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Varieties of the Locked-in Syndrome

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Summary. The locked-in syndrome (LiS) was broken down on the basis of neurological symptoms in 12 patients. The criteria of classical LiS are total immobility except for vertical eye movements and blinking. If any other movements are present one should consider the condition as incomplete LiS. Total immobility, including all eye movements, combined with signs of undisturbed cortical function in the EEG led to the concept of total LiS. The anatomical basis for this condition consists of lesions in both cerebral peduncles which interrupt the pyramidal and corticobulbar tracts, the supranuclear fibers for horizontal gaze and the postnuclear oculomotor fibers. As to the course, chronic and transient LiS have been described.

Key words: Locked-in syndrome – Oculomotor disturbances – EEG in pontine lesions – Basilar artery thrombosis.

Zusammenfassung. Anhand der klinischen Symptome von 12 Patienten wurde das Locked-in-Syndrom (LiS) in das klassische LiS mit vollständiger Bewegungslosigkeit, ausgenommen vertikaler Blickbewegungen und Blinzeln, und in das inkomplette LiS mit weiteren Bewegungsresten unterteilt. Bei absoluter Immobilität mit Einschluß aller Augenbewegungen, jedoch Zeichen für ein erhaltenes Bewußtsein im EEG, wurde ein totales LiS angenommen. Die anatomische Grundlage für diese Form sind bilaterale Läsionen in den Pedunculi cerebri mit einer Unterbrechung der Pyramidenbahn, der corticobulbären Bahnen, der zentralen Fasern für horizontale Blickbewegungen und der postnukleären Oculomotorius-Fasern. Als Verlaufsvarianten wurden das chronifizierte und das transiente LiS unterschieden.

Introduction

A locked-in condition was first mentioned by A. Dumas in his novel "The Count of Monte Christo." The writer described "a corpse with living eyes." Despite several quite extensive reports in the medical literature [7, 21, 23, 47, 48, 75] the

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condition did not become popular among neurologists until Plum and Posner [66] introduced the term "locked-in syndrome" (LiS). The term denotes a neurological condition consisting of tetraplegia and paralysis of all cranial nerves except vertical eye movements [22, 61, 66]. Consciousness is fully preserved and can be demonstrated by voluntary blinking. The patients are able to communicate complex ideas by blinking Morse code [22]. Synonyms are pseudocoma or de-efferented state. Pathologically, ventral pontine lesions are the most frequent cause, but it is not appropriate to use the term "ventral pontine syndrome" as a synonymon since lesions in other locations may lead to similar conditions [44].

For theoretical as well as practical reasons one has to separate LiS from coma and prolonged coma-like states. The differential diagnosis from coma should be easy by means of the voluntary eye movements. Patient's awareness of himself and his environment is considered normal with LiS but seriously reduced or even abolished with akinetic mutism [10, 40, 61, 71, 72] or apallic syndrome [27, 39, 40, 45, 71]. The only way to prove these statements clinically is by the meaningful blink communication of LiS patients. The EEG is an important help but not conclusive per se as will be discussed later.

Since our interest directed to this not too rare brainstem syndrome, we have observed 12 cases. The present paper aims to describe the variety as well as the course of the symptoms.

Case Reports

The cases are summarized in Table 1. Exceptional cases are reported in greater detail.

Case 5. M.E., male, aged 41, was a known hypertonic for 10 years. On 7 July 1975 he had sudden violent headache and was found unresponsiveness shortly thereafter. He was considered comatose in a regional hospital. The cerebrospinal fluid (CSF) was bloody. On 20 July 1975 he was transferred to our clinic and found to be partially locked-in. He was able to signal by blinking that he was fully conscious from the very beginning of the disease. His eyes filled with tears on seeing his relatives. Tetraparesis was incomplete; he also could communicate via weak arm movements. He could look upward, otherwise no voluntary eye movements were possible. Perception of pain and temperature was diminished on the left side.

The EEG was slightly abnormal with diffuse 4–7 cps activity. The 9–10 cps α-rhythm was reactive to bright light. Cerebral panangiography revealed moderate diffuse sclerotic abnormalities.

