Corpus callosotomy for intractable seizure associated with Lennox-Gastaut syndrome

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Introduction

The Lennox-Gastaut syndrome is a severe childhood-onset epileptic encephalopathy characterized by several seizure types, diffuse slow spike-wave discharges and generalized fast rhythms on electroencephalogram (EEG), cognitive impairment and personality disorder.

Beaumanoir A. et al. 2005

Because of the seizures in patients with LGS are multiple and usually prognostically unfavorable to conventional antiepileptic medications, nonpharmacological therapeutic approaches, such as ketogenic diet, vagus nerve stimulation and corpus callosotomy are becoming a more widely used.
Corpus callosotomy is a palliative disconnection procedure that consists of microsurgically sectioning the corpus callosum to disrupt the bilateral propagation of epileptic discharges, thereby decreasing the frequency and severity of seizures.

Oguni H. et al. 1991

The overall goal of the current study is to evaluate the long-term surgical effect following corpus callosotomy in patients suffering from Lennox-Gastaut syndrome with special emphasis on different seizure types.
Methods

A total 107 patients underwent corpus callosotomy at the Severance Children’s Hospital between October 2003 and July 2011. The medical records were retrospectively reviewed for all patients. In this study were included 79 patients who underwent corpus callosotomy for intractable seizure associated with LGS. All patients failed multiple appropriate trials of antiepileptic medication and had no clear lateralizing data indicating the seizure onset hemisphere. Seizure frequency and severity were causing physiological injuries from falls which significantly impact on the quality of life.
Results

<table>
<thead>
<tr>
<th>Variable</th>
<th>Value</th>
<th>Gender, male</th>
<th>Female</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>49 (62%)</td>
<td>30 (48%)</td>
</tr>
<tr>
<td>Age at seizure onset</td>
<td>Mean ±SD</td>
<td>2.5 ±2.7</td>
<td>Range 0.1-9</td>
</tr>
<tr>
<td>Age at surgery</td>
<td>Mean ±SD</td>
<td>8.9 ±4.5</td>
<td>Range 2.8-20.5</td>
</tr>
<tr>
<td>Duration of epilepsy</td>
<td>Mean ±SD</td>
<td>6.4 ±3.9</td>
<td>Range 0.9-17.4</td>
</tr>
<tr>
<td>Follow-up period</td>
<td>Mean ±SD</td>
<td>3.4 ±2.5</td>
<td>Range 0.5-8.4</td>
</tr>
</tbody>
</table>

- Atonic seizure: 3.80%
- Generalized tonic Sz.: 12.66%
- Myoclonic seizure: 11.39%
- Epileptic spasms: 39.24%
- Atypical Absence: 32.91%
## Results

Postoperative seizure outcome according to Engel’s classification

<table>
<thead>
<tr>
<th>Follow-up period</th>
<th>6 months</th>
<th>1 year</th>
<th>2 years</th>
<th>3 years</th>
<th>4 years</th>
<th>≥ 5 years</th>
</tr>
</thead>
<tbody>
<tr>
<td>Engel Class I</td>
<td>30 (38%)</td>
<td>19 (27.1%)</td>
<td>19 (32.8%)</td>
<td>15 (30%)</td>
<td>11 (33.3%)</td>
<td>5 (18.5%)</td>
</tr>
<tr>
<td>Engel Class II</td>
<td>14 (17.7%)</td>
<td>17 (24.3%)</td>
<td>15 (25.9%)</td>
<td>16 (32%)</td>
<td>9 (27.3%)</td>
<td>8 (29.6%)</td>
</tr>
<tr>
<td>Engel Class III</td>
<td>17 (21.5%)</td>
<td>13 (18.6%)</td>
<td>10 (17.2%)</td>
<td>8 (16%)</td>
<td>3 (9.1%)</td>
<td>5 (18.5%)</td>
</tr>
<tr>
<td>Engel Class IV</td>
<td>18 (22.8%)</td>
<td>21 (30%)</td>
<td>14 (24.1%)</td>
<td>11 (22%)</td>
<td>10 (30.3%)</td>
<td>9 (33.3%)</td>
</tr>
<tr>
<td>Total</td>
<td>79</td>
<td>70</td>
<td>58</td>
<td>50</td>
<td>33</td>
<td>27</td>
</tr>
</tbody>
</table>
MR imaging findings

