

The Locked-In-Syndrome by Philippe VAN EECKHOUT ( 1997)

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The Locked-In-Syndrome (L.I.S.) is a rare neurological disorder which often puzzles the doctor. It may occur at any time during daily life. It tests our knowledge and illustrates the limitations on what we can achieve. It poses a medical and economic problem but, above all, a human one.

The first description of a state resembling L.I.S. occurs in literature. In 1884, Alexander DUMAS in The Count of Monte Christo, described this condition as : "a dead body with living eyes". His character, Mr NOIRTIER de VILLEFORT, victim of a cerebral attack, survived and learned to communicate by closing and opening his eyelids and by vertical movements of his eyes. Several years later, in 1868, Emile ZOLA wrote about a woman " mute and paralyzed, her face a death mask with two living eyes. Only these moved, rolling rapidly in their sockets".\_

Medical books talked about this syndrome only in 1941. In 1947, for the first time, a neurologist and a neuro-surgeon diagnosed a case of Locked-In-Syndrome. Their study cited an injury to the vertebral artery resulting from a stroke to brainstem following a manipulation of the cervical vertebrae by a chiropractor.

In 1966, the term "Locked-In-Syndrome" was introduced by PLUM and POSNER. Literally, this term means "locked in the interior". PLUM and POSNER defined the Locked-In-Syndrome as supranuclear motor disafferentation which produces a paralysis of the four limbs and of the last cranial nerves while not interfering with consciousness. The paralysis of the motor pathways hinders oral or gestural communication.

Other names were proposed such as : ventral pontine syndrome, state of supranuclear motor disefferentation, false coma by cerebro-medullary spinal disconnection.

The Locked-in-syndrome results from an extensive injury to the brainstem, most often at the level of the pons. The injury is to the long pathways which crisscross brainstem as well as to the vital nerve centers and the nuclei of certain cranial nerves from the third (III) to the twelfth (XII) pairs. The reticular formation is generally preserved. Clinically, the damage is to the pontine base extending, in varying degrees, to its top. This accounts for the motor deficiency rendering the patient tetraplegic due to the bilateral interruption of the descending corticospinal motor pathways. The majority of L.I.S. patients suffer from rigidity of decerebrate posturing (extension of the arms and legs), either spontaneous or caused by a painful stimulus.

In the initial phase, these patients additionally have major respiratory problems associated with breathing insufficiency. Consciousness and vigilance continue to exist. Therefore, it is necessary to think about determining the only voluntary movements which will allow the patient to communicate such as :

- opening the eyes (done by raising the upper eyelid),
- blinking the eyelids,
- vertical movement of the eyeballs.

The deficit of the last pair of cranial nerves is evidenced by weakness of the facial, tongue and pharyngeal muscles with anarthrie. Problems with swallowing and making sounds are great. In certain cases, swallowing occurs automatically when water or other liquids are placed in the patient's mouth and saliva is produced. Likewise chewing and sucking movements can be induced as reflex actions by oral stimulation. Troubles with eye movements depend essentially on the exterior of the pontine injury and on whether the injury is on one side or both sides. The most usual eye problem is paralysis of side to side movement but not a vertical movement. This is because the upper part of the mesencephalic structure has been preserved.

In the majority of L.I.S cases, the only voluntary spontaneous muscular movements are :

- opening and closing the eyelids,
- simultaneous vertically eye movements.

These movements are possible thanks to the partial preservation of nerve XI and of the mesencephalic reticular substance.

Pupils are generally small but they react to light. Although convergence may be affected, it is often normal but there may be abnormal movements. The most typical is "eye bobbing", a brisk deviation of the eyes toward the bottom and then a slow movement upward toward the resting position. The diagnosis of the L.I.S. must be clinical. It is often postponed until after a period during which the patient is thought to be in coma.

PLUM and POSNER emphasize the confusion that sometimes arises between the L.I.S. and akinetic mutism. There are however different conditions :

- Akinetic mutism is a subacute or chronic alteration of consciousness with cycles of awakening-sleeping. Patients look awake but do not speak or move, and this cannot be explained by damage to motor pathways as is the case of L.I.S.

The akinesia is recognizable by the absence of voluntary spontaneous movements, of movements initiated by command, even as a result of painful stimuli.

- Absence of speech and of mobility of limbs contrast with the presence of certain signs of wakefulness. Patients open their eyes spontaneously or after stimulation. Eye movements are joined together, sometimes appearing to follow a visual stimulus (blinking at something menacing).

Akinetic mutism concerns the frontal lobes and the reticular formation in varying degrees. PLUM and POSNER go on to state that is necessary to take care to distinguish between a spirit burdened by a body which cannot be made to obey and a typical akinetic mutism in which consciousness has been altered.

