International Encyclopedia of Rehabilitation

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Locked-in Syndrome

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Locked-in Syndrome (LIS) results from a lesion to the brainstem, most frequently an ischemic pontine lesion. It leads to particularly severe impairments resulting from the complete disruption of the motor pathways controlling eyes, face, trunk and limbs movements, as well as breathing, swallowing and phonation. Consciousness and cortical functions are preserved. Care and rehabilitation of the affected individuals, described as being "locked in", present great challenges. Access to communication is the main goal of treatment. Considering the current life expectancy of persons with locked-in syndrome, the top priority for the rehabilitation team is to help them reach the highest standards of quality of life possible.

Alexandre Dumas first described LIS in 1844 in his novel "The Count of Monte-Cristo". His character, Mr. Noirtier de Villefort, victim of a stroke, survived and learned to communicate by closing and opening his eyelids and by vertical movements of his eyes. In 1966, Plum and Posner described a false coma resulting from supranuclear motor de-efferentation that they called 'locked-in syndrome' (Plum and Posner, 1983).

Definition

In 1995, the American Congress of Rehabilitation Medicine defines LIS as a syndrome characterized by preserved awareness, relatively intact cognitive functions, and by the ability to communicate while being paralysed and voiceless (American Congress of Rehabilitation Medicine, 1995). This syndrome is defined by five criteria:

1. Sustained eyes opening and preserved vertical eye movement
2. Preserved higher cortical functions
3. Aphonia or severe hypophonia
4. Quadriplegia or quadriparesis
5. Primary mode of communication that uses vertical eye movements or blinking

Pathophysiology

LIS occurs as a result of bilateral lesions in the ventral portion of the pons. Damage to the corticospinal tracts (paralysis of all four limbs) induces selective supranuclear motor de-efferentation. The associate impairment of corticobulbar tracts leads to ocular motility disorders or paralysis of the IV and VI cranial nerves. The preservation of the levator of the upper eyelid and the upward gaze indicates that the mesencephalon and the oculomotor nerve (III) are intact. Damage to the nuclei of the facial nerve (VII) and lower cranial nerves explains the presence of facial diplegia, anarthria, dysphagia, tongue paralysis, as well as aphonia and respiratory failure.

Etiology

Ischemic strokes are the most common cause (Laureys et al, 2006; Patterson and Grabois, 1986; Doble et al., 2003). They most commonly occur following a basilar artery thrombosis with secondary occlusion of the perforating arteries. The other causes of LIS are hemorrhages and head trauma (pontine contusion or axonal damage). Correlation between vertebral artery dissection and cervical manipulation raises a number of concerns even though it is not fully supported in the literature (Smith et al., 2008; Ernst, 2007; León-Carrión et al., 2002).

There are less frequent causes that lead to similar clinical picture, such as tumours, central pontine myelinolysis, demyelinating diseases or infectious conditions. Other conditions like severe forms of Guillain-Barre syndrome or amyotrophic lateral sclerosis can present a LIS-like clinical picture without necessarily involving a pontine lesion.

Epidemiology

The actual prevalence rate of LIS is not specifically documented in the literature. Individuals affected in this manner probably represent less than 1% of all strokes, though the incidence rate is probably underestimated. Diagnosis is not always reported as some individuals die during the acute phase, while some others recover so rapidly that diagnosis is not maintained (Smith and Delargy, 2005).

During the acute phase, infections, most commonly pneumonia, are the most common cause of death (40% of the cases). The initial stroke is the primary cause of death in 25% of the cases (Doble et al., 2003; Smith et al., 2008; Casanova et al., 2003).

More than 85% of individuals are still alive after ten years (Doble et al., 2003; Casanova et al., 2003). This is supported by the authors' clinical experience (Beaudoin and De Serres, 2008).

The average age of onset of LIS varies between 17 and 52 years old (Doble et al., 2003; Bruno et al., 2008; Beaudoin and De Serres, 2008; Casanova et al., 2003). The youngest patients have a better prognosis for survival.

