

Recovery from Locked-in Syndrome after Posttraumatic Bilateral Distal Vertebral Artery Occlusion

Jose M. Cabezudo, M.D., Ph.D., Javier Olabe, M.D., Alfredo Lopez-Anguera, M.D., and Fernando Bacci, M.D., Ph.D.

Service of Neurosurgery, Complejo Sanitario "Virgen de Lluch," Palma de Mallorca, Spain

Cabezudo JM, Olabe J, Lopez-Anguera A, Bacci F. Recovery from locked-in syndrome after posttraumatic bilateral distal vertebral artery occlusion. *Surg Neurol* 1986;25:185-90.

A 55-year-old man developed a delayed locked-in syndrome after a mild head injury. Angiography showed bilateral distal vertebral artery occlusion. Anticoagulant therapy and energetic medical management to promote collateral circulation to the structures in the posterior fossa led to a functional recovery. A review of the reported cases of posttraumatic locked-in syndrome has allowed the authors to differentiate between two types: the first is due to primary brainstem injury and the second is due to secondary brainstem ischemia. Both types have different modes of onset, mechanisms of production, angiographic findings, and prognosis. It is concluded that, with early diagnosis and vigorous medical management, expectations for a useful recovery are high, especially in those cases of posttraumatic locked-in syndrome due to secondary brainstem ischemia.

KEY WORDS: Head injury; Neck injury; Vertebral occlusion; Angiography; Anticoagulation; Locked-in syndrome

The term "locked-in syndrome" (LIS) was coined by Plum and Posner [35] to describe a state of selective motor de-efferentation, characterized by quadriplegia, mutism, sparing of vertical eye movement and blinking, and paralysis of the lower cranial nerves without affecting consciousness. Sensation is preserved and communication with the patient is possible through blinking or vertical eye movements or both. The most common cause of locked-in syndrome is primary vascular disease of the ventral pons or medulla oblongata [2,7,10,15,17-20,23,24,26], but cases due to other causes such as tumor [20,31], postinfectious polyneuropathy [11,13], central pontine myelinolysis [1,29,34,40], air

embolism [32], diazepam toxicity [14], and trauma [8,9,12,17,27] have been reported.

Although at first it was thought that the condition was irreversible and fatal, some recent reports indicate that there are patients with a fully developed locked-in syndrome who recover from it with variable degrees of success [11-14,24,26,29,31,32,40]. As far as we know, after reviewing the literature, only five cases of posttraumatic locked-in syndrome have been reported to date [8,9,12,17,27].

This report describes the case of a patient with locked-in syndrome after a mild head injury in whom cerebral angiography showed bilateral distal vertebral artery occlusion and who recovered from it with medical management.

Case Report

A 55-year-old man was involved in an automobile accident on February 18, 1983 when his car was hit on the left side by another car about 700 m from our hospital. Upon arrival in the emergency room, he was conscious and complained of pain in the left side of his forehead and in his neck. He had a bruise on the left side of his forehead. Neurological examination was normal. Roentgenograms of his skull and cervical spine were normal. He was released from the hospital, but before he could leave, he vomited several times and became obtunded.

When first seen by us, he was obtunded but easily arousable. Neurological examination showed only a slight degree of dysarthria. Less than 1 hour later, abnormalities of the ocular movements, consisting of transient episodes of disconjugation of the eyes in the horizontal plane and nystagmus of the left eye, were seen for the first time. A computed tomography (CT) scan was normal. The patient was admitted for neurological observation. He remained the same until 20 hours later when he became lethargic, quadriparetic, and mute. Respiration was difficult due to accumulation of secretions in the upper respiratory tract. The eyes were deviated

Address reprint requests to: Dr. Jose M. Cabezudo Artero, Apartado de Correos no. 382, Palma de Mallorca, Islas Baleares, Spain.