He died suddenly the day after the admission to our hospital. The autopsy revealed an infarction within the upper ventral pons spreading to both cerebral peduncles for 2 mm. There was also recent hemorrhagic destruction of the entire pons and the adjacent medulla oblongata.

Comment. The patient experienced two consecutive pontine hemorrhages, the first one leading to a partial LiS with remnants of extremity motions.

Case 6. L.M., female, aged 71, was admitted to our hospital on 6 July 1975. No previous history was obtainable because of her unresponsiveness and the lack of any relatives. There was slight opening of both eyes on command and secondary to painful stimuli. There was questionable vertical movement of the right eye, otherwise no motions could be observed. The eyes were slightly divergent, the pupils pinpoint and unreactive. No communication was possible. Muscle tone was increased in the lower limbs, and plantar response was extensor bilaterally.

The EEG was moderately abnormal with diffuse 3–7 cps activity accentuated over both temporal regions. With passively opened eyes the 8–9 cps α-rhythm was attenuated. Her

Table 1. Clinical and pathological findings in 12 patients with locked-in syndrome (LiS)

Case and initials	Age (years)	Sex	Neurological status	EEG	Outcome	Pathological diagnosis
1. M.A.	68	M	Classic LiS, ocular bobbing, occasional groaning	Slightly abnormal, diffuse slowing	Died after 11 days	Basilar artery thrombosis, ventral pontine infarction
2. C.E.	44	F	Classic LiS, decerebrate posturing	Normal	Died after 3 days	Thrombotic occlusion of basilar artery, hemorrhagic pontine infarction
3. W.R.	42	F	Incomplete LiS, remnants of right arm motility, free eye movements, nystagmus, decerebrate posturing	Not done	Died after 34 days	Thrombotic occlusion of basilar artery, infarcted transsection at midpontine level
4. R.D.	65	M	Incomplete LiS, quadroparesis, decorticate posturing, ataxic breathing, skew deviation	Normal	Died after 15 days	Hemorrhagic infarction of pons
5. M.E.	41	M	Incomplete LiS	Slightly abnormal, diffuse slowing	Died the next day	Bilateral infarction of ventral pons and cerebral peduncles
6. L.M.	71	F	Total LiS	Moderately abnormal, diffuse slowing	Died after 4 days	Basilar artery thrombosis, infarction of both cerebral peduncles
7. B.H.	66	F	Total LiS	Normal	Died after 14 days	Pontine hemorrhage
8. S.A.	67	F	16 November 1977: Classic LiS 22 December 1977: Incomplete LiS	Normal	Still alive (Dec. 1978)	
9. B.M.	27	F	21 February 1977: Classic LiS, convergence preserved 20 April 1977: Incomplete LiS, eye movements free, palatinal, tongue, head and leg movements	Slightly abnormal, diffuse slowing	Still alive (Nov. 1977)	
10. P.R.	61	F	9 March 1978: Incomplete LiS, tetraparesis, some facial movements, skew deviation	Normal	Still alive. Moderate improvement, swallows, produces crude sounds, mimic reaction (June 1978)	
11. M.L.	46	M	Transient incomplete LiS	Moderately abnormal, diffuse slowing	Still alive. Marked improvement	
12. S.E.	67	M	Transient classic LiS	Slightly abnormal, diffuse slowing	Still alive. No permanent neurological deficit, further attacks of dizziness	

