

Case Report

Occurrence of bilaterally independent epileptic spasms after a corpus callosotomy in West syndrome

Katsuhiro Kobayashi^{a,*}, Fumika Endoh^a, Yoshihiro Toda^{a,b}, Makio Oka^a,
Hiroshi Baba^c, Yoko Ohtsuka^{a,d}, Harumi Yoshinaga^a

^a Department of Child Neurology, Okayama University Graduate School of Medicine, Dentistry and Pharmaceutical Sciences and Okayama University Hospital, Shikatacho 2-chome 5-1, Kita-ku, Okayama, Japan

^b Department of Pediatrics, School of Medicine, University of Tokushima, Tokushima, Japan

^c Epilepsy Center, Nagasaki Medical Center, Kubara 2-1001-1, Omura, Nagasaki, Japan

^d Asahigawaso Rehabilitation and Medical Center, Japan

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Abstract

We report a patient with intractable West syndrome whose epileptic spasms (ESs) were initially bilaterally synchronous, as is typical; after a complete corpus callosotomy, however, bilaterally independent ESs originated in either hemisphere. Activity of probable cortical origin associated with ESs was detected by observing ictal gamma oscillations. Brain MRI revealed no structural abnormality before surgery. This case suggests that ESs with a hemispheric origin may appear generalized because of synchronizing effects in the corpus callosum in some patients.

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1. Introduction

Epileptic spasms (ESs), or infantile spasms, characterize West syndrome in association with hypsarrhythmia in electroencephalogram (EEG). The generation of ESs is traditionally considered to involve the brain subcortical structures, particularly the brainstem, and pathological interaction with the cortex. However, many reports of successful ES treatment by cortical resection indicate that ESs originate from the cortex in at least

some patients [1,2]. It is unknown whether the ESs are focal or generalized [3].

We previously described a patient with Aicardi syndrome with complete agenesis of the corpus callosum, who exhibited bilaterally independent ESs with no mutual interaction; this suggested that the cortex plays a role in driving the brainstem to generate ESs [4]. This case was considered exceptional because of the presence of large-scale congenital brain malformations. However, we found that bilaterally independent ESs can occur after a callosotomy, which suggests that ESs with a hemispheric origin may appear generalized in some patients because of the synchronizing effects of the corpus callosum.

* Corresponding author at: Department of Child Neurology, Okayama University Hospital, Shikatacho 2-chome 5-1, Kita-ku, Okayama 700-8558, Japan. Tel.: +81 86 235 7372; fax: +81 86 235 7377.

E-mail address: k_koba@md.okayama-u.ac.jp (K. Kobayashi).

2. Case report

The female patient was born at 40 weeks gestation following an uneventful pregnancy; she weighed 2858 g and had no asphyxia. There were no abnormalities in her family history. At 4 months of age, ESs began in clusters with associated deterioration of head control.

The patient was admitted to Okayama University Hospital at 5 months of age. Mild hypotonia was observed. No etiological abnormalities were detected by a battery of tests including blood chemistry, blood gas analysis, cerebrospinal fluid, amino acid analysis, urinary organic acid analysis, and chromosomal analysis. Brain MRI revealed no structural brain abnormalities (Supplementary Fig. 1A). Hypsarrhythmia was typical and showed bilaterally synchronous grouping of discharges during sleep. She had about five ES clusters per day under the initial treatment with pyridoxal phosphate, and her ESs showed no asymmetric features either in symptomatology or on the ictal EEG. The patient was diagnosed with West syndrome, and treated with a protocol in which low-dose tetracosactide Zn (synthetic adrenocorticotrophic hormone (ACTH)) at a dose of 0.005 mg/kg/day was initially administered for 2 weeks, and then a higher dose of ACTH (0.025 mg/kg/day) was administered for an additional 2 weeks.