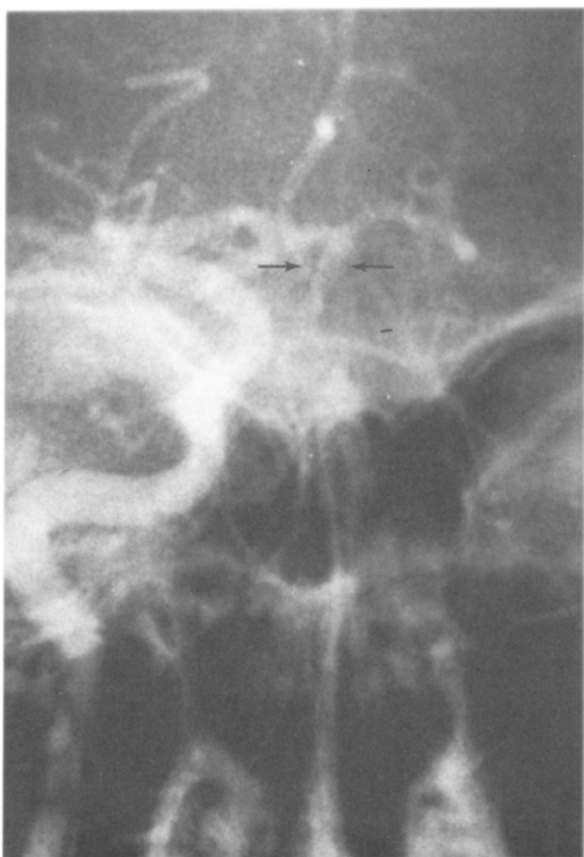


Figure 1. Left carotid angiogram. Arrows point to the distal basilar artery filling in a retrograde fashion.

towards the left, with both pupils equal and reactive. The plantar response was extensor on the left side. The ocular fundi were normal. A bilateral carotid angiogram was interpreted as normal, and the patient was admitted to the intensive care unit where he was intubated and respiration controlled.

On the next morning he showed facial diplegia, quadriplegia, and anarthria. The patient remained conscious, and he could blink when asked. Vertical eye movements were unimpaired, as was the gaze towards the left. However, when looking to the right, the right eye did not abduct, and the left eye showed nystagmus. There was loss of movement of the tongue and palate, and of gag reflex. Both plantar responses were extensor. Sensation remained unimpaired. The previous carotid angiogram was reviewed, and retrograde filling of the distal basilar artery was seen (Figure 1). A transfemoral bilateral vertebral angiogram showed occlusion of both vertebral arteries beyond the origin of the posterior inferior cerebellar artery (PICA) (Figures 2 and 3).

Systemic anticoagulation with heparin was started immediately to prevent further progression of the thrombus, and measures to prevent hypotensive episodes that

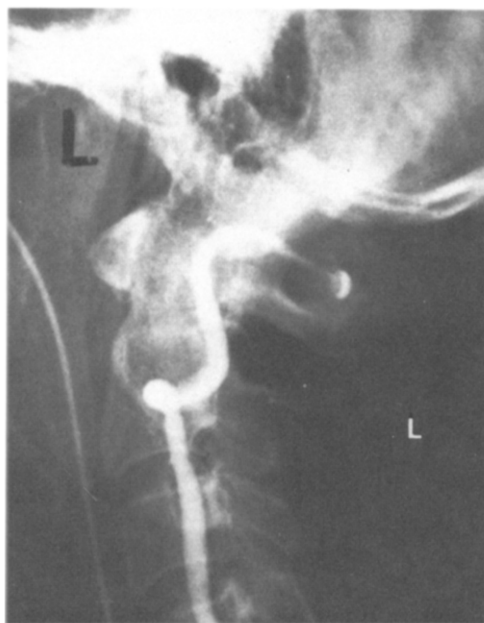
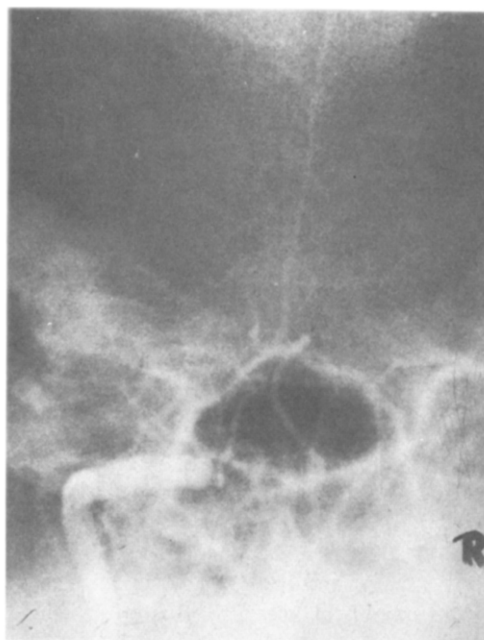


