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Case Report

Motor Function Improvement Following Intrathecal Baclofen Pump Placement in a Patient with Locked-in Syndrome

ABSTRACT

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We describe a patient with locked-in syndrome who had minimal volitional motor function and severe spasticity in all four extremities. The patient showed a significant improvement in volitional motor function following intrathecal baclofen pump therapy to control spasticity. This case study suggests that intrathecal baclofen pump therapy might improve motor function in select patients with locked-in syndrome.

Key Words: Locked-in Syndrome, Intrathecal Baclofen Pump, Spasticity, Motor Function

Locked-in syndrome (LIS) is a debilitating syndrome generally caused by injury of the ventral pons. Its main clinical manifestations are quadriplegia, lower cranial nerve paralysis, and mutism. Patients with classic LIS are unable to talk or move. Consciousness remains intact, and patients are only able to communicate by using eye blinking or eye movements.

There are three variants of LIS characterized in the literature: classical, incomplete, and total.¹ The classical type, originally noted by Plum and Posner,² includes patients with the signs and symptoms described above. Patients classified as having incomplete LIS are similar to the classical variety except that they retain limited motor function beyond vertical eye movements. The total category consists of patients who are totally immobile and cannot communicate.

Cases of immediate and complete recovery from transient LIS have occurred.³ However, lack of recovery after 6 mo is strongly predictive of chronic and persis-

tent LIS.^{4,5} Patients who survive beyond the first 2 mo have a significantly lower mortality rate.⁶ Rehabilitation efforts typically focus on establishing communication strategies, often with the use of computer-aided technologies, and establishing environmental controls. Relatively small motor function gains can give a patient with LIS the ability to interact with their environment by using computer-aided technologies, motorized vehicles, and greater spontaneous expression. We report a case of LIS in which intrathecal baclofen (ITB) pump therapy utilized for spasticity was found to have unanticipated benefits with regard to improved motor control.

Spasticity as described by Lance⁷ is “a motor disorder characterized by a velocity-dependent increase in tonic stretch reflexes (muscle tone) with exaggerated tendon jerks.” Quantifying spasticity can be challenging because of observer variability, fluctuating temporal measurements, and the plethora of different clinical scales in use. The modified Ashworth scale is one clinical scale widely used to quantify spasticity because it has relatively high interrater reliability.⁸

CASE REPORT

We describe the case of a otherwise healthy 37-yr-old man with a history of congenital coarctation of the aorta who underwent an elective repair of the coarctation via an endovascular stent placement. Immediately after the procedure, the patient was able to communicate normally. During the evening of the procedure, he became increasingly lethargic, had difficulty with vocalization, and developed right hemiplegia. The initial evaluation included a computed tomographic scan of the head and lumbar puncture, both of which were negative. A subsequent magnetic resonance image revealed an acute obstruction of the basilar artery in the posterior circulation and a ventral pontine infarction (Fig. 1). The patient’s symptoms continued to deteriorate

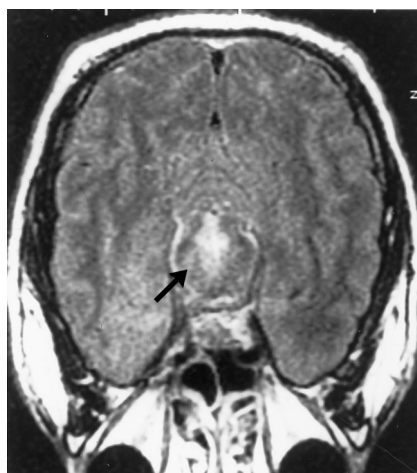


Figure 1: T2-weighted flair magnetic resonance image with an *arrow* pointing to the area of infarction in the ventral pons.

rate to the point that he was “locked in” and unable to move his extremities or verbalize. His volitional movements were limited to vertical eye movements and closing his eyelids. His acute hospital course was marked by mechanical ventilation for airway protection and gastrostomy tube placement for nutrition. Significant spasticity developed in all four extremities. The patient was transferred to a rehabilitation hospital for a multidisciplinary rehabilitation 6 wk after the brainstem infarction.

During his rehabilitation hospitalization, the patient exhibited persistent four-limb spasticity without observable volitional control. The patient’s reflexes were brisk in all four limbs and his lower extremities exhibited clonus bilaterally. No cogwheeling was evident on exam and his sensory exam was unreliable. The patient was able to respond with reasonable consistency to yes or no questions by using eye movements.