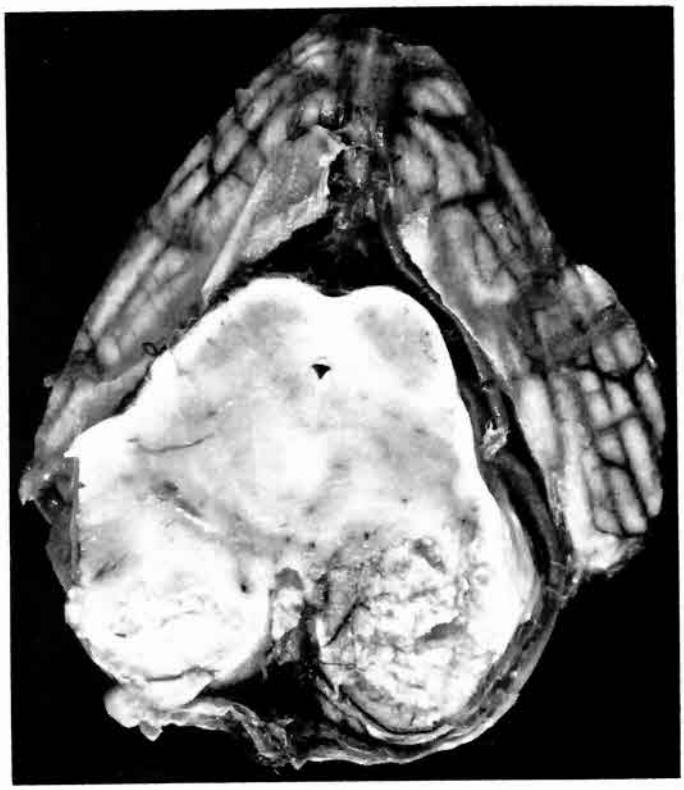


Fig. 1. Case 6. Transsection throught midbrain at level of III nerve nuclei. Infarcted areas in both cerebral peduncles, larger on left extending 10 mm in rostral direction. In caudal direction, infarction was in ventral pons. No abnormalities observed histologically 10 mm caudal to rostral pontine border

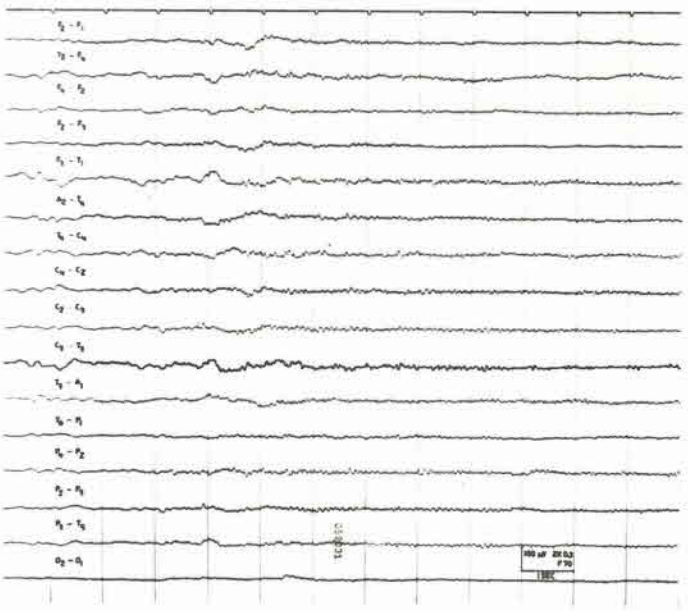
condition deteriorated on the next day. She was completely unresponsive, breathing became periodic and the muscle tone flaccid. She died 4 days after admission.

Subtotal thrombotic occlusion of basilar artery was found at autopsy. Both cerebral peduncles were destroyed by infarctions (Fig. 1).

Comment. Except for insignificant eye opening to external stimuli, this patient was totally unresponsive. The marginal movements and the reactive EEG α-rhythm made it possible to distinguish the condition from coma and to establish the diagnosis of total LiS.

Case 7. B.H., a woman aged 66, was under medical control for hypertonia for 5 years. On the day of admission she felt well until her husband found her unresponsive on the ground. She obviously had vomitted, was admitted cyanotic and remained in an unresponsive state. No spontaneous or reactive motions were observed. Only once, a mimic reaction was indicated to painful stimuli. Breathing was ataxic so artificial respiration was needed. The narrow pupils reacted slightly to light. The muscle tone was flaccid, tendon reflexes were normal and corneal reflexes abolished bilaterally. No pyramidal signs were present.

The CSF was bloody. Blood pressure varied frequently but was elevated to 220/120 mm Hg. Extreme sinus bradycardia was observed in ECG. The EEG (Fig. 2a) was normal with a reactive α-rhythm and normal sleep cycles. A normal driving response to intermittent photic stimulation was also observed (Fig. 2b).



