilepticus at 10 months of age, and her interictal electroencephalogram revealed hypsarrhythmia. She began to have spasms in clusters at 19 months. Although multi-anticonvulsant therapy and ST-ACTH therapy has been administrated, the efficacy was limited with more than 100 ES clusters 3 to 4 times a week.

Age 1 2 3 4 5 6 7 8

VPA ———— CLB ———— LTG ————  
ESM ———— KD ———— PER ———— VB6  
ST-ACTH ST-ACTH LT-ACTH

Since ES is transiently suppressed by ST-ACTH and ES is suppressed during pneumonia, we decided to try LT-ACTH therapy. After the introduction of LT-ACTH, seizures have been completely suppressed and her development has been slowly progressed.

Case 2 is a 13-year-old boy, who developed IESS with spasms in cluster at 7 months of age. Although his seizures were suppressed with ST-ACTH, the seizures continued to recur, and despite multi-anticonvulsant therapy and repeated ST-ACTH treatments, the seizures remained intractable.

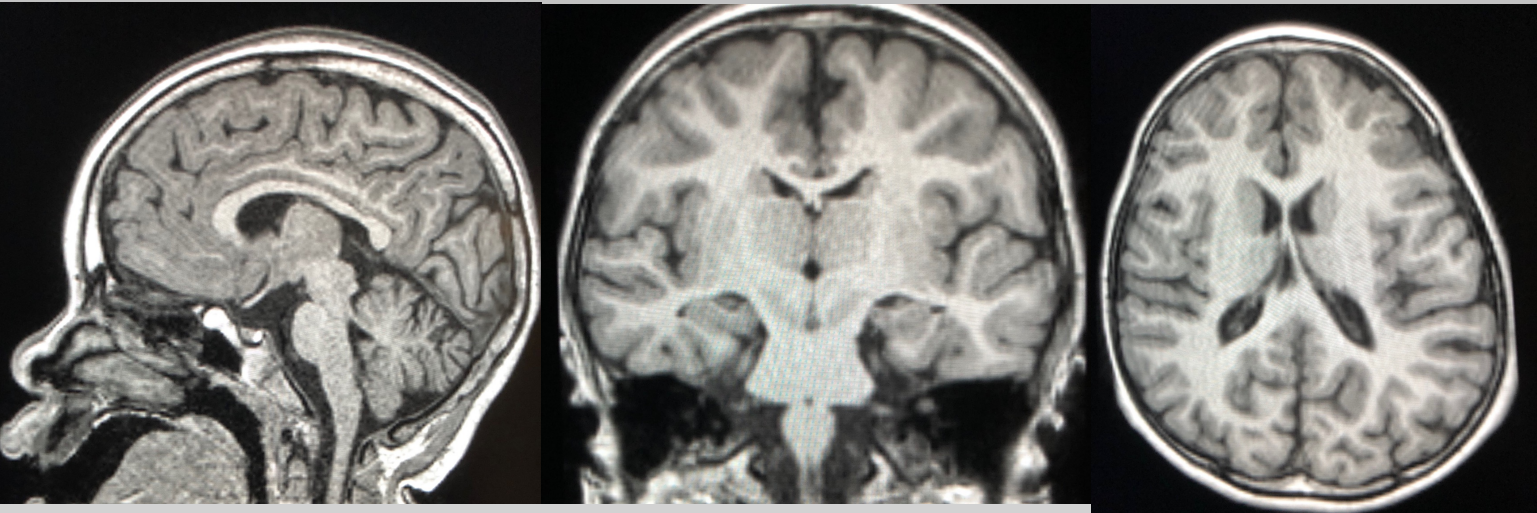
Age 1 2 3 4 5 6 7 8 9 10 11 12 13

VB6 ZNS ———— VPA ————  
VPA LEV NZP ———— CLB ———— SLM  
LTG VGB LCM  
ST-ACTH ST-ACTH ST-ACTH LT-ACTH

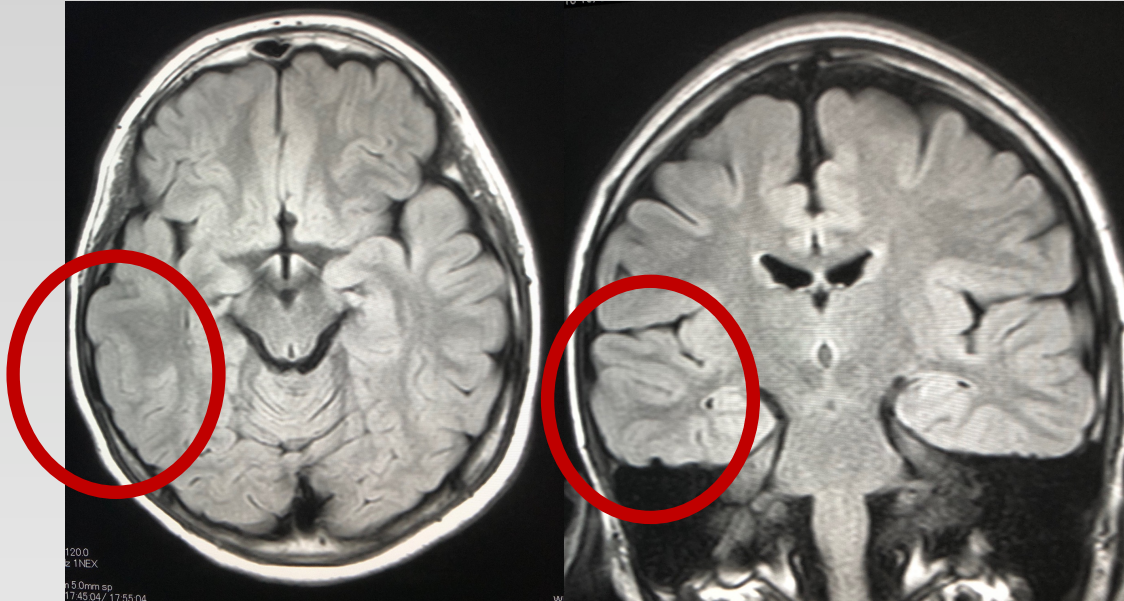
Although short-term ES suppression was obtained with ST-ACTH, the seizures soon recurred. After the introduction of LT-ACTH, the daily seizures decreased to weekly, but complete suppression was not achieved.