Semiology

It may sometimes be difficult to diagnose LIS during the acute phase, especially in cases of LIS caused by a traumatic brain injury with an initial coma. Some patients present complete LIS at first, in which no voluntary eye movement is possible. This clinical picture can thus lead to incorrect diagnoses of prolonged coma, vegetative state, minimally conscious state or akinetic mutism.

Alertness often fluctuates, especially during the acute phase (Gutling et al., 1996). Several authors reported that cognitive functions were generally preserved although cognitive impairment may be present (Smith et al., 2008; Smith and Delargy, 2005; Ruff et al., 1987). Attention and memory disorders are observed in nearly half the cases, especially in individuals with a post-traumatic LIS (León-Carrión et al., 2002; Ruff et al., 1987; Garrard et al., 2002). Emotional lability is a common symptom (Garrard et al.,

2002). Apathy is sometimes observed and may persist in some cases (Beaudoin and De Serres, 2008). Recovery of cognitive functions is often observed in individuals during the first year (Bruno et al., 2008).

Residual vertical eye movement (upward gaze) enables the individual to communicate as early as the first days post-onset. Eye movements will be the basis of communication. Sight is often impaired; the combined presence of impairment to the VI nerve (lateral gaze) with internuclear ophthalmoplegia, nystagmus or ocular bobbing explains diplopia and sometimes persistent blurred vision (Beaudoin and De Serres, 2008).

Initially, the patient is most aphonic and the restrictive respiratory insufficiency requires the use of mechanical ventilation. The clinical picture usually encompasses severe oropharyngeal dysphagia, impaired closure of lips and drooling. Voluntary cough is often impossible, and sometimes there is no reflex cough (Beaudoin and De Serres, 2008). There is an ongoing risk of saliva aspiration. Reduction of vital capacity, saliva aspiration and lack of cough reflex are all factors that can trigger off pulmonary atelectasis and lead to infectious risks (Smith and Delargy, 2005). Aspiration pneumonias are more common during the acute phase, especially in mechanically ventilated patients or if comorbidities are severe.

Sensation is generally preserved, although impairments may be observed when the pontine lesion covers a larger area.

The clinical picture usually encompasses quadriplegia. The majority of classic LIS cases will recover some of their motor skills over time. However, spasticity is a major issue for many patients.

Classification

Bauer described three categories of LIS (Bauer et al., 1979).

- Complete or total LIS: Quadriplegia and anarthria. No eye movement.
- Classic LIS: Preserved vertical eye movement and blinking.
- Incomplete LIS: Recovery of some voluntary movements in addition to eye movements.

According to the authors' clinical experience in rehabilitation, individuals with classic LIS also recover some motor function of the cervical spine, face, as well as some movement in limb extremities, most commonly the thumb or the first digit of the foot.

Individuals with incomplete LIS recover more significantly. Swallowing and speech disorders usually improve over time, although a weak and dysarthric voice remains. These individuals also recover better control of the movements of their head, trunk and partial function of one upper limb.

Acute phase

Locked-in Syndrome is the most severe neurological condition to be admitted in hospital settings. In addition to respiratory tract monitoring and cardiovascular support, the

treatment priority is to select a surgical approach, thrombolysis or the prescription of blood thinners based on the type of vascular impairment documented during the initial assessment (Smith and Delargy, 2005).

The specialized nursing care associated with the early rehabilitation phase decreases mortality risks during the acute phase and maximizes functional status (Casanova et al., 2003). Once the medical condition is stabilized, the healthcare team ensures respiratory support through tracheostomy, oxygen therapy, respiratory physical therapy and mobilizations. They also monitor proper feeding and hydration of individuals through a gastrostomy tube and ensure prevention of venous thromboses, pressure sores or corneal ulceration.

Interventions at the level of communication begin during the early acute phase. In nearly half of the cases, the individual's family is the first to notice attempts to communicate with eye movements (León-Carrión et al., 2002).

Preservation of joint range of motion, which may require bracing, and proper positioning in bed are particularly important at this stage.

