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Narrative Review

Locked-In Syndrome: Practical Rehabilitation Management

Ellen Farr, MD, Kathryn Altonji, MD, Richard L. Harvey, MD

Abstract

Locked-in syndrome is a rare and devastating condition that results in tetraplegia, lower cranial nerve paralysis, and anarthria with preserved cognition, vertical gaze, and upper eyelid movements. Although acute management is much like that of any severe stroke, rehabilitation and recovery of these patients have not been previously described. Challenges relevant to this population include blood pressure management and orthostasis, timing and appropriateness of reinstating oral feeding, ventilatory support, decannulation after tracheostomy, bowel and bladder management, vestibular dysfunction, and eye care. Targeted rehabilitation of head, neck, and trunk stability to improve function, and proper fit in an appropriate wheelchair are essential to assist with mobility. Rehabilitation interventions should include a focus on distal motor control and upright tolerance training followed by balance and mobility exercises. In addition, special considerations must be given to developing early methods of communication through use of augmentative systems to call for help and express needs. These systems along with additional technology provide the basis to promote connectivity to family and friends through the use of social media and the internet. Establishment of communication, mobility, and connectivity is essential in promoting independence, autonomy, and improving quality of life. Overall, with specialized rehabilitative care and access to the proper equipment, long-term outcomes and quality of life in these patients can be favorable.

Introduction

First defined in 1966 by neurologists Plum and Posner, locked-in syndrome (LIS) was identified as a combination of tetraplegia, lower cranial nerve paralysis, and anarthria (speechlessness due to severe dysarthria) with preserved awareness, vertical gaze, and upper eyelid movements.[1] An early literary description of this condition is found in the 1844 novel *The Count of Monte Cristo* where Alexandre Dumas described a character as a "corpse with living eyes" who could not move but communicated to family with blinking and vertical eye movements.[2]

The prevalence of LIS is not well documented. A study in Dutch nursing homes estimated a prevalence of 0.17/10,000 whereas other sources estimate <1/1,000,000.[3,4] LIS cannot be well studied by systematic review because most of the literature consists of independent case reports. The goal of this paper is to review the etiology, clinical manifestations, and comprehensive rehabilitation of LIS based on 20 years of clinical experience through an organized program at a large academic center.

Anatomy

Classic LIS results from injury to the bilateral basis pontis containing the corticospinal and corticobulbar tracts, sparing the lateral and dorsal aspects (Figure 1). Damage to this area is responsible for complete motor tetraplegia and paralysis of the bilateral face. The fascicles for the abducens nerve and the paramedian pontine reticular formation are also injured resulting in impaired bilateral horizontal gaze. Sensation is usually preserved because the medial lemniscus and the spinothalamic pathways (which sit laterally) are spared. Vertical eye movement and eyelid control are also spared as they are primarily controlled by cranial nerve III (oculomotor), cranial nerve IV (trochlear), the rostral midbrain reticular formation, and pretectal areas of the midbrain. Finally, arousal and consciousness remain intact as they are modulated by the ascending reticular activating system, which is largely located in the midbrain and cerebral peduncles. Thus, the clinical presentation of LIS includes tetraplegia, oral motor and facial paralysis, and impaired horizontal eye movement with intact vertical eye

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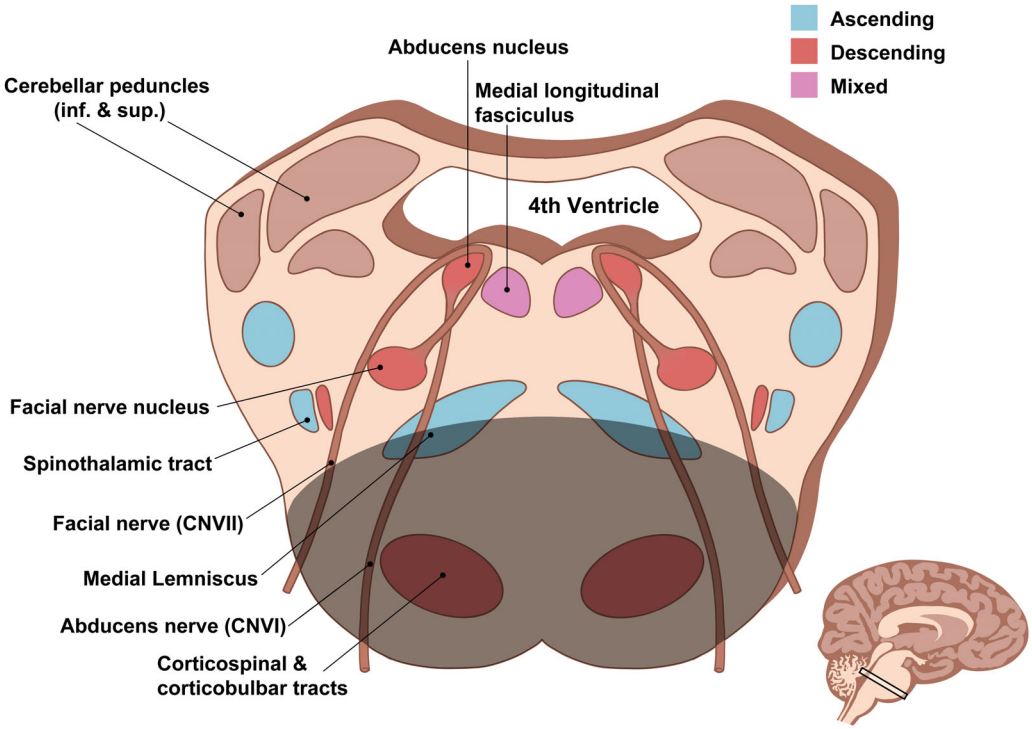

