

Is deep brain stimulation useful in Lance–Adams syndrome?

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Abstract

Lance–Adams syndrome (LAS) is a chronic post-hypoxic myoclonus that occurs after successful cardiopulmonary resuscitation. Although many drugs are available to treat this condition, the underlying mechanism of the disease is yet to be understood. Deep brain stimulation (DBS) has been attempted and proven to be partially successful in treating LAS in several cases. Here, we present a 40-year-old woman who developed myoclonus subsequent to cardiopulmonary arrest (CPA) that occurred after her first cesarean delivery at the age of 26 years. The patient underwent implantation of bilateral globus pallidus interna (GPi)-DBS about 14 years after disease onset. Regarding Unified Myoclonus Rating Scale (UMRS), 8% and 20% improvements were observed in action and resting myoclonus, respectively, with high-frequency stimulation as a result of the 3-year follow-up study. In this case, neuromodulation therapy applied 14 years after hypoxia-causing LAS was not sufficiently beneficial.

Keywords: Deep brain stimulation, myoclonus, cardiopulmonary resuscitation, post-hypoxic myoclonus

INTRODUCTION

Post-hypoxic myoclonus (PHM) is a rarely diagnosed disease and is difficult to treat. It is characterized by sudden, brief involuntary muscular jerking movements after cardiopulmonary resuscitation (CPR). PHM is of two types, namely acute and chronic.¹ In the literature, the chronic PHM has been described as Lance Adams Syndrome (LAS).^{2,3} Both the incidence and pathogenesis of the disease have not been clearly described, and only little is known about effective treatment strategies. Globally, around 150 patients have been diagnosed with LAS, and only five have been treated with bilateral globus pallidus interna-deep brain stimulation (GPi-DBS). In this article, we present an LAS case that did not benefit from bilateral GPi-DBS, unlike the five cases published in the literature.^{4,8}

CASE REPORT

A 40-year-old woman went into cardiopulmonary arrest (CPA) during an emergency cesarean delivery at the 36th week of pregnancy at the age of 26 years. The medical history of the patient was unremarkable, with no chronic disease

and no regular medication during pregnancy. After successful resuscitation, she regained consciousness on the 7th day. During the 2nd week, she developed involuntary jerky movements and was treated with several drugs, but without any significant beneficial effects. She had previously been treated with clonazepam, levetiracetam, lacosamide, and sodium valproate. She was referred to our hospital 14 years after the onset of myoclonus. She experienced myoclonus both at rest and at action and could not eat or walk by herself. Her Cattell 2A culture-free mental test score was 66, which denoted mild mental retardation. Negative and positive myoclonic jerks mostly originated from the flexor muscles in the upper and lower extremities and the abductor muscles in the trunk. Occasionally, the orbital muscles were also involved. It was observed that the myoclonic jerks became more frequent during tactile stimulation and action. More than one jerk occurred per second during action, and 1–2 times per 10 seconds at rest were recorded (Video 1). Surface electromyograms revealed a cortical and subcortical myoclonus pattern. Giant somatosensory evoked potentials were not noted in our case.

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Table 1: Preoperative and postoperative Unified Myoclonus Rating Scale (UMRS) scores

	Section	Preoperative	Postoperative 3 rd year
1	Patient questionnaire	33	30
2	Myoclonus at rest	30	24
3	Stimulus sensitivity	13	11
4	Myoclonus with action	112	103
5	Functional tests	26	23
6	Global disability score	4	4

Electroencephalogram (EEG) did not reveal any seizure activity. Magnetic resonance imaging (MRI) showed a millimetric ischemic area in the white matter in both cerebral hemispheres. Unified Myoclonus Rating Scale (UMRS) was 112 for action, 30 for rest, and 13 for stimulus sensitivity (Table 1). The patient underwent implantation of bilateral GPi-DBS about 14 years after disease onset. Stereotactic CT-derived targets to the posterolateral GPi guided the placement of the leads with the confirmation of intraoperative microelectrode recordings (MER) (Figure 1). Postoperative CT images superimposed on the preoperative MRI and we confirmed that both the leads were in the optimal position for GPi (Figure 2). The stimulation of GPi was started during the 1st postoperative week at high frequency.