Figure 2. Left vertebral angiogram showing occlusion of the vertebral artery.

would compromise collateral circulation were undertaken along with a strict nursing program to prevent pressure sores and cardiopulmonary complications. The next day, very discrete movements of the neck were seen, but they ceased on the same day. A tracheostomy was performed on February 26, and 4 days later he was started on dicumarol until discharged.

He remained clinically unchanged until 16 days later when voluntary horizontal neck movements were seen. Two days later he could clearly nod his head to answer questions. On March 15, 1983, 25 days after the acci-

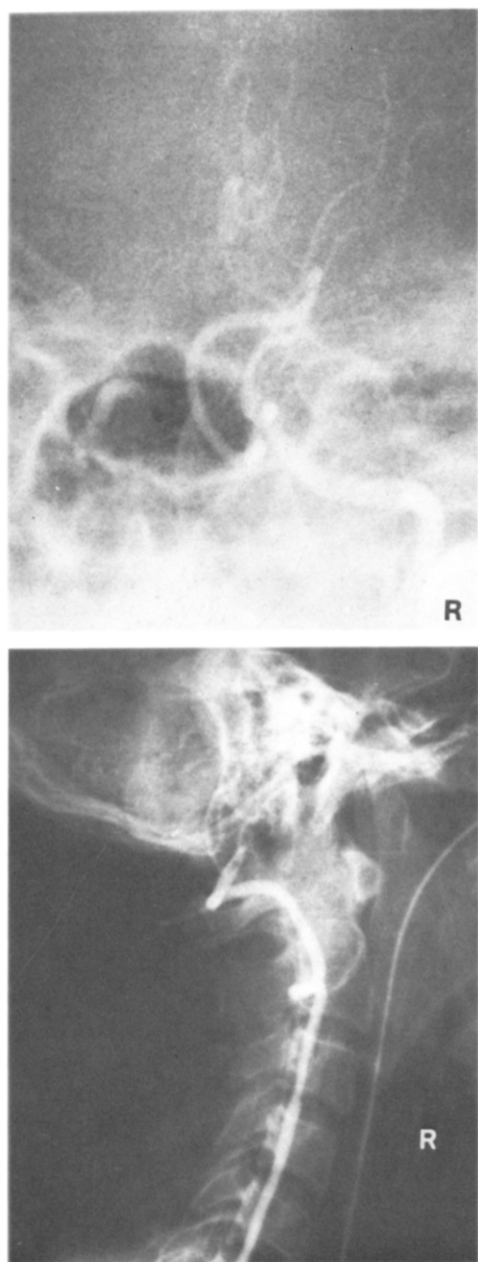


Figure 3. Right vertebral angiogram showing occlusion of the vertebral artery.

dent, movements of the limbs were first seen, and 5 days later he could move his arms and legs against gravity. The same day he was weaned from the respirator. He was moved to the rehabilitation unit for motor and speech therapy. Upon discharge on May 31, he had normal mentation except for a marked emotional lability of the bulbar type. He could speak with mild dysarthria. Ocular movements and swallowing were normal. Deep reflexes were hyperactive, and plantar responses were extensor bilaterally. Although he could stand by himself, walking

was impossible. He could recall only small fragments of his stay in the intensive care unit.

When last seen in December 1984, 22 months after the accident, he had a normal mental status with minimal emotional lability. His speech was almost normal, and ocular movements were normal. Swallowing was normal both for solids and liquids. Manipulation was good. Although he was able to walk unassisted, he occasionally needed canes due to poor coordination and spasticity in the legs.