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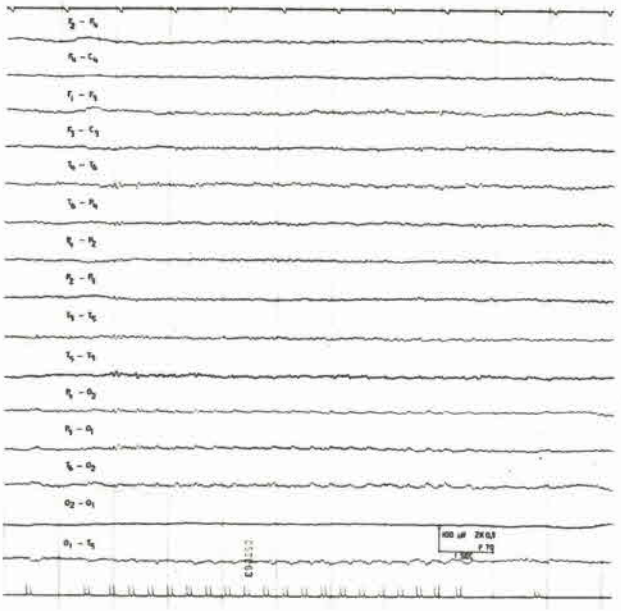

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Fig. 2a and b. Case 7. **a** Sleeping EEG on left section. After K complex 8—9 cps α-rhythm appeared over posterior regions. **b** Positive driving response to intermittent photic stimulation

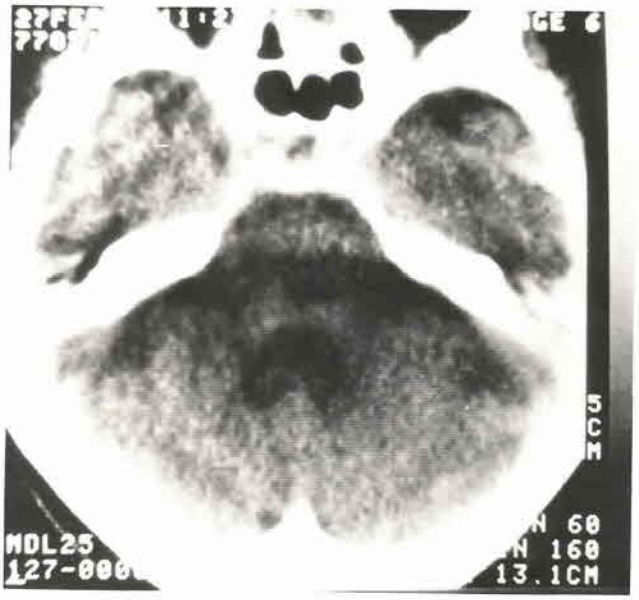

Fig. 3. Case 8. CT at pontine level. Symmetrical widening of fourth ventricle and small hypodense area within pons superimposed by hypodense artifact due to porus acusticus internus

Her condition did not change and she died 13 days after admission. The autopsy revealed extensive pontine hemorrhage with perforation into the fourth ventricle. More exact location of cerebral damage was not possible because of the bloody involvement of the entire brainstem.

Comment. With extensive pontine hemorrhage a totally unresponsive state was accompanied by normal waking and sleeping EEG indicating normal cortical function. This condition may also be called total locked-in syndrome.

Case 8. S.A., a woman aged 67, underwent an operation for varicose veins 14 days before admission. On 7 November 1977 she was found mute and motionless in her bed. On neurological examination there were some weak perioral and right arm motions, so a nearly classical LiS was diagnosed. Her physical condition was otherwise excellent.

CT was normal at this time, but 3 months later a hypodense area was evident within the upper pons and slight cortical and subcortical atrophy (Fig. 3). The EEG was normal. During the record yawning was noted by the EEG technician.

Summarizing the clinical and laboratory findings, a vascular accident of the ventral pons was assumed. The patient made a remarkable recovery. Palsy of horizontal gaze was transformed to an internuclear ophthalmoplegia (19 January 1978) and eventually normalized (20 March 1978). Paresis decreased gradually. During the first 2 months, the patient cried frequently in a crude and penetrating manner. On 20 March 1978, the patient pronounced single words, swallowed mashed food and moved all extremities but she could not stand or walk. Crying had disappeared. After this date, the progress was still going on but less dramatically.

Comment. This patient sustained an infarction of the pons leading to a nearly classical LiS. The condition became chronic but improved to an incomplete LiS.

