

# Letters

## OBSERVATION

### Recovery From Locked-in Syndrome

Locked-in syndrome may be classified into 3 categories: the classic form, the incomplete form (patients have other voluntary movements), and the total form with a completely motionless patient including absence of vertical eye movements.<sup>1</sup> Clinical

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Video at [jamaneurology.com](http://jamaneurology.com)

experience has shown that classic locked-in syndrome from pontine infarction may improve, but rarely, to functional independence.<sup>2</sup> In a series of 14 patients with classic or total locked-in syndrome seen 3 to 6 months following neurorehabilitation, motor recovery was found in 21%; return of swallowing in 42%; verbal communication in 28%; bladder and bowel control in 35%; and weaning of the ventilator in half of the patients.<sup>3</sup> Most dramatic is early recovery from locked-in syndrome usually reported after recanalization of the basilar artery<sup>4</sup> but improvement may be the result of major fluctuation of deficit in the early defining stage.<sup>5</sup> We present an improvement of a patient in a nearly complete locked-in syndrome.

**Report of a Case** | A 22-year-old healthy man who exercised 3 to 4 hours daily presented with an acute basilar artery occlusion. Pontine and cerebellar infarctions were found despite aggressive treatment with intravenous thrombolysis and endovascular thrombectomy (**Figure, A**). Follow-up vascular imaging confirmed complete recanalization. He remained awake and attentive, with a gaze preference to the left and minimal horizontal eye movements to the right. He had a complete flaccid quadriplegia. Vertical eye movements and blinking were preserved. He had a trace of jaw and oropharyngeal movement (**Video**). His stroke was found to be due to relative dehydration in the setting

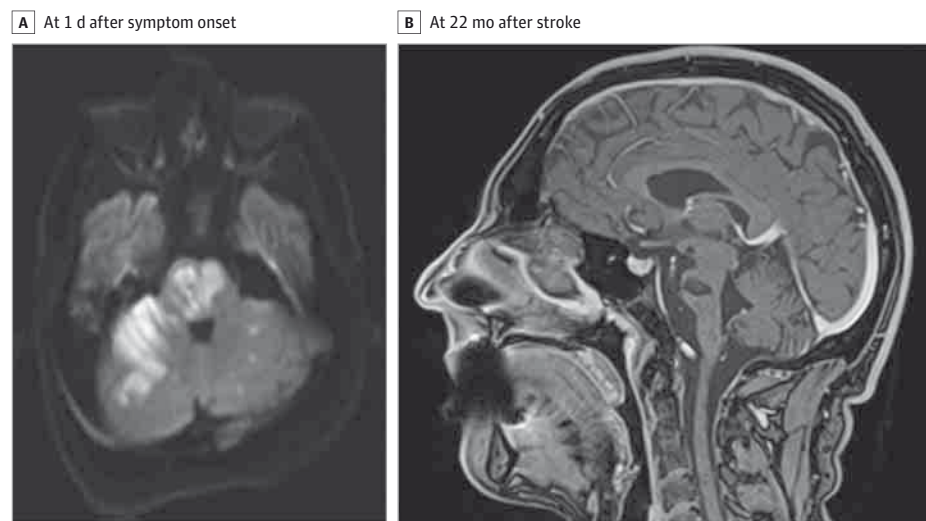
of a heterozygous mutation for factor V Leiden. At 3 months, he was weaned off the ventilator and tracheostomy was removed. There was a severe dysarthria and some cephalic, but no trunk control. At 6 months, he was able to sit with support of his hands. At 10 months, he was able to ambulate with a walker. At 17 months, he was able to climb stairs. His trajectory of recovery was very gradual but eventually led to a substantial recovery (**Video**). His follow-up magnetic resonance imaging scan showed a large punched-out infarct in the ventral pons (**Figure, B**).

**Discussion** | Locked-in syndrome is due to a ventral pons lesion, which deafferentiates the patient for most motor function (absent horizontal eye movements, no grimacing, no swallowing, no head movements, and no limb movements). Patients with locked-in syndrome have their eyes open, blink, and may have spontaneous vertical eye movements. A large proportion of patients die of pulmonary complications or withdrawal of support, but patients may survive for decades. Using serial videos, we were able to document that a patient with a nearly classic syndrome (but incomplete owing to some bulbar function) may improve dramatically over a prolonged period. Perhaps the presence of some oropharyngeal function early may be the prelude to recovery of speech and motor function. Imbalance due to spastic ataxia remained but most other motor functions recovered well.

**Conclusions** | We documented the clinical trajectory of a patient with a large pontine and cerebellar infarct and were able to confirm substantial improvement. Such an improvement may be more likely in incomplete forms of the syndrome.

Sara Hocker, MD  
Eelco F. M. Wijdicks, MD, PhD

Figure. Pontine and Bilateral Posterior Cerebral Artery Territory Strokes



A, Magnetic resonance imaging performed 1 day after the onset of symptoms depicting diffusion restriction in the bilateral posterior cerebral artery territories and pons. B, Follow-up magnetic resonance imaging scan performed 22 months after his stroke that shows a substantial infarct in the ventral pons.

