"Locked-in" Syndrome with Dissociated Nystagmus and Vertical Head Position Nystagmus

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Plum and Posner[1] use the term "locked-in" syndrome to describe a patient who, with bilateral basis pontis infarction, had no motion at all other than vertical eye movements. their patient was alert and communicated intelligently by using his eyes, differing from the classic akinetic mute who has the ability to move his extremities but usually does not do so.

Meanwhile, the term "akinetic mutism" was coined by Cairns et al[2] in 1941 to describe a patient with an epidermoid cyst of the third ventricle. In the original description, the patient did not speak or move but followed the observer with her eyes. "Usually there are no movements of a voluntary character" though the patient could be made to carry out simple commands and would withdraw from painful stimuli.

The purpose of this paper is to present a patient who had "locked-in" syndrome like an akinetic mute, but were alert and totally paralyzed save for mesencephalic-controlled eye and lid movements, their only means of communication, and to present the otoneurological comments on the pathophysiological mode in the central nervous system.

CASE REPORT

A 42-year-old man was intermittently treated for hypertension by a nearby doctor for last 4 years. Following sudden loss of consciousness, he was admitted to Emergency department of the Hospital where neurological evaluation revealed him to be comatose with difficulty in breathing.

Last midnight prior to the admission (October 28, 1972), he began to complain "abnormal sensation inside the head" and "numbness of the left arm". Shortly after he asked his wife to call his brother's help, he lost his consciousness and became unresponsive and he was in deep sleep with heavy snorring.

Physical examination at the emergency department revealed the patient was comatose and some difficulty in breathing with strong stridor. Pupils was anisocoric, and the right pupil was larger than the left. Response to the light was lost on both sides. Response to stick stimulation remained.

Respiration was irregular. Blood pressure was 180/120 mmHg. Auscultation and percussion was normal in the lungs and heart.

According to the request by the neurologist, tracheotomy was done immediately and the tracheal intubation through the tracheostome was done. He was placed on an automatic respirator.

A flaccid quadriplegia was present, and no response to stimulation of the trunk and limbs was observed in the following day.

Deep tendon and planter reflexes could not be elicited.

Thenafter, he was survived and was continuously maintained with the respirator, but unconscious status was unchanged.

Nasogastric feeding tube was placed and tracheostome was not closed because of disturbance of the swallowing and expectoration of the saliva or sputa up-to-date for last two and half years.

During his long hospitalization, he was in the status of so-called akinetic mutism with clinical diagnosis of the cerebral hemorrhage. His quadriplegia remained unchanged but he began to show somewhat voluntary eye movement (See Table 1, for summary of progress record concerning to the eye movement in broad sense).

Table 1. Progress record concerning to the eye movement.

11th day after onset	"Questionable movement of the left eyelid"
26th day	Blink-like movement of the eyelid, confirmed.
32nd day	He seemed to be able to open and close his eye by request, probably; reported.
39th day	Right oculomotor palsy (abducens and trochlear) suspected by ophthalmologist.
45th day	Nystagmus toward the right, horizontal or horizontal-rotatory (counterclockwise) was sometimes observed on only the right eye.
87th day	Right facial paralysis definite and preventive care for the corneal dryness, continuously.
96th day	Spontaneous nystagmus toward left, horizontal in character, was observed, sometimes. And downward vertical nystagmus was observed, too. But, no definite voluntary gazing was observed.
3 months later	He blinked two to three times when he was asked to close his eyes. But,he could not close the right eyelid.

Otoneurological evaluation was requested on the 96 th hospital day. Although thorough examination was hard to go over him at bedside in this akinetic mute, the report was made as follows: Right eye was covered with eye pad dressing. Right eye lid was not shut and not moved, showing right facial paralysis. Observation of nystagmus was tried to do on the left eye, only revealing that somewhat nystagmic eye movement to the leftward and some vertical downward nystagmus was observed on that date. No definite voluntary gaze eye movement of the left eye was observed. Ears, nose and throat were ordinarily normal finding, but ordinary audiometry was not performed.