During follow-up, the DBS parameters were adjusted to different frequencies, and the best response was obtained at a high frequency. During the last programming session, the parameters were adjusted to an amplitude of 3V, a pulse width of 60 μ s, and a frequency of 130 Hz. Although a minimal decrease was observed in the extremity and eyelid myoclonus after three years, no improvement was noted in the trunk myoclonus (Video 2). The UMRS motor scores are shown in Tables 1 and 2, along with the five other published LAS-DBS cases. The medications of the patient were maintained at the same dosage.

DISCUSSION

After successful CPR, several neurological symptoms such as PHM may appear depending

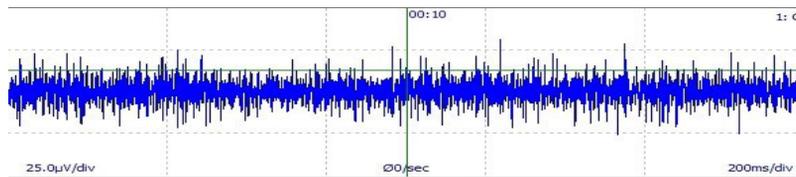


Figure 1. Perioperative microelectrode recordings of the patient; the target point for globus pallidus interna

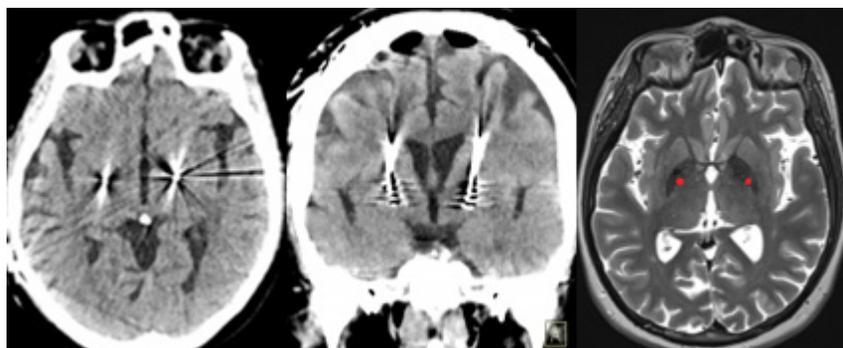


Figure 2. Postoperative CT images was fused with the preoperative MRI, and the ends of the leads were identified with red dots.

Table 2: Published cases of Lance Adams Syndrome treated with bilateral globus pallidus interna-deep brain stimulation (GPi-DBS)

Study	Etiology	Time to DBS	Target	Preoperative UMRS	Postoperative UMRS	Improvement	Follow-up
Asahi <i>et al.</i> (2015) ⁷	CPA	1 year	BL Gpi	Action: 25 Rest: 8 Stimulus Sensitivity: 5	Action: 5 Rest: 0 Stimulus Sensitivity: 0	Action: 80% Rest: 100%	6 months
Ramdhani <i>et al.</i> (2017) ⁸	Asthmatic attack and CPA	3 years	BL Gpi	Action: 52 Rest: 75 Stimulus Sensitivity: 0	Action: 32 Rest: 0 Stimulus Sensitivity 0	Action: 38% Rest: 100%	6 months
Mure <i>et al.</i> (2020) ⁴	ICA rupture and CPA	1 year	BL Gpi	Action: 90 Rest: NA Stimulus Sensitivity: NA	Action: 24 Rest: NA Stimulus Sensitivity NA	Action: 73% Rest: NA	2 years
Kim <i>et al.</i> (2020) ⁵	Drowning and CPA	3 years	BL Gpi	Action: 80 Rest: 32 Stimulus Sensitivity: 17	Action: 52 Rest: 3 Stimulus Sensitivity 8	Action: 35% Rest: 90%	5 years
Gao <i>et al.</i> (2020) ⁶	Drowning and CPA	5 months	BL Gpi	Action: 61 Rest: 2 Stimulus Sensitivity: NA	Action: 40 Rest: 0 Stimulus Sensitivity NA	Action: 35% Rest: 100%	1 year
Current case	During delivery-CPA	13 years	BL Gpi	Action: 112 Rest: 30 Stimulus Sensitivity: 13	Action: 103 Rest 24 Stimulus Sensitivity 11	Action: 8% Rest: 20%	3 years

