

Staged Total Callosotomy for Lennox-Gastaut Syndrome: A Case Report

Eun Hye Lee, MD¹, Mi-Sun Yum, MD², Seok-Ho Hong, MD³, Jeong-Kyo Lee, MD³,
Su Jeong You, MD⁴, Tae-Sung Ko, MD²

¹Department of Pediatrics, College of Medicine, Kyung Hee University; ²Department of Pediatrics, Asan Medical Center, Children's Hospital, University of Ulsan College of Medicine; ³Department of Neurosurgery, Asan Medical Center, University of Ulsan College of Medicine; ⁴Department of Pediatrics, Epilepsy Center, Sanggye Paik Hospital, Inje University College of Medicine, Seoul, Korea

Case Report

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Corresponding author: Tae-Sung Ko
Department of Pediatrics, Asan Medical Center, University of Ulsan College of Medicine, 388-1 Pungnap-dong, Songpa-gu, Seoul 138-736, Korea
Tel. +82-2-473-3725
Fax. +82-2-3010-3388
E-mail: tsko@amc.seoul.kr

We report a case with Lennox-Gastaut syndrome (LGS) who underwent staged total callosotomy with a favorable outcome. A 6-year-old boy began having myoclonic seizures at the age of 8 months and was diagnosed with LGS when he was 27 months old. Various antiepileptic drugs and a ketogenic diet failed to control his multiple types of seizures. At the age of 36 months, he underwent corpus callosotomy and achieved an immediate, post-operative seizure free state. However, 3 months later, various seizures relapsed and were refractory to additional vagus nerve stimulation. Remaining callosal fibers in the splenium noted on post-operative diffusion tensor imaging made us to perform a second operation, total callosotomy. The patient finally achieved a seizure-free state with electroencephalography (EEG) normalization noted after the staged total callosotomy. (2011;1:71-73)

Key words: Lennox-Gastaut syndrome; Callosotomy

Lennox-Gastaut syndrome (LGS) is a difficult-to treat type of epilepsy characterized by multiple types of seizures, generalized slow spike and wave activities or generalized paroxysmal fast activities in EEG, and progressive cognitive deterioration.¹ Because medical treatment usually fails to achieve satisfactory seizure control, multimodal management is often required. Surgery for this condition includes callosotomy, vagus nerve stimulation or resective surgeries for limited cases with focal lesions.^{2,3} Among them, corpus callosotomy is effective especially for treating drop attacks which are the most notorious type of seizures in LGS.^{4,5} In some medical centers, anterior callosotomy is often performed in order to avoid the complications of a disconnection syndrome, however, approximately 30% of these patients experience relapse of their seizures.⁶ In those cases, a second total callosotomy can be a useful treatment option. We report here the case of a patient with LGS who underwent staged total callosotomy, after which he achieved a seizure-free state.

Case Report

A 6 year-old boy was born at 34 weeks' gestation to healthy

parents with birth weight of 3,540 g after an uneventful pregnancy. He had no family history of epilepsy or focal neurologic sign, but he had delayed development in fine motor and language areas since before 12 months of age. His first seizures, myoclonic seizures, occurred at the age of 8 months, and he was started on antiepileptic drugs (AEDs). At the time of his admission to our hospital at the age of 27 months, he was suffering from multiple types of seizures including drop attacks, generalized tonic seizures, and atypical absence seizures. His EEG showed generalized slow spike and wave discharges with multifocal spikes (Figure 1), and he was diagnosed as having LGS. Brain magnetic resonance imaging (MRI) showed no abnormalities. Despite various antiepileptic drugs (valproate, topiramate, zonisamide, and clobazam) and a ketogenic diet, his seizures remained uncontrolled. Long term video EEG monitoring for 48 hours showed a dozen seizures a day. The seizure semiology consisted of suddenly dropping his head with losing his tone, thus suggesting myoclonic-atonic seizures. Ictal EEG showed diffuse generalized bursts of polyspike discharges with very brief electromyographic activities. His social quotient was reported to be 27. At the age of 36 months, he underwent anterior 2/3 corpus callosotomy. He achieved a post-operative seizure-free state for 3

seizures in 35%, whereas total callosotomy was effective in 77%. Sunaga *et al.*⁶ also demonstrated that control of drop attacks was achieved in 76% of their patients with total callosotomy and in 54% of those with anterior callosotomy. They also reported that patients with anterior callosotomy had a higher relapse rate than patients with total callosotomy (31% vs. 7%). However, Mamelak *et al.*¹¹ reported that anterior 1/2-2/3 callosotomy is nearly as effective as extensive total callosotomy for reducing both drop attacks and generalized tonic clonic seizures. They proposed that the extent of callosotomy is not an important factor if at least 50-65% of the callosum is divided.

Considering the seizure outcome and the risk of complications, some centers prefer to perform an anterior callosotomy which first spares the splenium, and then to perform total callosotomy as a second step if the first surgery is not successful.^{5,9,11} In a report by Spencer *et al.*¹⁰ for 14 patients who failed to improve after anterior callosotomy and then underwent further total callosotomy, dramatic responses were noted for all seizure types. In Korea, Kim *et al.*⁹ have reported that a second total callosotomy performed in 4 patients who did not have clinical improvement after their first surgery, allowed 3 of them to become seizure-free and allowed >75% seizure reduction in the other patient. We suggest that staged total callosotomy is a valid treatment option for patients with LGS suffering from drop attacks.

It is interesting that the EEG performed at 4 months post-operatively showed completely normal findings in our report. This finding supports the hypothesis that corpus callosum has a role in generating epileptiform discharges. Matsuo *et al.*¹² performed a quantitative analysis of EEGs in patients who underwent corpus callosotomy; they found that corpus callosotomy decreased the total amount of spike discharge and the duration of burst activity in addition to disrupting bilateral synchrony. They suggested that corpus callosum not only transfers epileptic activities but also has a mutually facilitatory effect that enhances the epileptogenic state of both hemispheres. However, the precise role of the corpus callosum in epileptogenesis remains to be determined.

Here we report the case of a patient who experienced relapse of seizures after anterior callosotomy but finally obtained freedom from seizures with EEG normalization after the second total callosotomy.

We, therefore, suggest that although surgery can initially be limited to the anterior 2/3 of the corpus callosum, if the first operation is unsuccessful, further total callosotomy should be considered.

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