Figure 1. Axial view through the midpons highlighting relevant pathways. Shaded area represents area of infarct in locked-in syndrome patients.

movement and full sensation.[5] This distinguishes LIS from other similar appearing conditions such as disorders of consciousness (DOC) including coma, unresponsive wakefulness syndrome, and minimally conscious syndrome.

Etiology

Traditionally, LIS is caused by an ischemic infarction or hemorrhage affecting the base of the pons, but it can also stem from traumatic brain injury, tumor, central pontine myelinolysis, brainstem abscess or encephalitis, air embolism, toxins, or heroin abuse.[6]

LIS Classification

The classification of LIS was published in 1979 by Bauer and others as a series of cases where each category of LIS was defined by the clinical presentation (Table 1). Each classification was further subdivided into transient or chronic.[7,8]

Acute Care Management

Because LIS can be mistaken for DOC, care should be taken during initial examination to evaluate eye function and attempt communication through instructed eye movements. Nearly all patients presenting with LIS require urgent respiratory care including intubation and ventilator support, with management in a neurointensive care unit (NICU).

Tracheostomy will be necessary if prolonged intubation is required, and because patients with LIS typically have severe oral motor paralysis and dysphagia, placement of a long-term percutaneous feeding tube should be an early consideration. As with all strokes, extremes of blood pressure (BP) should be avoided with a target systolic BP <220. Interventions such as tissue plasminogen activator, thrombectomy, and ventriculoperitoneal shunt are other acute considerations; however, these are beyond the scope of this review and will not be discussed further.[9,10]

Rehabilitation Management

The rehabilitation management discussed here is unique to LIS patients. Although certain aspects are

Table 1
Classifications of locked-in syndrome

Classification	Description
Classical LIS	The 1966 definition of quadriplegia and anarthria with preserved awareness and vertical eye movements.
Incomplete LIS	Refers to any case in which there are remnants of voluntary movement, or eye movements beyond conjugate up-gaze. Improvements in classical LIS would lead to the incomplete form.
Total LIS	Indicates a patient with full immobility including eye movements but with retained consciousness. This would require further testing to differentiate from coma or unresponsive wakefulness syndrome.

LIS = locked-in syndrome.

shared with spinal cord injury or traumatic brain injury care, the particular pattern of injury in these patients manifests as distinct deficits not routinely cited in the literature, specifically treatment of eye care, bulbar weakness, communication impairments, and ventilatory dysfunction.

Rehabilitation begins in the NICU with proper positioning, establishment of functional communication using eye movements, and mobilization as soon as medical stability allows. Initial rehabilitation requires a thorough examination followed by targeted rehabilitation interventions as clinically indicated.

Rehabilitation Oriented Examination

As with most strokes, the physiatrist will know the location and usually the cause of the stroke before performing a physical and neurological examination. The goal of the physiatric examination is to identify ongoing medical concerns and neurological impairment, which in turn indicate realistic activity goals for the patient. Any voluntary movement noted can be used functionally, either for mobility, self-care, communication, or operation of an adaptive device.

Before examination, it is critical that the examiner introduce him- or herself within the patient's field of view and explain the purpose of the examination. Because patients with LIS are typically immobile with intact sensation and limited field of view, any contact with another person can be misunderstood or even interpreted as threatening. Thus, orienting the individual to who is present and the goals of the encounter will put the patient at ease for the events to follow.

A "top-down" approach to the examination is best (Table 2). Important points include eye movements, lid closure, facial control, and ventilatory control. With incomplete LIS, eye movements beyond conjugate upgaze should be noted. The lack of ability to achieve full eyelid closure on either side is an important finding to note on facial examination, as is the ability to move lower face and lips. Facial sensation is usually intact but partial loss is not unusual. Ventilation is typically diaphragmatic only and often consists of involuntary tidal breathing exclusively. Asking the patient to take a deep voluntary sigh and hold the breath after exhalation determines whether some control of ventilation remains. If only involuntary breathing is noted, reflexive coughing and yawning may still occur. The strength of a voluntary or involuntary cough as well as spontaneous swallowing can help reduce tracheal suctioning needs.