**Author Affiliations:** Division of Critical Care Neurology, Mayo Clinic, Rochester, Minnesota.

**Corresponding Author:** Sara Hocker, MD, Division of Critical Care Neurology, Mayo Clinic, 200 First St SW, Rochester, MN 55905 (hocker.sara@mayo.edu).

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### Corticosteroid-Induced Paraplegia—A Diagnostic Clue for Spinal Dural Arterial Venous Fistula

A man in his 60s was evaluated for progressive myelopathy. His symptoms evolved insidiously over a year and included groin numbness, incomplete bladder emptying, and leg weakness. Examination revealed spastic paraparesis. Magnetic resonance imaging (MRI) of the cervical and thoracic spine demonstrated a T2 hyperintense lesion extending from T6 to the conus but no evidence of abnormal flow voids over the surface of the cord and no evidence of gadolinium enhancement. Magnetic resonance imaging of the brain was normal. His leg weakness worsened after walking and improved with rest. The following blood results were negative or normal: vitamin B<sub>12</sub>, copper, West Nile virus, human T-cell lymphotropic virus types 1 and 2 antibodies, human immunodeficiency virus antibodies, methylmalonic acid, Lyme serology, erythrocyte sedimentation rate, lupus panel, antineutrophil cytoplasmic antibody, and VDRL. A lumbar puncture revealed a normal profile aside from mildly elevated total protein (0.068 g/dL; range, 0.015-0.06 g/dL; to convert to grams per liter, multiply by 10.0); notable negative cerebrospinal fluid

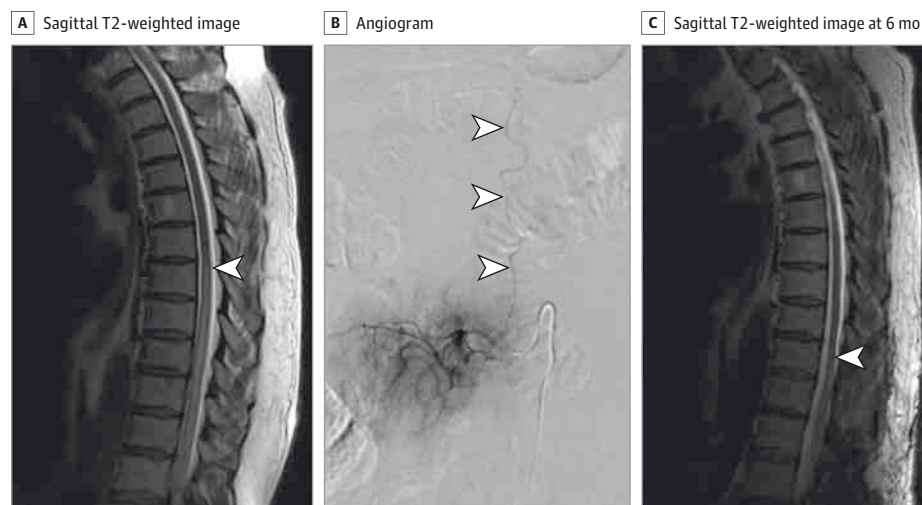
results included oligoclonal bands, IgG index and synthesis rate, neuromyelitis optica-IgG, paraneoplastic panel antibodies, angiotensin-converting enzyme, cryptococcal antigen, and cytology. An angiogram showed no evidence of arteriovenous malformation.

He was treated with intravenous methylprednisone for presumed transverse myelitis. However, approximately 1.5 hours after administration, he experienced acute paraplegia. A repeated cervical and thoracic MRI showed no changes. Seventy-two hours later, paraplegia resolved. A month later, his gait deteriorated further and he underwent a T10-T11 laminectomy. An intraoperative fluorescein angiogram revealed normal findings. The spinal cord appeared normal; spinal cord biopsies were nondiagnostic. One month later, he was treated again with methylprednisone. As before, approximately 1.5 hours after the infusion, he experienced acute paraplegia, which resolved after 48 hours. After this episode, his bladder function worsened and he required self-catheterization. His gait further deteriorated such that he required a wheelchair. A positron emission tomographic scan to detect systemic sarcoidosis or other inflammation yielded negative results.

He was referred to our institution. His medical history confirmed these details; symptoms were confined to the legs; and he had no pain and no upper extremity symptoms. He had severe sensory ataxia and spastic paraparesis, right worse than left; he was incontinent of bladder and bowel. There was a sensory level at approximately T12 affecting all sensory modalities.

Considering the worsening of myelopathic symptoms after walking and improvement at rest and worsening following intravenous corticosteroid, the working diagnosis was spinal dural arterial venous fistula (SDAVF) despite the previous negative angiogram result. A magnetic resonance angiogram spine showed dilatation of posterior pial venous plexus associated with cord expansion and intramedullary edema (Figure, A), strongly suggesting the presence of venous hypertension. An SDAVF originating from the right L2 pedicle was identified

**Figure. Radiologic Evidence of Spinal Dural Arterial Venous Fistula Ablation**



A, Sagittal T2-weighted image of the thoracic spine demonstrates mild cord expansion associated with intramedullary hyperintensity extending into the conus (arrowhead). B, Angiogram demonstrating L2 spinal dural arterial venous fistula (arrowheads). C, Sagittal T2-weighted image of the thoracic spine 6 months following fistula ablation showing markedly diminished signal abnormality (arrowhead).