Discussion

Since 1969 when Plum and Posner [35] coined the term "locked-in syndrome," many reports referring to different aspects such as etiology, clinical evolution, physiological observations, and pathological findings have been published. From these reports it was generally assumed that locked-in syndrome was a devastating condition that ended, sooner or later, in death, with the accompanying emotional distress for the patient and family, and frustration for the medical attendants. Although there were reports of patients surviving as long as 2 to 3 years, they did not experience such a significant neurological improvement as to warrant a more optimistic view [17,20].

In 1972, the first case of recovery from locked-in syndrome was reported by Davis et al [14] in a patient with locked-in syndrome secondary to diazepam toxicity. Since then, as many as 18 cases of transient locked-in syndrome have been reported [8,11-13,20,24,26,27,29,31,32,40]. Of these, three occurred in patients with posttraumatic locked-in syndrome [8,12,27]. To our knowledge, angiography was obtained in only 15 cases of locked-in syndrome [2,7-10,13,15,17-20,24]. Of these, only six cases showed occlusion of the vertebral or basilar arteries or both [2,8,10,17,18,20].

Only five cases of posttraumatic locked-in syndrome have been reported to date [8,9,12,17,27] (Table 1). Ours is the sixth case. Of the six cases, four recovered to a different extent [8,12,27], one remained unchanged after 2 years, [17] and one died [9]. Angiography was performed in four patients, and showed significant vertebrobasilar occlusions in three, including ours [8,17].

The first case of posttraumatic locked-in syndrome was reported by Feldman in 1971 [17]. A 36-year-old woman was involved in an auto-train accident. She did not lose consciousness, but was lethargic and complained of pain in her head, neck, and shoulder. She had a hematoma over the left zygoma and a laceration over the left eye. She suffered a progressive deterioration, and the next morning she was found to be "locked-in". Angiograms were interpreted as indicative of throm-

Table 1. *Reported Cases of Posttraumatic Locked-in Syndrome*

Author (year)	Sex	Age	Time to onset of LIS	Angiographic findings	Time to onset of recovery	Outcome
Feldman [17] (1971)	F	31	24 hours	RVA occlusion with secondary embolization of BA	No recovery	Remained unchanged 2 years later
Marti-Vilalta et al [27] (1977)	M	21	Initial coma	Not obtained	4 hours	Normal status
Britt et al [9] (1977)	M	21	Initial coma	Normal	No recovery	Died
Chang et al [12] (1979)	M	25	Initial coma	Not obtained	10 weeks	Moderate recovery
Bivins et al [8] (1981)	F	43	48 hours	Proximal LVA occlusion, secondary BA occlusion	3 weeks	Good recovery
This report	M	55	24 hours	Distal occlusion of RVA and LVA	19 days	Good recovery

Abbreviations: F, female; M, male; RVA, right vertebral artery; LVA, left vertebral artery; BA, basilar artery; LIS, locked-in syndrome.

bosis of the right vertebral artery with secondary embolic occlusion of the basilar artery. Over the next 5 months, she regained most functions of the motor cranial nerves, but remained tetraplegic and mute.

In 1977, Marti-Vilalta et al [27] reported the case of a 21-year-old man who was rendered "locked-in" after being dragged by a car. He was reported initially to be in coma. The clinical condition cleared in 6 hours. Roentgenograms of the skull and cervical spine were normal. They postulated as the pathophysiologic mechanism that of a transient ischemia of the ventral pons due to compression of the vertebral arteries after abrupt hyperextension-rotation of the neck. In the same year, Britt et al [9] reported the case of a 21-year-old man who was found apneic at the site of a car accident. On admission he was comatose and quadriplegic. He had an abrasion on the left side of his forehead. Roentgenograms of the skull and cervical spine were normal. Forty-eight hours after the accident a bilateral carotid and left vertebral angiogram showed no abnormalities. Five days later, the patient started to blink upon stimulation. By the third week, the patient was reported to be "locked-in." He died on the 14th week after the accident. Postmortem examination demonstrated cavitary necrosis of the ventral pontomedullary junction and bilateral disruption of the sixth cranial nerve. The proposed mechanism of injury was that of a partial traumatic pontomedullary disruption due to hyperextension of the head on the neck.