Table 2
"Top-down" physician exam approach

"Top-Down" Examination	
Head & Neck	**Positioning:** Upon introduction, note tonic neck rotation to either side. **Movement:** Examine of voluntary head and neck movement including cervical spine range of motion. **Tracheostomy:** If present, note the type and size, and examine for any signs of infection or adjacent skin breakdown.
Eyes	**Range of Motion**: Determine the maximum range of eye movements and if dysconjugate gaze or nystagmus are present. **Lid Closure**: Test ability to fully close the eyes.
Ears	Perform a hearing exam on both sides. Hearing is intact in most cases, but depending on lesion location, unilateral hearing loss may occur.
Face & Throat	Examine for movement and sensation in the face and lips as well as for any tongue and jaw movement, including voluntary swallow.
Cardiac	Perform a standard cardiac exam.
Respiratory	Perform careful auscultation of lung sounds. **Ventilatory Control**: Test for ventilatory control and cough strength. Ask the patient to take a deep voluntary sigh and hold the breath after exhalation. This indicates if a patient has some control of ventilation. **Cough**: If a spontaneous cough occurs, note if there is sufficient strength to clear secretions.
Abdominal	**Feeding Tube**: If present, note the size, and the type. Record in the medical record if the tube is "traction removable" (as tube labeling may rub off during hospital stay). **Skin**: Check the skin around the tube for signs of erythema, discharge, or breakdown.
Neurological	**Strength**: Perform a standard neurologic examination of strength. Grading is performed using the Medical Research Council system (0-5). Note abnormal flexor or extensor synergy or posturing. **Sensation**: May be intact throughout, however pin prick should be tested in all four limbs and each side of the trunk.
Skin	A thorough skin examination is critical as pressure ulcers are common.

Blood Pressure and Heart Rate Management

Upon presentation to acute inpatient rehabilitation, patients with LIS typically remain on one or more antihypertensive medications as optimal BP control is a critical part of secondary prevention. However, when transitioning a patient with LIS to wheelchair or tilt table early in the rehabilitation course, orthostatic hypotension is common and is more concerning medically than an elevated BP while in bed. Hypotension is partly because of profound weakness from stroke, prolonged bedrest, modification of vagal nerve function due to brainstem injury, and use of multiple antihypertensive agents. Resting pressure often remains within the normal range after reducing antihypertensives to address orthostatic hypotension. If in the process of weaning medications, one finds that supine systolic pressures are persistently 160 and above and orthostatic hypotension continues, then pressure stockings or an abdominal binder may be necessary, but only during therapy. With time and increased mobilization, pressure garments can be discontinued, and BP will remain stable on adjusted antihypertensive doses. The therapeutic approach to orthostatic hypotension includes progressive elevation on a tilt table with close monitoring of BP and heart rate, maintaining them in the appropriate physiological range.

Tachycardia is also common after LIS because of vagal dysfunction but will usually normalize over time.[11] In the meantime, use of beta-blockers to maintain a normal heart rate is beneficial and usually does not have a negative impact on BP.

Paroxysmal sympathetic hyperactivity (PSH) is rare but can occur in patients with LIS. Episodic PSH is managed using standard approaches.[12] If despite best efforts frequent episodes of PSH continue, an intrathecal baclofen pump may be the best management option.[13,14]

Ventilation and Respiratory Function

Protecting and maintaining a clear airway is the primary strategy for respiratory management in LIS. As in all patients with dysphagia, upright position during enteral or oral feeding is mandatory, and close attention to upper respiratory congestion and secretion management is critical. Because LIS patients have limited communication and ability to use hospital call lights, frequent (q30 minute) nursing checks are recommended. The presence of a family member or caregiver helps with close monitoring, even if just to notify staff when the patient needs suctioning.

Fortunately, most patients who survive with LIS will not require chronic ventilator care. Respiratory failure is a significant cause of early mortality within the first 30 days following bilateral brainstem stroke, but among those who survive, chronic ventilator use is uncommon, though a need for chronic tracheostomy has been reported at 36%.[6,15] All patients with complete LIS and many with incomplete LIS have a ventilation pattern that is limited to involuntary tidal volumes.[16,17]

The emergence of voluntary ventilatory drive predicts a favorable prognosis for tracheostomy weaning and the return of swallow function. In contrast, involuntary ventilation places the patient at risk for pulmonary atelectasis and therefore hypoxia. An aspiration event or pulmonary embolism in the context of bilateral atelectasis can be devastating, likely leading to rapid respiratory decline or respiratory arrest.

The prevention and treatment of atelectasis are best achieved using a mechanical insufflator-exsufflator, also known as a cough assist (CA).[18] This modality can be given via tracheostomy or face mask if air leaking can be prevented. The CA works by delivering high pressure air to inflate the lungs followed by a rapid negative pressure to facilitate lung deflation, acting like a mechanical cough or high velocity sigh. The lung inflation can open collapsed airways, and the deflation can mobilize pooled secretions upward. The latter can help secretion clearance from airways when combined with deep suctioning. Providing CA, with or without beta-agonist nebulizer treatment, can help minimize alveolar collapse and maintain optimal pulmonary toilet when given at a minimum of every 8 hours. It is unlikely that the risk of atelectasis can be fully mitigated until the patient recovers voluntary ventilation, an effective reflexive cough, and spontaneous swallowing of secretions. Once these clinical improvements are observed, a trial without CA can be considered. Successful weaning from CA usually occurs in tandem with tracheostomy weaning.