Otoneurological examination was done at bedside of the patient of the persistent "locked-in" syndrome on August 29, 1974 (one year and ten months after the onset). Otoscopic examination showed mild retraction of the tympanic membrane; otherwise normal. Peculiar dissociated spontaneous nsytagnus was observed such as the left eye showed horizontal and jerking nystagmus to the left and as the right eye showed right beating nystagmus of less frequent but relatively regular and small amplitude in character (Fig. 1). These nystagmus and eye movements were recorded by electronystagmography using surface disc electrdoes placed above, below, and at the inner and outer canthus of each eye; vertical lead and horizontal lead of each eye, respectively (Fig. 2).

Downward vertical head position nystagmus was elicited following a latency of about 11 seconds after his rightside head was down (Fig. 3). When his leftside head was down, the upward vertical nystagmus appeared following a latency of several seconds of proceeding horizontal rightbeating nystagmus.

Results of caloric stimulations to the external ear canal (by modified Hallpike method) was shown in Fig. 4.

Caloric testing with cold water (30°C) in the left ear produced calorically responsed nystagmus and/or eye movement but very peculiar in character and in its direction. Those are shown in Fig. 4.

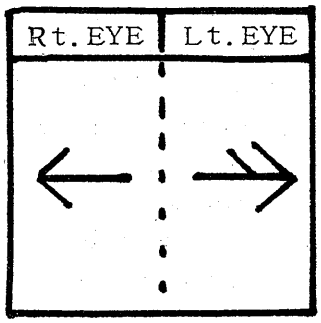
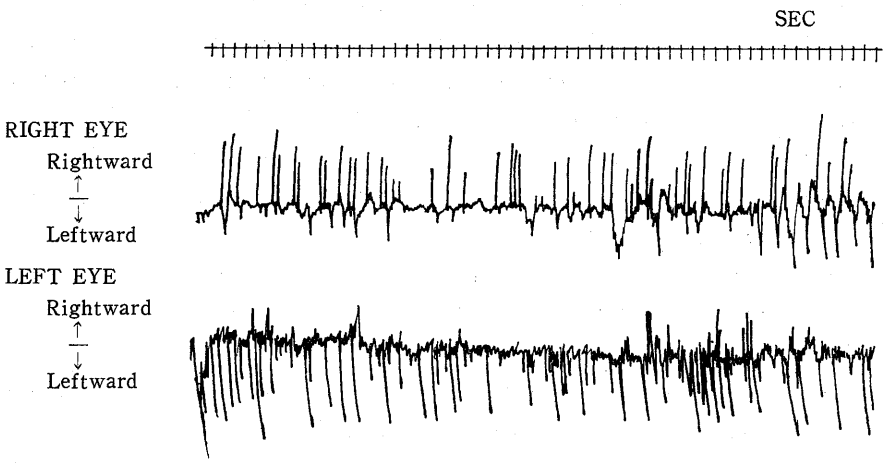


Fig. 1. DISSOCIATED SPONTANEOUS NYSTAGMUS



Simultaneous ENG recording (Traced from original ENG)

Fig. 2. DISSOCIATED SPONTANEOUS NYSTAGMUS

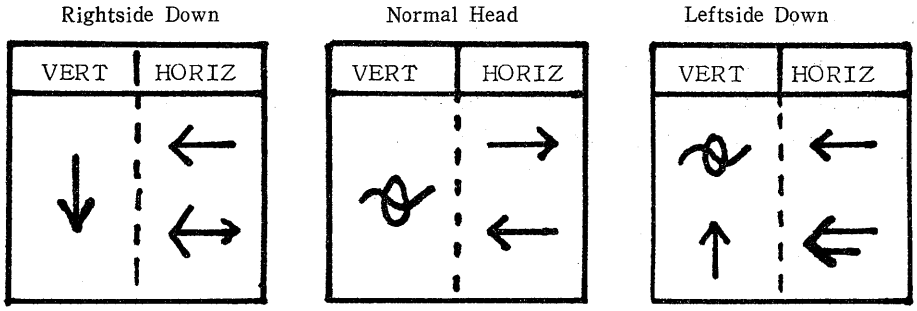


Fig. 3. NYSTAGMUS and EYE MOVEMENT affected by HEAD POSITION (VERTICAL NYSTAGMUS elicited)