In 1979, Chang et al [12] reported another case of transient posttraumatic locked-in syndrome in a 25-year-old man who was found unconscious at the scene of an automobile accident. Roentgenograms of the skull and cervical spine were normal, as well as CT scan of the head. He had sustained multiple fractures on the left side of his body, including the left zygoma. He remained comatose for 6 weeks, when he was found to be "locked-in." Three months after the accident, the patient was

able to speak, to grasp his left hand, and to control his sphincters, but he was unable to walk. Based on the injuries on his face and on the findings of auditory evoked potentials, a direct brainstem injury due to hyperextension of the head was postulated.

In 1981, Bivins et al [8] reported the case of a 43-year-old woman who was involved in a minor automobile accident. She did not lose consciousness, but complained of pain in her neck and shoulder. Roentgenograms of the skull and cervical spine were normal. Two days later she became progressively quadriplegic, though remaining fully conscious. A CT scan showed an area of edema in the left cerebellar hemisphere. A four-vessel angiogram showed thrombosis of the left vertebral and basilar arteries. At that moment, 3 days after the accident, systemic anticoagulation was instituted. Three months later, she had recovered enough to be self-sufficient. Bivins et al believed that the mechanism of the injury was thrombosis of the left vertebral artery secondary to hyperextension and lateral rotation of the head followed by propagation of the thrombus into the basilar artery.

Our patient developed a locked-in syndrome within 24 hours after a minor head injury that did not render him unconscious at any time. Roentgenograms of the skull and cervical spine as well as a CT scan were normal. Angiography showed distal occlusion of both vertebral arteries beyond the origin of the posterior inferior cerebellar arteries, and retrograde filling of the distal basilar artery from the anterior circulation. Systemic anticoagulation was instituted. When last seen, 22 months after the accident, he showed moderate residual deficits and was able to cope well with the activities of daily life.

Although the number of cases of posttraumatic locked-in syndrome is not large, it can be seen from the preceding review that posttraumatic locked-in syndrome can be subdivided into two groups according to the mode of onset, mechanism of production, angiographic find-

ings, and clinical evolution. In the first group, locked-in syndrome is due to primary injury to the brainstem by the forces acting in the moment of the accident. The patients reported by Marti-Vilalta et al [27], Britt et al [9], and Chang et al [12] would fit in this group. These patients were rendered unconscious from the first moment. As coma subsided, clinical features of locked-in syndrome became evident. Angiography, when performed, did not show any abnormalities. The outcome, if recovery did not take place very soon [27], was very poor [9,12]. The mechanism of production is thought to be direct damage to the brainstem as demonstrated by the postmortem findings in the case reported by Britt et al [9].

In the second group, locked-in syndrome is due to delayed ischemia of the brainstem. The patients reported by Feldman et al [17], Bivins et al [8], and ours would fit in this group. These patients sustained minor head injuries without losing consciousness at any moment. Within hours after the accident there was a progressive deterioration leading to a locked-in syndrome. Angiography, performed in the three patients, showed occlusion of the vertebral and/or basilar arteries in all of them. Two patients recovered significantly [8], and one remained unchanged [17]. The mechanism of production was thought to be brainstem ischemia secondary to vascular damage that occurred in the moment of the accident.

In our case, the most probable mechanism of production was hyperextension and rotation of the neck with pinching of one or both vertebral arteries between the occipital bone and the atlas. Damage to the intima and media led to local thrombosis of these vessels. Progression of the thrombus led to the development of locked-in syndrome. This mechanism has been claimed by other authors, both after head injury and after chiropractic manipulation of the neck [8,16,17,22,25,28,30,33,36,38,39]. In our patient, we think that maintenance of flow through both posterior inferior cerebellar arteries and the retrograde filling of the basilar artery from the carotid circulation made recovery possible.