Classic pulmonary physical therapy with postural drainage can be incorporated as well. Postural drainage has limitations because many patients with LIS find certain positions uncomfortable and may feel vulnerable in some prone and head down postures. The role of diaphragmatic pacing in LIS is unknown as it has not been studied in this population, but given that most survivors do not require ventilator support, its role would likely be limited.[19]

Tracheostomy Management

The majority of patients with LIS will be admitted to acute rehabilitation with a tracheostomy tube. The optimal tube for airway management in LIS is a cuffed tight-to-shaft (TTS) tube. The TTS is preferred because it allows for effective use of the CA and pulmonary toilet when the cuff is inflated, and for optimal tracheal airflow during respiratory and voice training when the cuff is deflated and the tracheostomy tube is occluded.

Many patients with LIS can eventually be decannulated, with the decision to do so individualized to the patient's clinical and neurological status. Prognosis for successful decannulation is better for younger patients with LIS but more guarded for older patients and those with chronic lung disease (eg, chronic obstructive pulmonary disease). At a minimum, to be considered a candidate for decannulation, patients with LIS must recover spontaneous or voluntary swallowing of oral secretions and have an effective spontaneous cough. One process for successful tracheostomy weaning is shown in Figure 2.[20]

Swallowing and Nutrition

Patients with a bilateral brainstem stroke will present with prolonged dysphagia requiring a percutaneous feeding tube. Enteral feeding should be advanced very slowly from continuous to bolus feeds via a gastric port because nausea and vomiting, especially with movement, are common owing to vestibular dysfunction. In addition, there is a high risk of regurgitation and aspiration with reflexive cough, especially in patients with severe dysphagia.

Recovery of swallowing is possible in some patients. Coordinated swallowing of food and liquid requires adequate oral motor control for bolus formation and propulsion with a coordinated swallow trigger. Once spontaneous swallows are noted, introduction of small volumes of liquid and pureed foods by a speech language pathologist is reasonable. Advancement of feeding trials is based on bedside assessment supplemented by

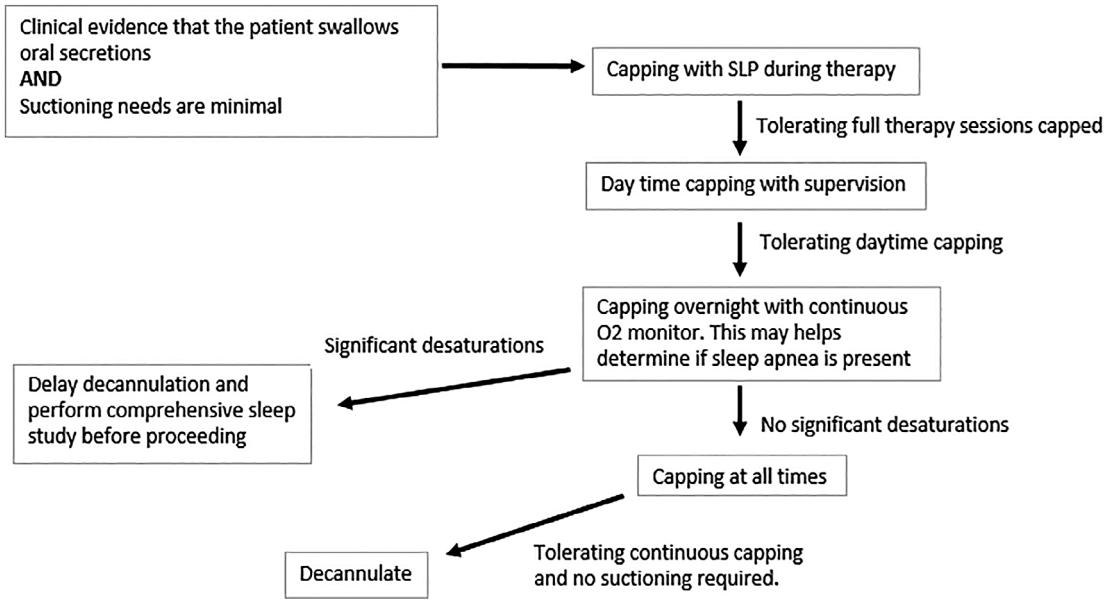

Figure 2. Recommended treatment algorithm for tracheostomy weaning in locked-in syndrome patients.[20] SLP = speech language pathologist.

diagnostic evaluations with video fluoroscopy or fiberoptic endoscopic evaluation of swallowing. Oral diet can be advanced in some cases to thin liquids, but caloric and fluid intake may remain limited and supplemental enteral feeding and fluid flushes may be required. Recovery of swallowing endurance can take time, but many LIS survivors eventually do have the feeding tube removed weeks to months after placement and usually after discharge from rehabilitation.