The prognostic for a L.I.S. is generally reserved and pessimistic; In the acute stage, the treatment of such patients requires much reanimation and nursing care. In fact, treatment in reanimation or intensive case is of an exceptional quality. It must be combined with psychological assistance and, above all, information about the short-term therapeutic project. The care-givers must not forget that their patients are conscious and able to communicate.

Treatment is based on specific reanimation measures on one hand and, on the other in certain cases, on special care directed to the cause. Treatment of the symptoms is necessary in all cases to insure that the vital functions are maintained. This is particularly true for ventilation where there is a need for respiratory assistance by incubation and then by a tracheotomy and repeated bronchial aspirations. Regular respiratory physiotherapy is indispensable. This must be carried out during the acute stage and sometimes for months thereafter.

Ventilation by the lungs is reduced and coughing is unproductive. This results in tracheobronchial congestion. In this case, measuring vital capacity is an important part of the examination. Deficiency in coughing together with problems of swallowing present a risk of tracheobronchial flooding since the respiratory command is neither automatic nor voluntary. While automatic bulbar control may remain, it is impossible to voluntarily modify respiratory rhythm and amplitude. This means that there is a bilateral interruption of the passages by which cortical control is exercised. Finally, speaking about *hemodynamic*, a drastic fall in arterial pressure must be avoided. The reassuring competence of reanimation services stops once the patient has attained respiratory independence with a tracheotomy and also cardiac autonomy.

For administrative reasons, the L.I.S. patient is forced to leave reanimation or intensive care "to be placed", in the best cases in a functional rehabilitation center or in a center for chronic traumatic patients or for the aged. Often such type of new center is "unfindable" despite the best efforts of the family and the social welfare worker.

If it can be found, the L.I.S. patient must receive continued care for the symptoms previously described. A second therapeutic project must be to improve his respiratory autonomy and to try to reestablish his swallowing. The two most important people to do this are the physiotherapist and the speech therapist, under the surveillance of the doctor in charge. Cooperation between these two therapists is always beneficial and morale-building for the patient.

**SWALLOWING** : This act in its entirety assures the transfer of solid and liquid food and of saliva from the mouth to the stomach by way of the pharynx and the esophagus. The influx which triggers off the nerve movements of swallowing travels the length of the motor and sensitive fibers of six pairs of cranial nerves. The

sensitive and sensory information is conveyed to the corresponding cranial nuclei situated primarily at the level of the *bulb rachides* in the brain stem.

These nuclei may be activated by the cerebral cortex for "voluntary swallowing" or by the peripheral receptors in the mouth and the pharynx for "reflex swallowing".

Usually, there are three phases in swallowing :

1. The time in the mouth, oral or oropharyngeal, is the preliminary phase. It is a conscious complex motor action which can be set off voluntarily or as a reflex action.
2. The pharyngeal time : the air pathway closes and the digestive one opens. Closing of the epiglottis by the action of the vocal cords protects against "false roads" which could cause pneumonia by aspiration or even death by suffocation, one of the usual causes of death of L.I.S. patients. Respiration is suspended during this conscious phase even though it is a reflex action and involuntary.
3. The time in the esophagus on the way to the stomach is an involuntary, unconscious, reflex action.

Swallowing, despite its frequency and its usual character in normal people, is highly sophisticated and closely connected with other vital functions :

- breathing.
- salivating.
- chewing.

Swallowing and breathing are coordinated in order to protect the respiratory tract. Swallowing is preceded by a short intake of breath and the apnea continues during the pharyngeal time. Breathing begins again only during the third phase of swallowing. There is a structural and functional neuro-physiological interdependence between swallowing and breathing. Since both use the same passage (the oropharynx), coordination between the two functions is vital in order to avoid "false roads".

Therefore, with L.I.S. patients, work on swallowing begins by introducing a drop of water into the mouth, placing it between the lips and directly on the tongue, using a syringe when the mouth opening is too small. Most of the time, the mouth is totally inactive due to the sonde and the tracheotomy. Thus it is a drop of water which first permits reanimation of the face and, in the second step, facilitates

relaxation of the oral-facial muscles. Once these muscles are relaxed, the patient becomes conscious of the existence of his laryngeal muscles and their functioning. Thus, the tongue, the soft palate and the pharyngeal pillars are stimulated. Often a fit of coughing ensues and this requires that the patient be suctioned in order to free him from the obstructing mucus.