With regard to treatment, we think that if a patient develops signs or symptoms of brainstem dysfunction after head trauma, and CT scan rules out the existence of an intracranial hematoma or other space occupying lesion, vertebrobasilar angiography should be performed to exclude the existence of thrombosis in that territory. If there are signs of arterial damage or if thrombosis has already occurred, anticoagulation should be instituted immediately to prevent development or progression of the thrombus. That this procedure is useful has been demonstrated by Sherman et al [39], who found that mortality dropped from 44.4% to 16.7% when anticoagulation was used in a group of patients

with progressive vertebrobasilar stroke due to sudden change in head position. Efforts to augment cerebral blood flow should be undertaken by avoiding head elevations and hypotensive episodes, or by pharmacologically raising blood pressure if necessary. In addition, adequate supportive care and rehabilitation should be instituted. In the case of locked-in syndrome due to primary brainstem injury, management is the same except for anticoagulation, which is not used. Regarding surgical treatment, although several techniques to revascularize the structures of the posterior fossa have been described [3-6,37,41,42], their role in posttraumatic locked-in syndrome of the ischemic type is not clear, but they could be performed in selected cases as reported by Caplan [10].

In conclusion, we think that patients with posttraumatic locked-in syndrome, especially those with a delayed onset and progressive course, deserve vigorous medical management and, most important, a nonfatalistic attitude; this syndrome seems to be more benign than previously thought, and expectations for useful recovery are high.

The authors are indebted to Mr. Antony Bauzá for the photographic work.

References

1. Adams RD, Victor M, Mancal EL. Central pontine myelinolysis. *Arch Neurol Psychiatry* 1959;8:154-172.
2. Al-Wardi DAM, Adams AH, Hamilton AE. Four cases of "locked in" syndrome and review of the literature. *Bull Los Angeles Neurol Soc* 1975;40:60-70.
3. Ausman JI, Lee MC, Klassen AC, Seljeskoj EL, Chou SN. Stroke: what's new? Cerebral revascularization. *Minn Med* 1976;58:223-7.
4. Ausman JI, Nicoloff D, Chou SN. Posterior fossa revascularization: Anastomosis of vertebral artery to PICA with interposed radial artery graft. *Surg Neurol* 1978;9:281-6.
5. Ausman JI, Diaz FG, de los Reyes RA, Park H, Patel S, Boulos R. Anastomosis of occipital artery to AICA for vertebrobasilar junction stenosis. *Surg Neurol* 1981;16:99-102.
6. Ausman JI, Diaz FG, de los Reyes RA, Park H, Patel S, Boulos R. Superficial temporal to proximal superior cerebellar artery anastomosis for basilar artery stenosis. *Neurosurgery* 1981;9:56-60.
7. Bauer G, Gerstenbrand F, Rumpl E. Varieties of the locked in syndrome. *J Neurol* 1979;221:77-91.
8. Bivins D, Biller J, Laster DW, Mclean WT. Recovery from post-traumatic locked in syndrome with basilar artery occlusion. *Surg Neurol* 1981;16:230-4.
9. Brit RH, Herrick MK, Hamilton RD. Traumatic locked in syndrome. *Ann Neurol* 1977;1:590-2.
10. Caplan LR. Bilateral distal vertebral artery occlusion. *Neurology (Cleveland)* 1983;33:552-8.
11. Carrol WM, Mastaglia FL. "Locked in coma" in postinfective polyneuropathy. *Arch Neurol* 1979;36:46-7.