Eye Care and Vision

Severe facial nerve palsy following pontine stroke can limit voluntary eye closure on one or both sides and exposure keratopathy is common leading to keratitis, infection, and blindness if left unattended.[21] All patients with LIS should be offered saline eye drops as needed because they are unable to wipe debris from their own eyes. If incomplete eyelid closure and widening of the palpebral fissure are observed, then eye drops should be scheduled every 4 hours while awake, and ophthalmic ointment applied every night with the eye closed and covered by a sterile gauze eye pad. The eyes should be examined daily for signs of redness, corneal abrasions, and infection. Once effective palpebral closure is noted both voluntarily and during sleep, eye care strategies can be withdrawn.

Vestibular Dysfunction

In classic LIS, patients will have retained conjugate upgaze and lack all other eye movements. For patients with incomplete LIS, partial or complete horizontal movements may be noted, though often dysconjugate, leading to diplopia with certain directional gazes that may respond partially to corrective prism lenses. Diplopia combined with movement can lead to motion sickness. Furthermore, some LIS patients may have injury to vestibular nuclei, the cerebellar peduncles, or cerebellum resulting in true central vestibular dysfunction. Together these can lead to problems with recurrent nausea and vomiting that can interfere with therapy, increase the risk of aspiration pneumonia, and compromise nutrition. The primary treatment is to allow for accommodation overtime, and acute rehabilitation may have to be delayed in those with severe vertigo. In mild to moderate cases, vestibular dysfunction can be addressed as part of the acute rehabilitation program with some combination of habituation training, functional mobility, and balance interventions as well as vestibular adaptation in the form of gaze stabilization exercises and substitution.[22] Associated nausea can be managed with antiemetic drugs (eg, meclizine, metoclopramide, etc) with careful monitoring for confusion and other cognitive side effects. Emesis should be avoided as it markedly increases the risk for chemical pneumonitis or pneumonia in these patients. If frequent emesis persists despite pharmacotherapy, placement of a gastrojejunal tube with slow feeding through the j-port is often the best solution.

Bladder and Bowel Management

People with bladder and bowel dysfunction are characteristically incontinent with uninhibited bladder but otherwise have normal micturition. Urinary retention early after stroke is common but will resolve spontaneously during recovery.[23] If urinary retention is ongoing, short periods of bladder reset with placement of an indwelling

catheter may facilitate resolution. Use of tamsulosin may be beneficial for males with a history of benign prostatic hypertrophy keeping in mind that this drug can contribute to orthostatic hypotension. A trial of bethanechol is an additional option, but its efficacy in stroke-related urinary retention is unclear and may cause abnormal cramping, diarrhea, and an increase in airway secretions.

Patients with LIS are also at risk of constipation. Maintaining good hydration with judicious use of laxatives is appropriate.

Spasticity and Splinting

Severe spasticity and decorticate posturing are uncommon in LIS. Contractures are generally preventable by proper positioning in the bed and wheelchair. However, when hypertonia emerges, splinting may be beneficial. The effectiveness of splinting to prevent contracture and control spasticity is not well established[24] but is probably worthwhile in the context of a comprehensive rehabilitation program that includes stretching and therapeutic exercise. If a patient has resistance to passive ankle dorsiflexion either from spasticity or soft-tissue stiffness, the use of static-progressive ankle-foot orthoses (AFO) on both lower limbs while in bed is recommended. Standard solid ankle AFOs are beneficial for providing ankle stability during therapy, especially in a stander or on the treadmill. Resting hand splints should be applied when wrist and finger flexor spasticity is noted.

Spasticity is managed according to usual standards combining enteral agents for general hypertonia and botulinum toxin for focal spasticity. Sedation and weakness are risks, but when necessary these two methods along with regular mobilization, stretching, and other therapeutic exercise are usually sufficient to control spasticity and facilitate motor recovery. In some cases, an intrathecal baclofen pump may be necessary.[25]

Pain Management

Most pain reported by patients with LIS is in response to body position and points of pressure in the bed and wheelchair. Frequent repositioning throughout the day and night are necessary for comfort and skin protection. Focal areas of neuropathic pain are rare in classic LIS with injury limited to the base of pons, but if the lesion involves the adjacent lateral spinothalamic tracts, central poststroke pain may occur and will need to be treated pharmacologically.[26]