Most often, L.I.S. patients have serious respiratory complications. Pulmonary hygiene and respiratory reeducation are constant concerns both in the hospital and after leaving it. Anomalies of the respiratory function include partial or total paralysis of the thoracic and abdominal muscles deficiencies in the volume of air exhaled and inhaled, and also a non-synchronization between respiratory and laryngeal movements. Good breathing is that which can easily vary to adapt to different circumstances as required. The form of respiration results from three factors : rhythm (slow, rapid), type (abdominal, thoracic, mixed), and amplitude. It is possible to adapt automatically to respiratory needs.

Working with breathing helps the patient to return to a normal respiratory rhythm and to develop his respiratory capacity by aiding him to exhale through the mouth. Thus, the mouth speech therapist or the physiotherapist must make him conscious of the difference between exhalation and inhalation. During inhalation, air enters the lungs. It leaves during exhalation and causes the vocal cords to vibrate. The therapist presses on the thorax at the patient's rhythm and then, rapidly, uses different rhythms : slow and fast. By pressing on the thorax, he forces the patient to exhale. Then he releases this pressure, inhalation is automatic. The therapist's action on exhalation tends to increase the amplitude of the respiratory movement. The aim is to prolong the exhalation long enough to obtain a sound.

The voice may be thought of as a sound exhalation . In phonation, exhalation is active. Air is pushed out of the lungs by the action of the expiratory muscles. The active exhalation necessary to produce the voice is called phonatory breath. The vibration of the vocal cords permits the emission of noise, of sounds which lead to language. The L.I.S. patient has lost all the control of the motor command allowing him to speak. The work of reeducating the phonatory breath is carried out with the help of the respiratory exercises. It requires active participation from the patient and a

lot of energy from the speech therapist. It is important that the therapist transmits the patient his own hope of seeing the patient escape from the "locked-in" state.

At first, the therapist partially blocks the tube and then blocks it and the nose completely. In order to breathe the patient opens his mouth, emits moans and guttural sounds, then coughs during exhalation. These are favorable factors for the patient's recuperation.

L.I.S. patients have a *dystonie oromandibulaire* which impedes opening their mouths. Exercises in opening and closing the mouth and in contracting the oral-mouth sphere must be performed regularly.

During a second phase, the physiotherapist mobilizes the lips of the patient in order to relax the muscular tension and to promote articulation. Special attention is paid to relaxing the lip muscles, to decontracting the entire oral sphere, and to working on lip musculation. Finally, passive exercises of the praxies are done to work on *mandibulaire* and lip mobility. Throughout the reeducation sessions, the speech therapist helps the patient to place his lips and tongue in the proper position to emit such and such a phoneme. When reeducating the L.I.S. patient, the therapist becomes aware that the patient's emotional state favors verbal expression and permits voluntary attainment of phonemes or words.

Although we have been speaking about the importance of respiratory, swallowing and phonatory exercises, these are not to obtain oral expression again at any price since, for many L.I.S. patients, this is impossible. These are, above all, to make the patient more comfortable. Our principal goal is to liberate the thought process of the patient, which is intact but locked in and too often captured and canalized trying to manage respiratory problems and/or swallow saliva.

In order to have communication, it is necessary for the persons who are speaking to be motivated, to emit and to receive. The consciousness and the capacity of the L.I.S. patient to enter into communication with the world around him may be verified by the opening of his eyes and the orientation of his glance. The first contact to be made with these patients is through a code using the fluttering blinks or the vertical movements of their eyes.

To establish a yes-no eye code, the following eye movements can suffice : "yes" can be indicated by one blink and "no" by two. The principal aim of reeducation is to reestablish a genuine exchange with the L.I.S. patients by putting into place various codes to permit them to reach a higher level of communication and thus achieve an active participation. With sufficient practice, it is possible for them to communicate complex ideas in Morse Code using only eye movements.

FELDMAN has described the case of a patient who used jaw and eyelid movements to communicate in Morse Code.

Usually, to communicate, it is necessary to establish a code based on blinking the eyelids. At present, three alphabetical systems are used :

**1. *Vowel and consonant method***

The alphabet is divided into 4 groups : Vowels, Consonants 1 (B to H), Consonants 2 (J - Q), and Consonants 3 (R to Z). The user says : "Vowel" and then Consonants 1, 2, 3 and the patient blinks his eyelid to indicate the chosen group :

<u>V (Vowel)</u>	<u>Consonants</u>	<u>C1</u>	<u>C2</u>	<u>C3</u>
A		B	J	R
E		C	K	S
I		D	L	T
O		F	M	V
U		G	N	W
Y		H	P	X
			Q	Z

**2. *Alphabetical system using a grid of letters***

		<u>Consonants</u>					<u>Vowels</u>		
		1	2	3	4	5	1	2	
1		B	G	L	Q	V	1	A	O
2		C	H	M	R	W	2	E	U

3	D	J	N	S	X
4	F	K	P	T	Z

3	I	Y
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*Example* : To designate the letter "B" (1-1), the patient blinks his eye once, pauses, and then blinks one time again. If he wishes to designate a vowel, he raises his eyes before blinking. After using this system for a certain length of time, both the patient and the person communicating with the patient know it by heart.

