Pallidal Deep Brain Stimulation for Lance-Adams syndrome

A 79-year-old woman presented with a history of cardiac arrest due to internal carotid artery rupture following carotid endarterectomy after successful cardiopulmonary resuscitation. However, within 1 month, the patient developed sensory stimulation-induced myoclonus in her face and extremities. Because her myoclonic symptoms were refractory to pharmacotherapy, deep brain stimulation of the GPi was performed 1 year after the hypoxic attack.

Continuous bilateral Pallidal Deep Brain Stimulation with optimal parameter settings remarkably improved the patient's myoclonic symptoms. At the 2-year follow-up, her Unified Myoclonus Rating Scale score decreased from 90 to 24. In addition, Mure et al. observed burst firing and interburst pause patterns on intraoperative microelectrode recordings of the bilateral GPi and stimulated this area as the therapeutic target.

The results show that impairment in the basal ganglion circuitry might be involved in the pathogenesis of myoclonus in patients with Lance-Adams syndrome ¹⁾.

A 23-year-old male with chronic medication refractory PHM following a cardiopulmonary arrest related to an asthmatic attack who improved with bilateral globus pallidus internus (GPi) deep brain stimulation (DBS). Ramdhani et al. reviewed the clinical features of PHM, as well as the preoperative and postoperative Unified Myoclonus Rating Scale scores and DBS programming parameters in this patient and compare them with the three other published PHM-DBS cases in the literature.

This patient experienced an alleviation of myoclonic jerks at rest and a 39% reduction in action myoclonus with improvement in both positive and negative myoclonus with bilateral GPi-DBS. High frequency stimulation (130 Hz) with amplitudes >2.5 V were needed for the therapeutic response.

They demonstrated a robust improvement in a medication refractory PHM patient with bilateral GPi-DBS, and suggest that it is a viable therapeutic option for debilitating post-hypoxic myoclonus ²⁾.

The first case of a patient who developed action myoclonus after experiencing perinatal anoxia and was successfully treated by chronic deep brain stimulation (DBS) of the thalamus (thalamic DBS).

The effectiveness of chronic thalamic DBS in this patient supports the concept of involvement of the thalamus in post-perinatal anoxic myoclonus $^{3)}$.

Asahi T, Kashiwazaki D, Dougu N, et al. Alleviation of myoclonus after bilateral pallidal deep brain stimulation for Lance-Adams syndrome. J Neurol. 2015;262(6):1581-1583. doi:10.1007/s00415-015-7748-x $^{4)}$.

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Yamada K, Sakurama T, Soyama N, Kuratsu J. Gpi pallidal stimulation for Lance-Adams syndrome. Neurology. 2011;76(14):1270-1272. doi:10.1212/WNL.0b013e31821482f4 ⁵⁾.

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