Pathological Laughing and Crying

Within the pontomesencephalic region of the brainstem is a network of nuclei associated with the generation of movements involved in laughing and crying that are typically inhibited by descending fibers from the amygdala and premotor cortex. Pontocerebellar inputs are additionally involved in adapting the execution of laughing or crying to the cognitive and situational context. Following a bilateral brainstem infarction, the pontomesencephalic network can be disrupted or disconnected from descending or cerebellar inputs. The result is pathological laughing and crying that are typically appropriate for the patient's mood but out of proportion to the reported level of emotion.[27] We choose this term based on support from the literature regarding the characteristics of this disorder and because LIS is a true bulbar injury, making use of the more common term *pseudobulbar affect* rather unsuitable. A patient may laugh excessively at only mildly funny jokes or cry uncontrollably with minor setbacks in progress, and notably full facial expression of the emotion is observed even when voluntarily movement is lacking. When these symptoms become disturbing to the patient or family, serotonin reuptake inhibitors are the first and best choice for management, starting with a low dose and titrating until emotional expression is less extreme. The use of dextromethorphan-quinine is a secondary choice for management.[28]

Communication

The importance of establishing an optimal means of communication has been previously emphasized. Within the rehabilitation hospital, communication includes both a system to call for assistance as well as techniques to express wants and needs. For the former, the presence of a family member or caregiver around the clock can be most effective to ensure the patient's needs are communicated to staff in a timely manner. However, it is beneficial to determine if a low-pressure touch pad can be used to call staff either with a head turn or finger press. If communication is limited to eye movements only, regular nursing checks at least every 30 minutes are necessary.

A simple method to express yes and no is mandatory for communication of wants and needs. It is usually best to continue use of any system used prior to rehabilitation if available. If no consistent system had been established, using "*eyes up for yes and eyes closed for no*" can be easily taught to patient and family. A picture board can be used for a quick needs assessment and an AEIOU board can be used for more complex communication (Figure 3A). In both cases the examiner must point to each item sequentially along a column or row until the patient indicates the correct letter or icon (eg, with an upward gaze). Patients with at least some horizontal and vertical eye movements can use the E-TRAN board, which requires first a gaze to one of six color blocks to establish the group to which a letter belongs, followed by a gaze to a color block again to indicate which letter in the group (Figure 3B). The E-TRAN board is often more efficient to use if complex communication is necessary, but effective use requires good attention, working

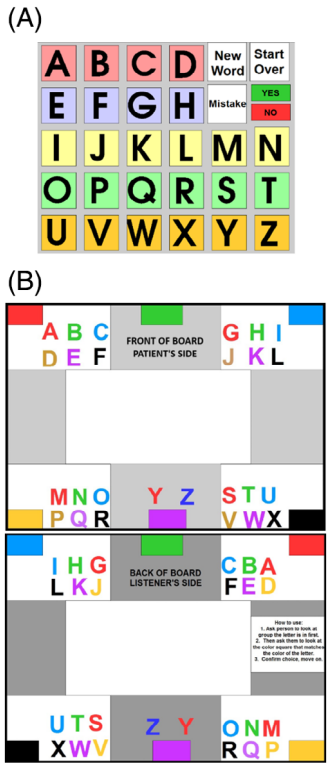

Figure 3. (A) Example of an AEIOU communication board. (B) TOP: E-TRAN board patient side. BOTTOM: E-TRAN board caregiver side.

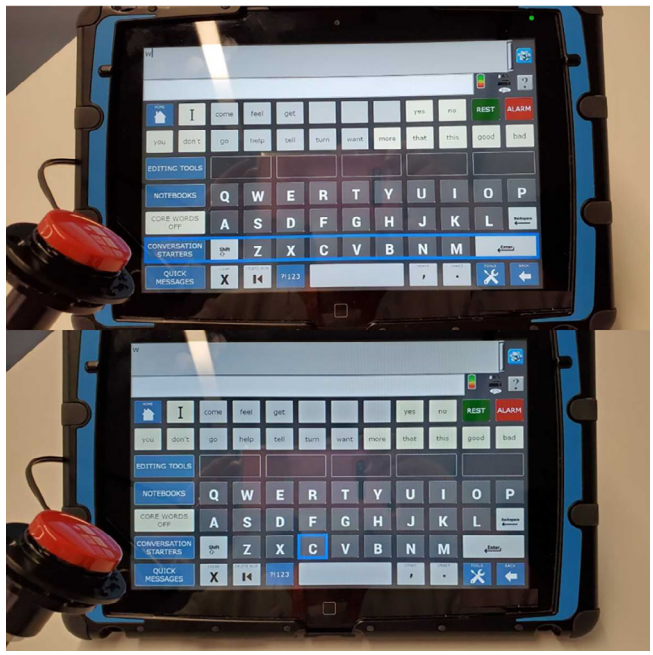

Figure 4. Switch Scanning example. TOP: Switch scanning screen with row selection highlighted. BOTTOM: Switch selection with final selection highlighted.

memory, and the ability to hold gaze on the target until the communication partner understands the selection. Eventually patients and caregivers become quite efficient with these simple letter systems. Many patients will ultimately use a purely verbal AEIOU system where the communication partner verbalizes the memorized letter sequence until the patient indicates the correct letter has been spoken.