12. Chang B, Morariu MA. Transient traumatic "locked in" syndrome. *Eur Neurol* 1979;18:391-4.
13. Cherington M. Locked in syndrome after "swine flu" inoculation. *Arch Neurol* 1977;34:258.
14. Davis LE, Wesley RB, Juan D, Carpenter CCI. "Locked in syndrome" from diazepam toxicity in a patient with tetanus. *Lancet* 1972;1:101.
15. Dehaene I, Martin JJ. "Locked in" syndrome. A clinico-pathological study of two cases. *Eur Neurol* 1976;14:81-9.
16. Easton J, Sherman D. Cervical manipulation and stroke. *Stroke* 1977;8:594-7.
17. Feldman MH. Physiological observations in a chronic case of "locked in" syndrome. *Neurology* 1971;21:459-78.
18. Gilroy J, Lynn GE, Ristow GE, Relcryn RJ. Auditory evoked brainstem potentials on a case of "locked in syndrome." *Arch Neurol* 1977;34:492-5.
19. Halsey JH, Downie AW. Decerebrate rigidity with preservation of consciousness. *J Neurol Neurosurg Psychiatry* 1966;29:350-5.
20. Hawkes CH. "Locked in syndrome": report of two cases. *Br Med J* 1974;4:379-82.
21. Hopkins L, Budney J, Spetzler R. Revascularization of the rostral brainstem. *Neurosurgery* 1982;10:364-9.
22. Kamshepolisky J, Danielson H, Flynn RE. Vertebral artery insufficiency and cerebellar infarct due to manipulation of the neck. *Bull Los Angeles Neurol Soc* 1972;37:62-6.
23. Karp JS, Hurtig HI. "Locked in" state with bilateral midbrain infarcts. *Arch Neurol* 1974;30:176-8.
24. Khurana RK, Genut AA, Yannakaris GD. Locked in syndrome with recovery. *Ann Neurol* 1980;8:439-41.
25. Kreiger B, Ozaki H. Vertebrobasilar distribution infarction following chiropractic cervical manipulation. *Mayo Clin Proc* 1980;55:322-32.
26. McCusker EA, Rudick RA, Griggs RC. Recovery from locked in syndrome. *Ann Neurol* [Abstract] 1980;8:91.
27. Marti Vilalta JL, Roic-Arnall C. Transitory posttraumatic "locked in" syndrome. *Lancet* 1977;2:1351-2.
28. Mehalic T, Farhat SM. Vertebral artery injury from chiropractic manipulation of the neck. *Surg Neurol* 1974;2:125-9.
29. Messert B, Orrison WW, Hawkins MJ, Quagliery CE. Central pontine myelinolysis. Considerations on etiology, diagnosis and treatment. *Neurology* 1979;29:147-60.
30. Mueller S, Sahs A. Brain stem dysfunction related to cervical manipulation. *Neurology (Minneap)* 1976;26:247-50.
31. Murray TJ. Locked in syndrome caused by a tumor. Another comment. *Neurology (Minneap)* 1976;26:1185-6.
32. Newman RP, Manning EJ. Hyperbaric chamber treatment for "locked in" syndrome. *Arch Neurol* 1980;37:529.
33. Okawara J, Nibbelink D. Vertebral artery occlusion following hyperextension and rotation of the head. *Stroke* 1974;5:640-2.
34. Paguirian A, Lefken EB. Central pontine myelinolysis. *Neurology* 1969;19:1007-11.
35. Plum F, Posner JB. Diagnosis of stupor and coma. Ed 1. Philadelphia: Davis, 1969:92-3.
36. Robertson J. Neck manipulation as a cause of stroke. *Stroke* 1981;12:1.
37. Roski R, Spetzler R, Hopkins L. Occipital artery to posteroinferior cerebellar artery bypass for vertebrobasilar ischemia. *Neurosurgery* 1982;10:44-9.
38. Schneider RC, Crosby EC, Russo RH, Gosch HH. Traumatic spinal cord syndromes and their management. *Clin Neurosurg* 1973;20:424-92.
39. Sherman D, Hart R, Easton J. Abrupt change in head position on cerebral infarction. *Stroke* 1981;12:2-6.
40. Stockard JJ, Rossiter VS, Wiederholt WC, Kobayashi RM. Brain stem auditory-evoked responses in suspected central pontine myelinolysis. *Arch Neurol* 1976;33:726-8.
41. Sundt TM, Piepgras DG. Occipital to posterior inferior cerebellar artery bypass surgery. *J Neurosurg* 1978;48:916-28.
42. Sundt TM, Piepgras DG, Houser O, Campbell J. Interposition saphenous vein grafts for advanced occlusive disease and large aneurysms in the posterior circulation. *J Neurosurg* 1982;56:205-15.