Rehabilitation Phase

Individuals with LIS have the highest level of disability among stroke survivors. Early recognition of LIS state is important for rehabilitation. The active participation of patients to the decision-making process regarding their rehabilitation is essential.

Communication

The establishment of a reliable code to indicate yes and no, usually constitutes the basis of communication. Several individuals initially use eye movements and blinking to communicate. As language skills are usually preserved, persons with LIS make efficient use of spelling via an alphabetical code in which their communication partner presents one letter at a time. The person with LIS would then indicate the desired letter by moving their eyes or blinking. The letters may be grouped in several ways as alphabetical order or frequency of use of the different letters. Basic messages can also be added (De Serres and Martel, 2008; Beukelman et al., 2007).

Early on, the rehabilitation team should ensure that individuals with LIS are equipped to draw attention to themselves in order to indicate a potential need or a desire to communicate. The recovery of a reliable movement, will enable the activation of a switch, that could be use to access communication devices (as computer with synthetic voice) and thus allow alternative methods to verbal communication and access to better control of the environment (Beaudoin and De Serres, 2008; De Serres and Martel, 2008).

According to the authors' experience, persons with LIS whose recovery is poor tend to choose to communicate via visual code when interacting with familiar people since it is the fastest method. Computers or communication devices are mostly used with unfamiliar partners of communication, as well as written communication and electronic mail (Beaudoin and De Serres, 2008). Persons with incomplete LIS are usually able to express themselves through multimodal communication methods, which may include visual scan

features, residual speech skills (phonation and articulation), the use of communication devices, computers, and in some cases, handwriting (De Serres and Martel, 2008).

The majority of persons with LIS living at home use a personal computer to engage in conversations, prepare messages, play, write or use Internet. This versatile tool helps these persons to resume meaningful leisure activities as well as social and family roles (Bruno et al., 2008; Beaudoin and De Serres, 2008).

Swallowing and breathing

Initially, patients are essentially fed through a feeding tube as 'oro-glosso-laryngo-pharyngeal' paralysis may significantly worsen the risk of aspiration. Following the initial clinical and radiological assessment of their swallowing ability (videofluoroscopy), most patients gradually start with eating smooth purees and thickened liquids.

Making a decision about oral feeding is not only based on the imaging results, but also on the presence or not of risk factors for aspiration pneumonia. Tracheostomy, mechanical ventilation, cardiac failure, chronic obstructive pulmonary disease, uncontrolled emotional lability, improper positioning during feeding or prior conditions of undernutrition or dehydration are all factors that may lead the healthcare team to remain cautious and delay, at least for a little while, feeding through the mouth.

Some individuals may be suitable for weaning from their tracheostomy as their condition improves during the first months.

In some cases, the use of anticholinergic drugs, scopolamine or botulinum toxin injections at the level of the parotid glands will enable better saliva control in dysphagic patients.

According to the literature and based on the authors' experience, optimal oral hygiene care, delivered before and after meals, combined with compensatory strategies, is known to minimize the risk of pneumonia and enable an early attempt to provide small amounts of clear water to patients under supervision. The desire and determination of people to feed orally leads the rehabilitation team to consider earlier feeding attempts. Many of them are successful, which goes beyond the healthcare team fears and predictable physiological functions.

Positioning and mobility

Progressive verticalization as well as positioning in bed and wheelchair ensure comfort, visual contact and promote the functional use of the emerging voluntary movements (De Serres and Martel, 2008). Exercises to maintain range of motion, as well as breathing, eyes, head, trunk and limb control exercises are performed throughout the rehabilitation process.

Stretching exercises, use of antispasmodic drugs or botulinum toxin injections in a context of spastic dystonia help to decrease spasticity and facilitate caregivers' work, especially regarding personal care, positioning and transfers.

As sensation is often preserved, healthcare personnel should pay very special attention to ensure the comfort of patients when in bed or in wheelchair. A judicious selection of cushions, mattresses, wheelchairs and other technical aids is required. The presence of sensation contributes to decreasing the risk of developing pressure sores in this population with extremely reduced mobility.