Computer based augmentative and alternative communication systems (AAC) are available that use eye tracking or switch scanning (Figure 4). Eye tracking systems can be used to spell words and work best for those with both conjugate horizontal and vertical eye movements. Caregivers require sophisticated training to learn to troubleshoot eye tracking communication systems because proper patient and device positioning are necessary for success and recalibration is often needed. Switch scanning uses voluntary movement (head or digit) to sequentially scroll through pictures or letters. Switch scanning is more reliable than eye tracking but less efficient. Devices that combine the use of eye tracking and switch scanning may work well for some patients. Though these systems allow for expression in a computer-generated voice, many patients choose a simple letter board over high-tech systems. Ultimately the determination of the best technology must be based on the individual's personal needs, ability, available support, and functional goals.

The newest innovation in communication is the brain-computer interface (BCI) using noninvasive or implanted electrodes to collect neural signals that drive computer-based AAC. This technology is presently in the investigative phase but may lead to clinically useful solutions in the future.[29] The most common BCI technology uses the P300 event-related potential, which is a neural response that occurs during active visual engagement within a field of visual targets (eg, gazing at a letter among a choice of letters). The peak latency following this visual discrimination is 300 ms. The P300 event-related potential can be used to spell words and sentences for a computer-generated voice.[30]

Head Control and Cervical Strength

A detailed discussion of the rehabilitation of head control and cervical strength in the neurological patient is beyond the scope of this paper; however, in incomplete LIS it is common to see some voluntary cervical muscle activity. A key clinical focus then is to improve cervical strength and head control with a goal to achieve independent control of head position during daily activities. Therapy will initially focus on head turning and lifting from a flat surface while supine, but then progress to head control in gradually more upright positions. Independent head control reduces the need for assistance during bed to chair transfers. For example, lacking head control during transfers requires an additional caregiver to provide head and neck support. Head control is critical for upright sitting in a wheelchair, operating a wheelchair using head movement, using higher tech AAC, and scanning the environment when driving a power wheelchair (PWC).

Figure 5. Example of a locomotor training system with pedals and functional electrical stimulation.

Mobility

In addition to the critical role of cervical strength and head control in advancing mobility, tolerance of upright position must be achieved before PWC training. Tilt table training strengthens cervical muscles, enhances trunk stability, and improves or maintains ankle flexibility. Once a patient can tolerate upright position, mobility training may include use of a standing frame, locomotor training with or without functional electrical stimulation (Figure 5), gait training on a treadmill with body weight support,[31] or over ground walking, depending on the amount of motor recovery and balance observed.

The majority of LIS patients will require the long-term use of a wheelchair. For those who achieve only minimal recovery, a tilt-in-space wheelchair is the ideal choice for providing upright position, easy pressure relief, and transportation. If adequate head control is achieved, PWC mobility may be appropriate using a head array to control wheelchair direction and speed. Finally, if the patient achieves arm and hand recovery sufficient to control a switch or joystick, a PWC can be prescribed with these control options. Wheelchair training may require more time than usual because of limited range and coordination of head or hand for operating a wheelchair controller. Sip and puff systems used in cervical spinal cord injury typically work poorly in classic LIS because these patients lack oral motor strength and good respiratory control. Eye gaze can work for communication, but in the context of moving through space in a power chair it becomes more challenging when eye movement is limited and complicated by diplopia.

Other Equipment

A profound concern for a person with LIS is the risk of social isolation and the constant dependence on others. Fortunately, today we all have the capacity to stay connected to the world and others via the internet and social media. In addition, computer access vastly contributes to a person's educational and vocational goals.[32] The challenge is to provide functional computer access using available assistive technology (AT), which unfortunately has been traditionally underused, mismatched for the individual's needs, and underfunded.[33] Environmental control systems are another technology that helps to reduce dependence on others, giving persons with LIS the ability to turn on and off lights or a computer, to choose what to watch on television, or to call for assistance.

The approach to prescribing AT, like with communication, must be client centered focusing on the individual's strengths and abilities. A therapist with knowledge of the technology market and expertise in matching AT to individuals is critical. Multiple options may be appropriate for any one individual depending on the task goal and available motor function (Table 3). A simple environmental control system is a switch that can be controlled by a head or limb for turning on or off appliances, changing channels on a television, or selecting icons on a computer screen. Once a switch is selected, it will need to be mounted for access usually on a wheelchair or in a workspace. More specific switch options may be available depending on the patient's residual motor control. Such systems include electronic head pointing, electronic gaze systems, or modified upper extremity systems (Table 4).