Case 11. M.L., a man aged 46, was admitted with multiple injuries after an accident by a high pressure tube. He was comatose for 17 days with a posttraumatic midbrain syndrome. After intensive care including dehydrating measures he became alert but partially locked-in. The

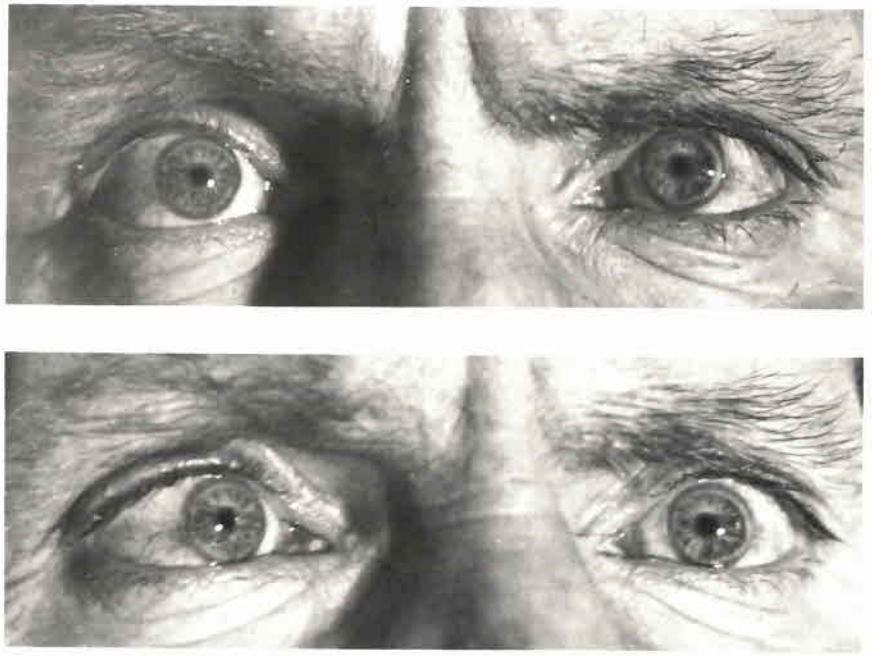

Fig. 4a and b. Case 11. Photographs of bilateral VI nerve palsy. **a** Attempt to look to right; **b** attempt to look to left. At rest both eyes squint toward the midline but each eye attains a midposition on attempting to look to the respective side

disturbances of ocular motility consisted of bilateral abducens palsy (Fig. 4). Weak facial, tongue and finger movements were observed, otherwise the patient was tetraparetic and mute.

CT revealed a small left frontal subdural hematoma not requiring operation. There was also a small hypodense area within the left upper pons. The EEG was moderately abnormal with slow (7 cps) basic rhythm and diffuse 2–6 cps activity. The basic rhythm was reactive to exogenous stimuli.

With physical treatment the patient made an excellent recovery. When discharged after 3 months, he had a slight diffuse brain syndrome with slight ataxia of gait. The ocular motor disturbance was unchanged. The remembrance for the locked-in condition was present but somewhat blurred.

Comment. Coma due to herniation after multiple brain concussions by an accident with a high pressure tube was followed by a partial LiS. Posttraumatic basilar artery thrombosis was presumed. The patient made an extensive recovery so the LiS was called transient.

Case 12. S.E., a man aged 67, was suffering from attacks of dizziness and optic sensations for some years. These attacks also occured on turning the head. After one attack he was unable to move or to speak but could open his left eye and gaze in a vertical plane. The right eye had been enucleated after a world war II injury. The patient's condition improved rapidly. After a few hours he spoke and moved all extremities although moderate tetraparesis was still demonstrable. Plantar responses were extensor bilaterally. The patient remembered all details of the locked-in condition and related vividly his feelings during his general flaccidity.

The EEG was slightly abnormal with diffuse 3—7 cps activity, accentuated over the temporal regions. X-ray of the cervical spine revealed marked arthrotic derangements with narrowing of the intervertebral foramina. Cerebral panangiography revealed multiple stenotic changes of both vertebral arteries.