In some cases, recovery of upper limb motricity in persons with incomplete LIS, evolving from distal to proximal, enables them to participate in activities of daily living (ADL) and feeding process.

Loss of postural reflexes and balance disorders, associated with the importance of paresis in lower limbs, does not usually enable independent gait, except in rare cases. However, some individuals with incomplete LIS are successful in performing weightbearing during pivot transfers (Beaudoin and De Serres, 2008).

Following the increased tolerance for sitting and powered wheelchair skills training, most patients can move around independently at the end of the rehabilitation process in a powered wheelchair with head control (classic LIS) or manual joystick control (incomplete LIS) (Beaudoin and De Serres, 2008).

Sphincter control

Sphincter retraining enables continence for most individuals. However, severity of paresis and transfer dependence often leads the healthcare team to suggest the use of indwelling catheters, which allows more freedom for mobility and leisure activities.

Cognitive functions

Apathy, often present at the beginning of the rehabilitation process, is a barrier to both the patient and healthcare team. When it hinders optimal rehabilitation outcomes, introducing psychostimulant drugs is appropriate. Serotonin reuptake inhibitors are the preferred choice for the individual with significant lability.

Recovery and prognosis

Recovery of horizontal eye movements prior to the first four weeks is associated with a good neurological prognosis (Bauer et al., 1979).

According to the authors, recovery of the oral-motor area (speech and swallowing) begins later and spreads over a longer period than seen among other stroke patients. The same is true for recovery of movement in distal parts of limbs of individuals with classic LIS (Janjua et al., 2005; De Serres and Martel, 2008).

The great majority of patients are weaned from their tracheostomy during the first months. One third will be able to utter isolated comprehensible words after the first year (Casanova et al., 2003).

During inpatient rehabilitation, more than 50% of individuals start feeding orally and their gastrostomy is removed during the first year (Bruno et al., 2008; Beaudoin and De Serres, 2008; Casanova et al., 2003).

Level of independence in activities of daily living is directly related to motor recovery. If the majority of these individuals can move around independently, most often in powered wheelchair, personal care is another matter entirely. Very few of them reach a level of motor recovery enabling them to feed and perform ADLs on their own (Beaudoin and De Serres, 2008). Spasticity often remains a significant problem for many individuals, even after several years. The authors' experience shows that individuals with LIS usually do not suffer from a pain syndrome during the chronic phase.

In several countries, most of these patients go back to live at home (Doble et al., 2003; Casanova et al., 2003). The burden is then on the family and financial aid is rarely sufficient to compensate the numerous needs and environmental modifications required to live at home. Despite the degree of their impairments, these persons demonstrate extraordinary creativity in resuming their parental and social roles (Beaudoin and De Serres, 2008).

Quality of life

Doble (2003) published the most comprehensive study on the long-term survival rate in persons with LIS who were included in an initial cohort 11 years earlier: 54% never considered euthanasia, 46% considered it and none of them chose not to be resuscitated. According to the authors' experience, even though some patients consider the idea of dying at the beginning of their rehabilitation process, most of them affirm their desire to live after they have overcome the initial emotional shock. In the light of these facts and of what is found in the literature, thinking that these patients should have chosen death is no longer valid.

Access to communication via increasingly sophisticated technological tools associated with family support enhance the quality of life of these persons and enables them to remain active in society (Dupont et al., 1992). As mentioned by Smith and Delargy, few of them return to the labor market (Smith and Delargy, 2005; Beaudoin and De Serres, 2008). However, their level of social participation and leadership is sometimes remarkable, as demonstrated by some of their achievements: some offer conferences, create mutual aid funds or write books (see list in appendix).

The great majority of individuals with LIS have a strong desire to live and reach a level of quality of life way beyond what most people would foresee based on the severity of their impairments (Doble et al., 2003; Bruno et al., 2008; Dupont et al., 1992; Beaudoin et al., 2010). These persons are a remarkable model of inspiration for healthcare teams and their loved ones.

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Appendix: Books written by persons living with LIS consequences

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