Progressive spasticity of his hip flexors and knee flexors developed (3/4 on the modified Ashworth scale) and resulted in the patient’s heels being in contact with his buttocks. Painful spasticity affecting all four limbs interfered with positioning, passive range of motion, and attempts to establish improved communication or environmental control. Trials of oral

medications were undertaken, including oral baclofen, tizanidine, and dantrolene. The patient continued to experience an abnormal resting position with his lower extremities flexed at the knee without being able to be stretched to an extended position. The patient was unable to perform head movements such as flexion or extension, lateral rotation, and shrugging of his shoulders on a consistent basis. Oral medications were attempted, but they were limited by dose-related sedation. Because of his persistent wide-spread spasticity refractory to oral medical therapy, a test dose of 50 μg of baclofen was injected intraspinally. The patient showed a substantial reduction in spasticity of 1–2 points on the modified Ashworth scale in affected limbs. The patient subsequently had an ITB pump placed 16 wk after transfer. An infusion rate of baclofen beginning at 50 $\mu\text{g}/\text{day}$ was gradually increased. Four days after initiating ITB pump therapy, the patient had a marked decrease in spasticity most evident in his lower extremities. Furthermore, the patient demonstrated left thumb abduction to command with a 3/5 strength rating. The baclofen dosage was titrated to 230 $\mu\text{g}/\text{day}$, which appeared to provide optimal control of spasticity. The patient continued to exhibit further motor function gains, including being able to perform flexion and extension movements and lateral rotation of his head on a consistent basis and being able to shrug his left shoulder.

His subsequent rehabilitation hospital course was notable for gradual return of spasticity with attendant loss of motor control. This was attributed to ITB tolerance, and improved with a progressive increase of dosage to 330 $\mu\text{g}/\text{day}$. This restored control of the patient’s spasticity, and his motor control returned to his new baseline. The patient now utilizes head nodding and shoulder movements routinely for communication. Effort is being made to develop more consistent control of switches with his left thumb.

DISCUSSION

This case illustrates the potential of ITB pump therapy to unmask volitional motor function that may be hidden in extreme cases of spasticity. It has been shown that continuous ITB decreases spasticity in individuals with long-standing hemiplegia.⁹ This patient showed both a significant reduction in spasticity after continuous ITB and also improved motor function. Although in many individuals the return of slight motor function may seem insignificant, in the case of LIS, these movements can be critical. In the case of the patient described, limited movements allowed the patient a much broader range of spontaneous expression without the use of technological aids. Furthermore, limited motor function of his left thumb enhanced efforts to operate computer-aided devices that could aid in many different functions, including operating a motorized wheelchair, communication, and environmental control.

We distinguished spasticity in our

patient from other forms of increased muscle tone on the basis of the clinical exam. The patient had brisk reflexes in all four extremities and lower extremity clonus bilaterally. Furthermore, tone in his lower extremities was velocity dependent, consistent with Lance's definition of spasticity.

Although it is possible that the patient's improved motor control was simply coincidental, this seems unlikely. The patient was noted to regain function 4 days after the ITB pump therapy was initiated. Furthermore, when the patient's spasticity worsened because of ITB tolerance, his control of head movements declined, only to improve with an increase in ITB dosage. This case illustrates the potential use of ITB therapy in some patients with LIS to unmask motor function that is being hindered by severe spasticity.

REFERENCES

1. Bauer G, Gerstenbrand F, Rump E: Variables of the locked-in syndrome. *J Neurol* 1979;221:77-91
2. Plum F, Posner JB: *The Diagnosis of Stupor and Coma*. Philadelphia, Davis, 1966
3. Khurana RK, Genut AA, Yannakis GD. Locked-in syndrome with recovery. *Ann Neurol* 1980;8:439-41
4. Katz RT, Haig AJ, Clark BB: Long-term survival, prognosis, and life care planning for 29 patients with chronic locked-in syndrome. *Arch Phys Med Rehabil* 1992;73:403-8
5. Patterson JR, Grabois M: Locked-in syndrome: A review of 139 cases. *Stroke* 1986;17:758-64
6. Haig AJ, Katz RT: Locked in Syndrome: A Review. *Curr Concepts Rehabil Med* 1986;3:12-4
7. Lance JW: Symposium synopsis, in Feldman RG, Young RR, Koella WP (eds): *Spasticity: Disordered Motor Control*. Chicago, Year Book Medical Publishers, 1980
8. Bohannon RW, Smith MB: Interrater reliability on a modified Ashworth scale of muscle spasticity. *Phys Ther* 1987;67:206-7
9. Meythaler JM, Guin-Renfroe S, Hadley MN: Continuously infused intrathecal baclofen for spasticity/dystonic hemiplegia: A preliminary report. *Am J Phys Med Rehabil* 1999;78:247-54

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