The patient was discharged without major neurological deficit. He experienced further attacks of dizziness and abnormal optic sensations. Surgical treatment was considered but eventually rejected because of the diffuse vascular changes.

Comment. The patient had several attacks of basilar insufficiency, one of them leading to a classic LiS. This condition was completely reversible so a transient LiS due to a TIA of the basilar artery was assumed.

Discussion

Our observations indicated that LiS is not a homogenous neurological entity but has numerous variations. Total immobility except for vertical eye movements and blinking, combined with preserved consciousness, are essential for the diagnosis of *classical LiS* (Cases 1 and 2).

If any remnants of voluntary motion other than those mentioned are present, one should consider the condition as an *incomplete LiS* (Cases 3—5). The terminology has the same logical basis as the differentiation between complete and incomplete transsection syndromes with spinal lesions. Such motor remnants have been reported by several authors [57, 61, 66]. Improvements of a classical LiS lead to the incomplete form. This will be discussed with chronic LiS.

Total LiS (Cases 6 and 7) is of special interest not only for theoretical but also for ethical reasons. This condition consists of virtually total immobility including all eye movements combined with preserved consciousness i.e. existing inner monologue and awareness of external and internal stimuli as far as the corresponding pathways for sensory perception are spared by the lesion. How can this be demonstrated in a totally de-efferented patient? First of all, total de-efferentiation does not mean coma. Jouvet's experiments [40] with curarized probands brought final proof. But there is no way to arrive at a final decision by means of neurological examination in an irreversible de-efferented patient. The EEG seems to be of crucial importance in this connection. As a matter of fact, it was the EEG which raised the question about the existence of a total LiS.

In general, the EEG is a useful tool monitoring the level of consciousness. The reactive α-rhythm over the posterior regions indicates normal alterness whereas the amount of diffuse slow activity shows but statistical relation to the depth of coma. The EEG of LiS is normal or slightly abnormal with diffuse slowing [22, 37, 56, 57, 61, 78]. To be sure, α-frequencies in the EEG, even if they are predominant, do not necessarely indicate consciousness. They occur in coma after cardiopulmonary arrest [8, 15, 31, 69, 76, 78], after intoxication especially with sopoforic drugs [46, 62, 69] and after high electrical injuries [32]. They are also intermingled in the EEG of posttraumatic coma [53]. These α-frequencies are different from normal α-rhythm in terms of spatial distribution and reactivity.

Predominant α-frequencies are also seen in clinically unresponsive states after brainstem lesions [13, 52, 55, 59, 67, 78]. They were accentuated over the fronto-temporal regions and mostly unreactive to various stimuli. The conflict between the normal looking EEG and the comatose behavior led to the supposition of two separate neuronal populations subserving alertness and synchronization of α-rhythm [25, 78]. However, in several unresponsive patients with brain stem

lesions there were hints in the EEG of preserved consciousness. The α-frequencies were found reactive to passive eye opening [51, 78], and a positive driving response to intermittent photic stimulation [52, 78, 80] or NREM sleep periods alternating with α-rhythm have been recorded [55, 78, 80]. Clinical responsiveness has been reported in a few apparently comatose patients [42, 48]. Lhermitte's [48] patient showed the clinical picture of deep coma; no voluntary eye movements were present, but the patient could signal his alertness by Morse code with weak movements of his wrists. Several authors stress the difficulty of demonstrating alertness in unresponsive patients and discuss the existence of a condition we call total LiS [9, 12, 32, 54, 80]. Cases 6 and 7 showed EEG signs of alertness. Case 6 also had residues of eye movements. We presume that both patients were not unconscious but totally de-efferented. Localizing aspects will be discussed below.

The LiS is a very serious condition, and most patients will be released from their suffering after a few days. However, some patients survived the acute phase and became chronic. There is usually moderate improvement. *Chronic LiS* has been reported by Feldman [22], Markand and Dyken [57] and Nordgren et al. [61]. Cases 8—10 meet the criteria of chronic LiS. The remarkle improvement in Case 8 is unique.