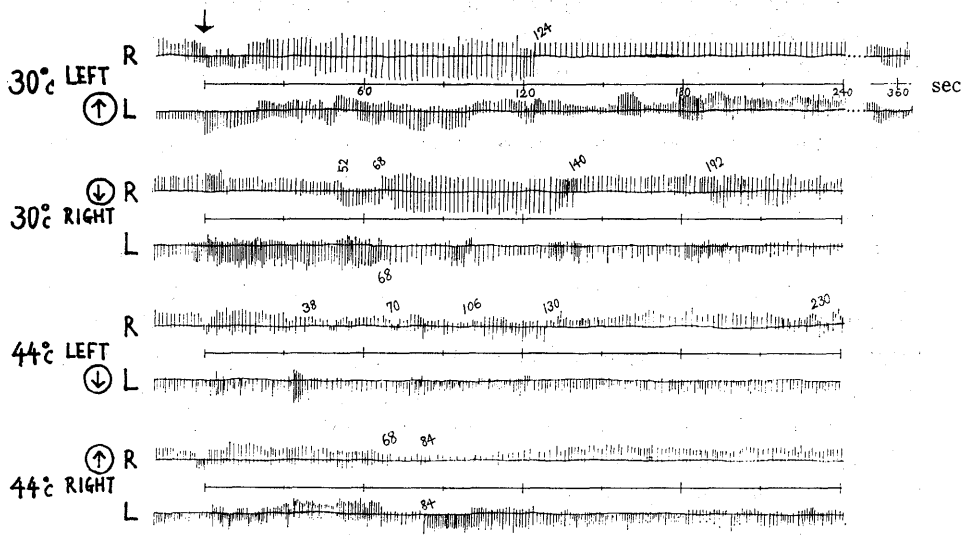


Fig. 4. Schematic presentation of Electronystagmograph recorded by horizontal lead of each eye, and induced by Caloric stimulations (Alternate cold and warm caloric test).

Note: 1) R=Right eye ENG horizontal lead.
L=Left eye ENG horizontal lead.
2) Arrow indicates the time of caloric stimulation started for 10 seconds.
3) For example, 30°C LEFT ⓘ means cold water (30°C) stimulation to the left ear, and expected direction of induced nystagmus to right-ward (pen moves to upward).

COMMENTS

These clinical results mentioned before indicate that this was the case of "locked-in" syndrome of speechless, motionless at all except for voluntary vertical eye movement, probably caused by bilateral lesions in the pons due to infarction (or relatively small hemorrhage) of basilar artery in the tegmentum of the rostal part of midbrain. These pathologic occurrence might be related to his chronic hypertension in his age for last 5 years.

It is reasonable to presume that although the lesion in the brainstem was bilateral and wide enough to cause this serious status of "locked-in" syndrome at the beginning of sickness, the imagined lesion of nowaday might be well getting smaller than the expected at the early period because there are several clinical signs to suggest "fairly working nuclei in the brainstem".

It is quite possible to imagine this from the literature of Dr. Feldman[3] who reported a chronic case of "locked-in" syndrome, in which patient had learned Morse code from her husband, seven months after accident, utilizing jaw movements and eye blinks to signal "dots" and "dashes". He explained that over the course of the next two to five months, the patient gradually recovered most functions of the motor cranial nerves, including neck extension.

On the basis of available clinical and pathological evidences of Nordgren, et al[4], who presented seven collected cases and who discussed intensively the results in 4 cases of them from the clinical and pathological standpoint of view, it seems likely that essential changes in the central nervous system are bilateral involvement in the basis pontis, extending from the caudal pons to the pontomesencephalic junction; caused by any pathology, i. e., infarction, hemorrhage and compression or ischemic changes in medulla, pons and/or midbrain.

The neurological signs and presumed pathological lesions in this case would be summarized to outline the imagined spread of the lesion (Refer Fig. 5. Table 2 and 3). Lower limit of the lesion might be at least above the medullary respiratory center which is located in the caudal pons. Upper limit might be around the lower margin of the pretectal area which functions vertical gaze. Involvement might not occupy total oculomotor nuclei and trochlear nuclei, but abducens nuclei, especially on the right side might be destructed including the right facial nucleus and its pathway and pontine tegmental horizontal gaze zone. In this midposition of the pons, cochlear and vestibular nuclei remained enough and are functioning fairly well, at least since the elicited caloric nystagmus are seen in spite of markedly distorted and also seemingly hearing acuity remained. Another main feature was dissociated eye movement and nystagmus with vertical head position nystagmus, those which might be caused by mesencephalic tegmentum including medial longitudinal fasciculus.