- Within normal limits: 26
- Diffuse cerebral atrophy: 23
- Cortical dysplasia: 13
- Hypoxic-ischemic Encephalopathy: 6
- Periventricular leukomalacia: 3
- Cerebral infarction: 2
- Pachygyria: 2
- Microcephaly: 1
- Arachnoid Cyst: 1
- Hypocampal sclerosis: 1
Postoperative seizure outcome based on Engel’s classification

- Atypical absence: 33.3% Engel Class I, 66.6% Engel Class II
- Epileptic spasms: 30.0% Engel Class II, 20.0% Engel Class III, 10.0% Engel Class IV
- Myoclonic seizure: 22.2% Engel Class I, 22.2% Engel Class II, 22.2% Engel Class III, 33.4% Engel Class IV
- Tonic seizure: 30.8% Engel Class II, 19.2% Engel Class III, 26.9% Engel Class IV
- Atonic seizure: 25.8% Engel Class II, 35.5% Engel Class III, 9.7% Engel Class IV, 29.0% Engel Class IV
Case Report (Girl, Born in January-2004)
She was born without perinatal complication. Her development has showed appropriate up to 2 years old. Since then, she showed language disability. Seizures have been onset at 3yr 2 months old. At that time, she showed two kinds of abnormal behaviors that the first type was brief motion arrest with upward gaze 3~4sec and the second type was upward gaze with tonicity of the both upper extremities 5~6sec, 1~2 times/day. The frequency was increased up to 5~10 times/day. She failed multiple trials of AED’s and also ketogenic diet.
PHYSICAL EXAMINATION

General examination
HC=49cm (25-50p), Ht=112cm (50p), Wt=16kg(10-25p)
Hand dominancy : Rt. side        Pathologic handedness (+)
Visual dominancy : unchcked

Neurologic examination
M/S : alert
Light reflex : +++/+++
Sensory : intact
Motor : Lt. Upper G 5 Lt. Lower G 5
       Rt. UpperG 5 Lt. Lower G 5
Reflex : (Lt/Rt) Bj ++/++,Tj ++/++, Kj ++/++, Aj ++/++
Babinski sign (-/-), Ankle clonus (-/-)

Development : IQ 30 (2009.11.20 age of 5 yr 10 mo)
Head control - 3mo Sitting- 6mo Independent walking – 11mo
Social and personal – delayed about 1year
Language - delayed about 1year
Mental retardation (+)
Total intelligent score : 43 - Moderate Mental Retardation

Language : 46 - Moderate Mental Retardation

Performance : 53 - Mild Mental Retardation

Visual – performance test : below 2year and 10month years old

Social score : 2year and 7month years old

Right handedness
Seizure profile

Ictus: Type A. myoclonic jerking
Type B. vacant staring and EBD upward with tonicity of both arms
Type C. head drop (recently most of seizures)

Aura: (-)
LOC: (+)
Onset: 3yr 2months
Frequency: 3-4/day
Post-ictal: alert
Imp:
- Lennox-Gastaut syndrome

Medication:
- Lamiotrigine 200mg #2
- Valproic acid 300mg #2
- Rufinamide 400mg #2
PostOP
Discussion

In our series the procedure significantly decreased or stopped seizure in 57 (72.2%) patients with an average follow-up period 3.4 years (ranged from 0.5 to 8.4 years).

The corpus callosotomies were successful in decreasing atonic and tonic seizures in many patients with Lennox-Gastaut syndrome, thereby physiological injuries from falls have also been reduced which significantly improved the quality of life. Patients with myoclonic seizures, epileptic spasms and atypical absence have also benefited from callosotomy.

In addition, corpus callosotomy is not only therapeutic, but also diagnostic procedure.

Callosotomy can help
- to identify of potentially epileptogenic hemisphere or foci, that may qualify for subsequent further resection
- may help to prevent convulsive status epilepticus.
In conclusion

The study demonstrated that corpus callosotomy is an effective palliative procedure for patients suffering from LGS that are not candidates for resective surgery. With the advent of new microsurgical techniques, the potentially serious intra-/post-operative morbidity risks are relatively low and corpus callosotomy is becoming a more commonly used as palliative surgical option for children and adolescence with intractable seizure associated with LGS.
Thank You for Your Attention!

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