Reports about LiS as a *transient* incident are scarce. Only Hawkes and Bryan-Smith [37] described a patient with periodes of total unresponsiveness but a normal EEG during these periods. Therefore, the authors presumed TIA of the basilar artery. The same diagnosis was made in Case 12. This patient was especially instructive because of his ability to tell about his experiences during the locked-in state afterwards. Case 11 represents a presumed posttraumatic basilar artery thrombosis with an excellent recovery.

Autopsy studies revealed brainstem damages in all reported cases of LiS. The lesions were localized mostly in the ventral part of the pons [7, 22, 37, 56, 57, 61, 66, 80]. Bilateral lesions of cerebral peduncles at the mesencephalic level are also known to produce LiS [20, 44]. Cases 5 and 6 (Fig. 1) are further examples. In all cases, no bilateral extensive destruction of the tegmentum rostral to the ponto-mesencephalic junction was be observed.

Neuronal structures necessary for the maintenance of normal consciousness are the cerebral cortex for higher integrative functions, the ascending reticular activating system for vigilance and the connecting fibers of both systems. The brainstem reticular formation includes the areas of grey matter in the bulbar tegmentum, pons and mesencephalon but excludes the cranial nerve nuclei. Its cranial limits are not clarified [40]. What part of the activating reticular ascending system must be destroyed, and how big must the lesion be to cause coma? There is much agreement between experimental works in animals [3, 24, 49, 50], amytal-injection experiments in man [1] and clinicopathological observations [12, 34, 63, 68, 68a, 73]. Brainstem lesions will not produce coma unless they pass the pontomesencephalic junction in the rostral direction and affect the tegmentum bilaterally. This statement is clearly confirmed by the autopsy findings in LiS. This holds also in the cases with bilateral lesions in the cerebral peduncles.

In order to explain the oculomotor disturbances with LiS it is necessary to epitomize the neuroanatomical basis. The following statements are extracted from the comprehensive reviews of Bender [4, 5], Clara [16], Plum and Posner

[66] and Walsh and Hoyt [77]. Pathways serving voluntary eye movements start at the frontal eye field (area 8α, β, ϑ) and descend within the corticobulbar tract. Projection fibers of the III, IV and VI cranial nerve have been traced through the genu of the internal capsule into the anterior third of the cerebral peduncles. They begin to leave the pyramidal system at the III nerve level. Furthermore, there are other nonpyramidal connections, and the pathways, especially for vertical movement, are not completely understood. No projection fibers enter the oculomotor nuclei directly but synapse with neurons of the mesencephalic and brainstem reticular formation. The postulated existence of a paraabducens nucleus [18, 65] has not been confirmed by others [11]. However, there exists a system of supranuclear integration within the *paramedian pontine reticular formation* (PPRF) [2, 17, 29, 77]. The medial longitudinal fasciculus is a further integrating system and is believed to be subordinated to the PPRF system [70].

The most common oculomotor disturbance of LiS is bilateral palsy of horizontal gaze. This symptom is indicative of a brainstem lesion [5, 77] and closer localization is within the PPRF [2, 6, 17, 29, 38, 41]. Bilateral lesions in the PPRF have also been held responsible for palsy of horizontal gaze in LiS [20], but lesions in the medial longitudinal fasciculus and VI nerves could not be rouled out completely. With palsy of horizontal gaze, vestibulo-oculomotor functions are mostly abolished. In some patients vestibulo-oculomotor reactions were preserved [9, 35, 58]. The dissociation of voluntary and induced horizontal eye movements was also observed in LiS [61], so there is good reason to assume that paralysis of horizontal gaze is produced by various lesions of the supranuclear fibers, of the PPRF, of the medial longitudinal fasciculus or the VI nerve nuclei. The observation of bilateral VI nerve palsy in Case 11 is further evidence for this view. The transformation of bilateral horizontal gaze palsy to an internuclear ophthalmoplegia (Case 8) is notheworthy insofar as internuclear ophthalmoplegia is believed to be due to lesions in the medial longitudinal fasciculus [70]. Considering the extent of damages with vascular ventral pontine syndromes, it seems not reasonable to confine the lesion producing the paralysis of gaze to the PPRF.