After this ACTH treatment, however, ES suppression lasted for only 1 month. The EEG also worsened, with hypsarrhythmia reappearing at around 3 months after ACTH treatment (Fig. 1A). Relapsed ESs occurred in clusters or in isolation with no asymmetric features and were intractable despite treatment with sodium

valproate, clonazepam and zonisamide (ictal EEG in Fig. 1B; corresponding movie in Video 1). Time–frequency analysis of this ictal EEG data showed bilateral gamma activities (Supplementary Fig. 2).

A complete corpus callosotomy was performed at Nagasaki Medical Center when the patient was 14 months of age (Supplementary Fig. 1B). Her ESs persisted after this operation, though they changed into bilaterally independent isolated spasms. Hypsarrhythmia remained in the EEG, but the discharge grouping during sleep became bilaterally asynchronous (Fig. 2A). Post-operative ESs involving either hemisphere occurred almost continuously at intervals of several minutes during wakefulness. A total of 137 ESs were captured by video-EEG monitoring (Table 1): ESs were considered to have a left and right hemispheric origin with associated unilateral gamma activities in 67 (48.9%) (Fig. 2B and Video 2, Supplementary Fig. 3) and 61 (44.5%) (Fig. 2C and Video 3, Supplementary Fig. 4), respectively.

The patient was admitted again to Okayama University Hospital at 18 months of age. Her spasms were suppressed using ACTH (0.025 mg/kg/day) treatment that continued for 6 weeks, followed by oral prednisolone (1 mg/kg alternate days) that lasted for 1 month. The patient has been seizure-free since then, although her EEG shows many multifocal spikes. She is now 5 years and 2 months old and is currently being treated with topiramate and levetiracetam. She has attained head control and visual following, but is hypotonic and cannot sit alone. She does not understand language. Vigabatrin is officially unavailable in Japan and cannot be administered to her.

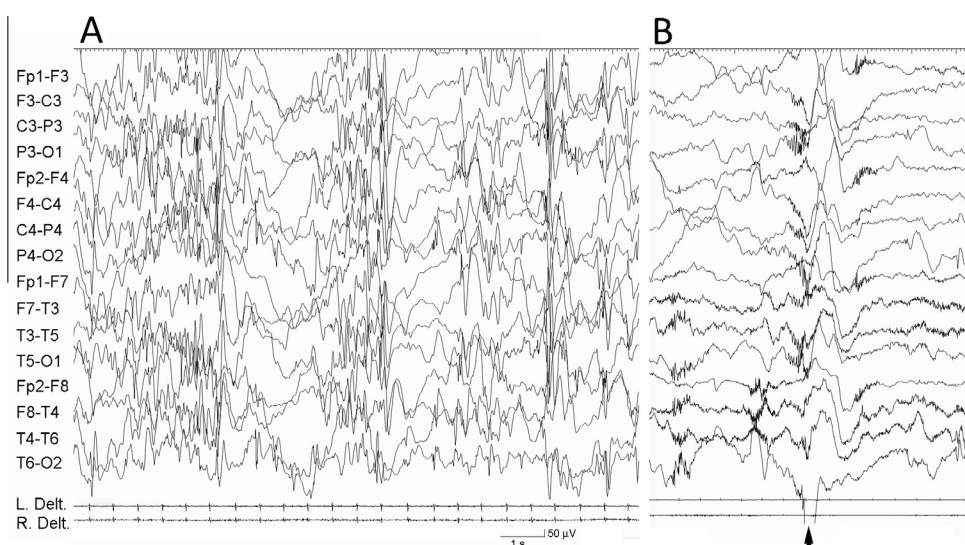


Fig. 1. Electroencephalogram (EEG) recorded at 10 months of age, before the corpus callosotomy. (A) Interictal sleep EEG showing hypsarrhythmia with a bilaterally symmetric grouping of discharges. (B) The ictal EEG of a bilaterally symmetric epileptic spasm (ES) in Video 1 showing diffuse bilateral high-amplitude slow waves and superimposed fast activity. The corresponding time–frequency analysis is shown in Supplementary Fig. 2.

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