## Brain MRI

Case 1



Case 2



## Conclusion

In both cases, seizures were suppressed or reduced due to the effects of LT-ACTH, no serious side effects were observed, and the quality of daily life improved.

## Reference

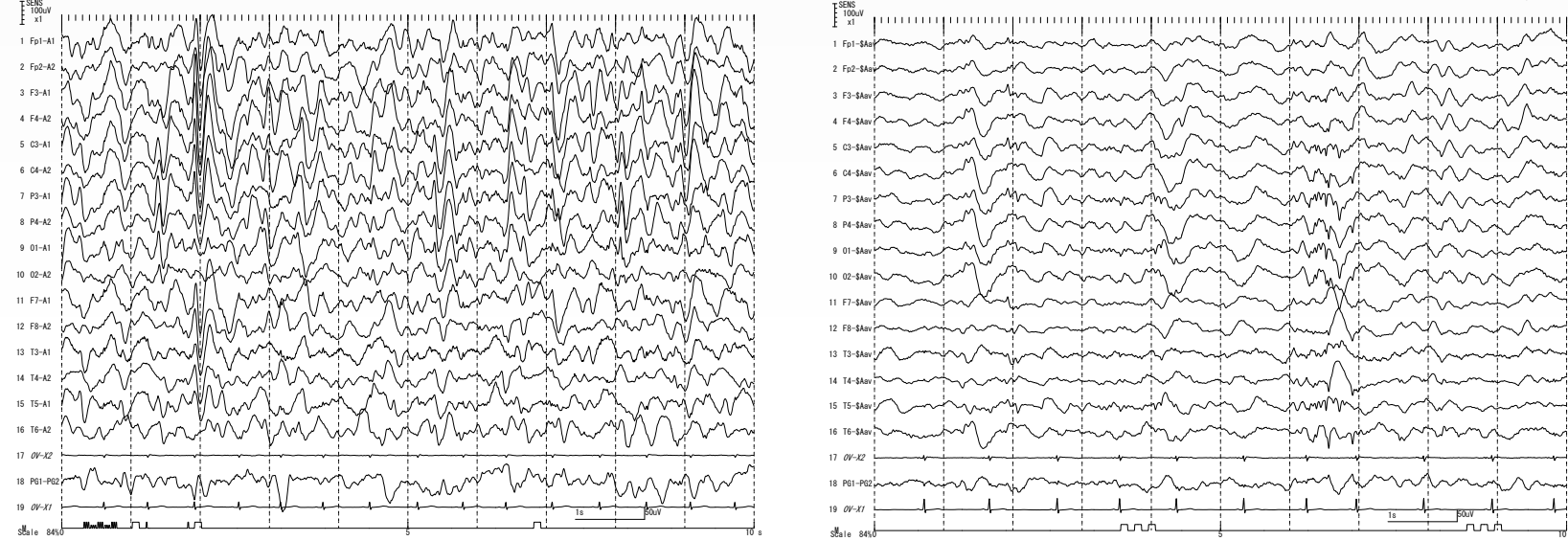
- Efficacy of long term weekly ACTH therapy for intractable epilepsy. Inui et al. Brain Dev. 2015
- Long-term weekly ACTH therapy for relapsed West syndrome in tuberous sclerosis complex: A case report. Nakata et. al. Brain Dev. 2016

## Contact

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## Chronological EEG findings of LT-ACTH period

Case 1



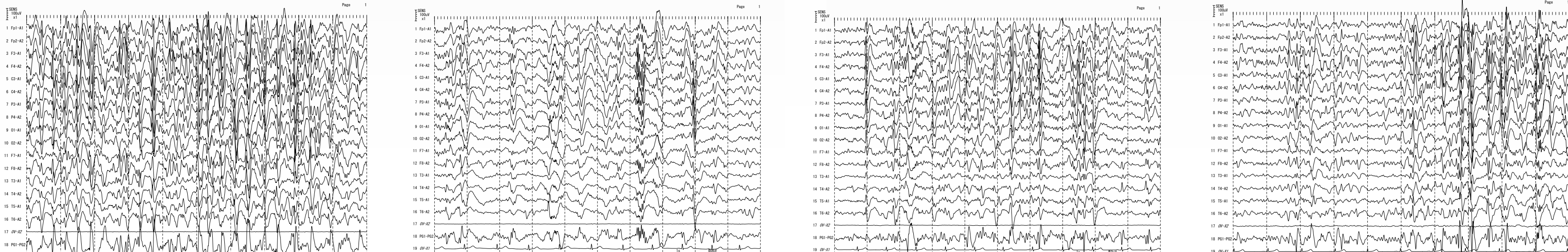
Before

2 weeks

1 month

1 year

Case 2



Before

2 weeks

1 month

1 year