Beside the horizontal gaze, there are several other oculomotor disturbances with LiS. Case 1 exhibited ocular bobbing. This symptom consists of sudden conjugate downward excursion of the eyes from their resting position and is associated with palsy of lateral gaze. The phenomenon is seen with pontine lesions [9, 23, 58, 60], which may also produce LiS [9, 20, 23, 57, 61, 80]. It is rarely observed with metabolic encephalopathy [64, 74]. Ocular bobbing indicates dysfunction of pathways serving vertical eye movements, but there are no verified facts about the pathophysiological mechanism [19, 36].

Further examples of oculomotor dysfunction with LiS are preserved vertical gaze and convergence [61; Case 9], skew deviation of eyes [57; Cases 4 and 10], hampered or obolished upward vertical gaze additional to palsy of horizontal gaze (Cases 4 and 10) and total immobility of one eye [14].

Karp and Hurtig's [44] patient with infarcts in both cerebral peduncles was tetraparetic, but the eye movements were totally undisturbed. In contrast Case 6 exhibited only very slight remnants of ocular motility. In this context, the question of localization of total LiS arises. In Case 6 the oculomotor immobility

may be explained by interruption of supranuclear pathways serving lateral gaze combined with partial destruction of postnuclear III nerve fibers. Prenuclear vertical gaze palsy results from bilateral lesions involving the pretectum and the midportion of the posterior commissure [5, 6, 43]. These structures were intact in Case 6. Therefore the immobility must be due to peripheral III nerve palsy. The pinpoint pupils in this patient are somewhat confusing. One might speculate that the postnuclear parasympathetic fibers were spared by the lesion because of their location near the midline. Pinpoint pupils with pontine lesions are believed to result from parasympathetic irritation in combination with sympathetic interruption [77]. Sympathetic descending pathways are located within the prerubral field and the red nucleus capsule [81]. These structures were certainly affected in Case 6. Therefore, Walsh and Hoyt's [77] explanation may be applicable.

To summarize, a totally de-efferented state with preserved consciousness seems possible with lesions in both cerebral peduncles interrupting the prenuclear paths for horizontal gaze and the postnuclear III nerve fibers. Case 6 demonstrates this possibility. The lesion in Case 7 was too extensive for discussion about localization.

A vascular etiology is most likely with LiS. Carful history taking often yields precursor symptoms like episodes of dizziness, slurred speech, double vision, ataxia and vomiting. Autopsy confirmed the vascular etiology. Cases 1, 2, 3 and 6 had brainstem infarcts due to basilar artery occlusion and Cases 4, 5 and 7 had pontine hemorrhages. Vascular accidents were also presumed in the surviving patients, corroborated by CT (Case 8, 10 and 11), by angiography (Case 12) or by both examinations (Case 9). Brainstem infarcts secondary to basilar artery thrombosis were the most frequent cause in the literature [7, 20, 37, 44, 57, 61, 80] followed by pontine hemorrhage [57, 61]. Some of Dinsdale's [21] cases with spontaneous hemorrhage into the posterior fossa also meet the criteria of LiS. Rarely does a tumor cause LiS [14, 30, 37]. The connection of LiS with heroin abuse [33] is only partially understood. A hypersensitivity similar to the transverse myelitis associated with heroin or opium abuse has been suggested.

The vascularization of the brainstem is so variable that syndromes cannot be identified appropriately by the name of the suspected artery [28]. Among 18 patients with basilar artery occlusion no distinct pattern of tissue destruction could be predicted [47]. Gauthier [26] found in his cases of basilar thrombosis bilateral infarcts of the ventral pons with sparing of the dorsal part. This pattern of lesion is commonly seen with LiS. What is the anatomical basis for the sparing of the tegmentum pontis in cases of basilar artery occlusion? For embryological, anatomical and pathological reasons Biemond [7] assumed that the tegmentum pontis is almost entirely supplied by the carotid arteries via the superior cerebellar artery. Therefore, basilar artery occlusion will not affect the tegmentum provided that the circulation is intact in the carotid artery system.

To summarize, a vascular accident, especially basilar artery thrombosis, is the most probable cause of LiS, and some kind of LiS is the most probable condition to occur with occlusion of the basilar artery.

Several stereotyped movements, which were incompatible with the extent of paresis of voluntary movements, were observed in the cases reported. The interesting phenomenon of involuntary motor pattern with LiS will be the subject of an other paper.

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