The patient indicates the position of the chosen letter with his eyes ; the user guesses the letter. The resulting dialogue can become remarkably rapid.

### 3. ***Different order of the alphabet***

Listing the alphabet in the order in which the letters are most frequency used in the French language, beginning with those used most often :

E - S - A - R - I - N - T - U - L - O - M - D - P - C - F - B - V - H - G - J - Q - Z - Y - X - K - W .

The user pronounces the letters beginning with the most frequently used, E, and continues until the patient blinks after hearing the desired letter which the user then notes.

It is necessary to begin over again for each letter necessary to form words and phrases. The rapidity of this system depends upon habit and the ability of the patient and the user to work together. The user may be able to guess at a word or a phrase before all the letters have been pronounced. It is sufficient for him to pronounce the word or the rest of the sentence. The patient blinks his eye once to indicate "yes" and twice to indicate "no".

### **The system of universal ocular guidance.**

This is a method for the L.I.S patient to communicate without requiring assistance. It uses eye designation. A camera films, the eye, determines its form and size as well as its reaction when following a specific direction. This eye takes over the role of the mouse. The patient clicks either by blinking the eye or by fixing his glance on a precise point.

A keyboard appears on the screen and thus the patient is able to create a text. This system permits the patient to use computer technology to solve certain difficulties inherent to his handicap. One L.I.S. patient, Mr. V., can print out documents and send faxes. With the help of a vocal synthesis and pre-recorded messages, he can hold a telephone conversation . *Contrôle domotique*, such as operation of HI-FI equipment, electrical apparatus (lighting, blinds, etc..) is also possible. The gentleman always uses a button that he can press with his right hand to turn the TV on and off, to regulate sound and select the stations.

It is also possible to multiply possibilities of self-expression by mimicking or gesturing in the case of a partial motor recuperation. Gesticulating and mimicking play a role modulating verbal interaction between social partners.

These patients may show small simple movements of the head and the eyebrows and short smiles after different kinds of stimulation and sometimes even spontaneously. These tiny motor movements enrich communication.

Shaking the head can mean different things. A single, brief shake could be a sign of attention and mean "yes", while a longer one from side to side could be the sign that there is a problem of understanding. Most often, it would indicate "no".

Certain L.I.S. patients have recovered or kept some motor capacities. Puckering the eyebrow expresses pain or effort.

Smiling differs from other facial expressions, not only in the quality of the emotion expressed but also for other reasons. It is the easiest facial movement that can be made voluntarily, the most used, and the best for beginning and continuing social communication.

Motor functions retained differ from patient to patient. It is essential to use these to complete the eye code communication system which is set up as the first objective by the speech therapist even before more specific reeducation has been begun.

Numerous therapists refuse to consider the possibility of verbal or non-verbal expression for L.I.S. patients because of their limited life span. Those around the patients, family and friends usually encourage the therapists to actively oppose such a pessimistic stand. They request assistance and advice but, in addition, provide the therapist with valuable information about the personality of the L.I.S. patient and his functional improvement.

On the best of cases, the L.I.S. patient can reintegrate into the family environment after many months of hospitalization. This necessitates special arrangements for equipment and daily care (nurse, speech therapist, physiotherapist, help, etc...)

J.D. Bauby, in particular, created an association, ALIS, for the Locked-In-Syndrome. In a letter dated February 13, 1997, he wrote that " ALIS had several objectives : to collect all existing information about the Locked-In-Syndrome, to allow L.I.S. patients, to communicate better, to set up ways to better inform them in order to break down their isolation and to make them full citizens of the 21<sup>st</sup> century.

ALIS has built itself up and organized. An Internet site, ALIS, is accessible : <http://www.alis.fr>.

Progressively more information concerning the pathology of the syndromes, the number and location of patients, lists of rehabilitation centers and specialists, advice on problems of care, reeducation, communication methods, environmental control and computer materials has been added to Internet. At Seminars, it is possible to raise questions and to obtain answers from specialists. "Communication" in all forms is the key-word for the Locked-In-Syndrome.

In this article, the emphasis has been placed on the necessity for swallowing and breathing exercises in order to provide at least minimum comfort to the L.I.S. patient.

Caring for L.I.S. victims is a way to give the intellectual facilities their proper place and consideration.