CPA: cardiopulmonary arrest; UMRS: Unified Myoclonus Rating Scale; BL: Bilateral; A: Action, R: Rest; SS: Stimulus sensitivity, NA: Not available

on the area of the hypoxic damage to the brain.⁹⁻¹⁰ The myoclonus in LAS has no steady correlation with EEG abnormalities.¹¹ In the previously reported LAS cases, spike waves, or multiple spike activities on the EEG, or a decrease in the EEG frequency were observed. However, the EEG may also have multiple spikes followed by slow waves, and these spike waves may be accompanied frequently but not always by myoclonic jerks.^{9,12} Myoclonus can be triggered by sensory stimuli in certain patients. Moreover, the negative myoclonus can occur in the lower extremities, impairing the posture and resulting in falls. Other neurological symptoms such as seizures, cerebellar ataxia, and cognitive impairment are commonly seen in LAS, similar to our case. Myoclonus can be positive/negative and subcortical/cortical in the LAS. Especially, action myoclonus tends to persist for the entire life and is difficult to treat, lowering the overall

function and quality of life. Medical therapies, such as levetiracetam, piracetam, clonazepam, or sodium valproate, either alone or in combination, have been used to treat LAS. In addition, new anti-myoclonic treatments promise to treat LAS.¹³ To date, over 150 cases have been reported in the literature. As mentioned earlier, five LAS cases have so far been treated with bilateral GPi-DBS (Table 2). GPi is the preferred target in myoclonus-dystonia patients and has also been used in the previously published LAS cases; hence, we chose it in our case. Asahi *et al.*⁷ reported a case of LAS after respiratory distress-induced CPA. They reported the first bilateral GPi-DBS case. In this case, GPi-DBS, which was applied five months after the injury, yielded beneficial results. Ramdhani *et al.*⁸ were the first to review DBS-treated LAS, and they stimulated bilateral GPi in an LAS patient with a CPA etiology and an interval of three years between the injury and DBS. In their

report, they published a successful clinical result, with a 38% improvement in action myoclonus at the 6-month follow-up. Recently, Kim *et al.*⁵ reported a case involving low frequency (110 Hz) GPi-DBS, with an interval of three years from injury to DBS. They also published a successful clinical result, with a 35% improvement in action myoclonus at the 5-year follow-up. Mure *et al.*⁴ reported a 73% improvement in action myoclonus, with GPi-DBS, administered one year after injury in a 79-year-old LAS case with internal carotid artery (ICA) rupture and CPA. Gao *et al.*⁶ showed that bilateral GPi-DBS provided 35% benefit in action myoclonus in a 33-year-old patient with LAS of asphyxia etiology. In these five published cases, the time of brain damage that caused LAS was a maximum of three years. In our case, GPi-DBS application partially reduced distal extremity and orbital myoclonus of cortical origin, but subcortical axial myoclonus did not decrease in our patient. In our case, we did not see a significant improvement in action or resting myoclonus during the 3-year follow-up following GPi-DBS 14 years after the injury. DBS is one of the options in the treatment of LAS; however, the procedure could not be viewed as the gold-standard. What we need to focus on here is the application of neuromodulation therapy in the early stages of the disease. We postulate that the use of neuromodulation surgery in the early period after brain injury would be beneficial.

DISCLOSURE

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Conflict of interest: None

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