Finally, it may be concluded that a main focus of lesion will be located in the midpons and rostal and ventral zone on both side, possibly heavier lesion on the right side.

Noda, et al[5] recently reported a case of "locked-in" syndrome and made the literature review, and they found their paper will be the first paper in Japan.

Meanwhile, we must recongnize there is some tendency to increase the number of patient of akinetic mutism or so-called "vegetable-like patient" in many hospitals through the nations.

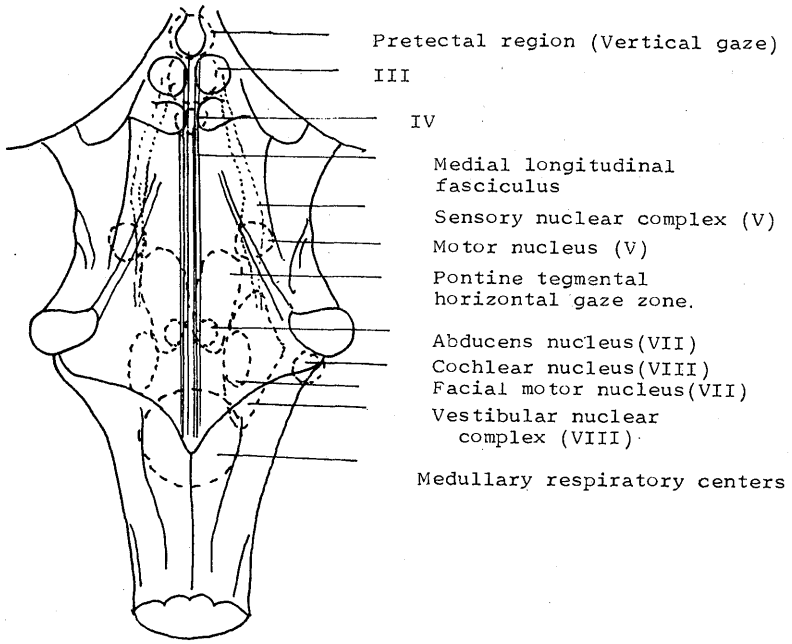


Fig. 5. Anatophysiological Consideration in the brainstem.

Table. 2. Dominant Clinical Manifestation of Symptoms —concerning to "locked-in" syndrome—

Acute quadriplegia	——	Involvement of transsection of cortico-spinal tract in the brainstem.
Aphonia and difficulty in swallowing	——	Lower corticobulbar tract, transsecting involovement.
Vertical eye movement, remained intact or sufficient to function	——	Supranuclear oculomotor tract, required to be intact.

Table 3. Peculiar Nystagmus and Localization of pathlogy, relating the Present Case.

Gaze paretic horizontal nystagmus	——	Pons and Tegmentum lesion
Dissociated and Non-conjugate Nystagmus	——	Pons and Mesencephalic tegmentum (Medial longitudinal fasciculus)

SUMMARY

"Locked-in" syndrome, similar to akinetic mutism but not same, was presented from the case of 42-year-old man who had not motion at all other than vertical eye movement by himself. The preesent case is unusual in that the syndrome has persisted for over two years and 10 months while the patient's general condition remains in good.

Otoneurological findings and comments on pathology in the central nervous system were presented.

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REFERENCES

1) Plum, F. and Posner, J. B.: The Diagnosis of Stupor and Coma. F. A. Davis, Philadelphia, 1966.
2) Cairns, H., Oldfield, R. C., Pennybacker, J. B. and Whitteridge, D.: Akinetic mutism with an epidermoid cyst of the 3rd ventricle. *Brain*, 64 ; 273-290, 1941.
3) Feldman, M. H.: Physiological observation in a chronic Case of "locked-in" syndrome. *Neurology*, 21 ; 459-478, 1971.
4) Nordgren, R. E. et al: Seven cases of Cerebromedullospinal disconnection; The "locked-in" syndrome. *Neurology*, 21 ; 1140-1148, 1971.
5) Noda, Y., Okayama, M. and Ogata J.: A case of locked-in syndrome. *Clinical Neurology*, (*Tokyo*) 14 ; 431-436, 1974.