Whether using a switch, head pointing, or eye gaze system, writing emails or posting on social media can be laborious because of the slow rate of character typing. Software is readily available to enhance typing efficiency such as expanding abbreviations into full words or sentences (eg, hru = How are you?). Word prediction is also helpful, where the software anticipates the word you are typing (commonly used with texting on smartphones). BCI systems such as the P300 event-related potential may be a faster method of typing for those who master the technology but is less readily available, costly, and demands a high level of attention from the user. Still, BCI has the potential to be a powerful tool for written communication and internet use in the future.[30]

Table 3
Types of switches available for assistive technology systems

Types of Switches
Mechanical "push button"
Light touch
No touch (proximity)
Electromyography or electroencephalogram driven

Outcomes

There are limited data on the long-term outcomes of patients with LIS. A series of retrospective phone surveys with 29 individuals who had survived LIS, or their caregivers, reported a 10-year survival rate of 83% and 20-year survival rate of 40%. Younger age at onset was predictive of longer survival, and quality of life was reported positively in those living in supportive home environments. Many in this cohort improved over time in object manipulation, oral communication, oral nutrition, and urinary continence.[34,35]

Motor Recovery

Unlike supratentorial strokes, LIS patients usually show recovery of limb movement from distal to proximal, often with trace isolated movements in fingers and toes appearing first and with persistent axial hypotonia. Recovery is asymmetrical, and a patient may regain far more motor ability on one side than the other. Classification of motor recovery in LIS is often described using five categories: no recovery, minimal recovery, moderate recovery, full recovery, and no neurologic deficit.[15]

Although there are no large multicenter studies available, multiple case series have described motor recovery in LIS. In 2003, Casanova reported outcomes in 14 patients who completed acute inpatient rehabilitation (10 classical LIS, 4 total LIS) and stratified based on those that had achieved full recovery, moderate recovery, minimal recovery, and no recovery based on Patterson and Grabois grading (Figure 6).[6,15] In a 1995 case series of 11 LIS patients, 10 regained precise distal motor control.[36] Doble described long-term upper limb function in patients with stable LIS finding that among 13 patients, four could trigger a switch, two could point or type, and two could lift an object.[35] In a more recent case series,

Table 4
Suitable assistive technology switches based on residual motor function

Switches Based on Motor Control
Breath Control: sip and puff
Bite & Release: mouth bulb (squeezable)
Gaze Control: electronic eye gaze system with augmentative communication device
Head Control: computer-mounted tracker camera, eyeglass appliances
Upper Limb Control: conventional mouse or modified hand-controlled mouse system

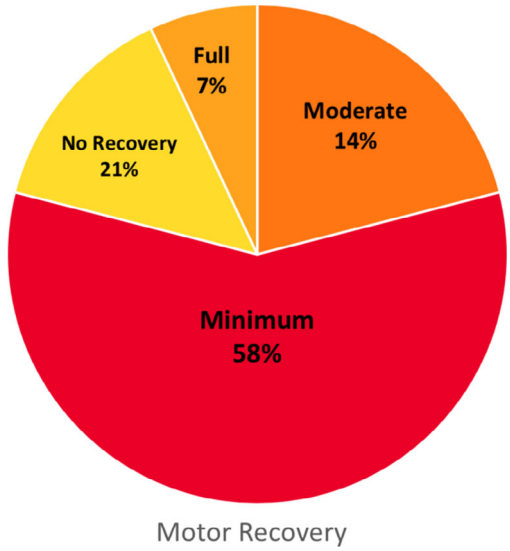

Figure 6. Casanova et al 2003 reported outcomes for 14 locked-in syndrome patients.

Hoyer and colleagues described nine patients with subacute or chronic incomplete LIS who participated in treadmill training, finding that five could walk or practice walking by the end of rehabilitation. All patients improved postural trunk and head control, physical endurance, and active mobility of the limbs. These case series demonstrate that although LIS can be physically devastating, a range of meaningful motor improvements can be regained for functional use.

The Impact of Balance

In stroke, sitting balance and trunk control on admission to acute rehabilitation are important predictors of independence in self-care and walking after stroke.[13] In LIS specifically, the axial trunk muscles are significantly affected with a negative impact on balance, usually well into the recovery process. However, we recently described a patient with incomplete LIS who made significant functional gains in self-care and mobility during inpatient rehabilitation that paralleled improvement in measures of balance.[37] Thus, gains in trunk control and balance predict improvement in overall function during rehabilitation care and is useful marker for overall prognosis.

Cognitive-Communicative Outcomes

Early after injury, cognitive function is limited by reduced attention, memory, and cognitive endurance. In chronic LIS cognitive function returns to normal, at least in those with pure pontine lesion.[38] New et al described the case of a patient with LIS who achieved full recovery (Patterson classification) and had neuropsychological testing performed at 6, 12, and 24 months after stroke. This patient had initial cognitive impairment and

Figure 7. Doble et al 2003 surveyed patients through their caregivers about life satisfaction, mood, and end of life.

distractibility but achieved improvement in verbal IQ to estimated premorbid levels by 12 months and executive function by 24 months.[39]

In the cohort reported by Casanova et al, 58% achieved stability in breathing patterns and were decannulated. Of these patients, 28% achieved verbal communication; 43% reached communication through devices such as a switch, letter board, or computer by hand, finger, or head movements; and 28% could communicate only through eye blinking.[6]

Life Satisfaction

The optimization of motor and neurological outcomes is an important purpose of rehabilitation because even limited physical function can improve quality of life and improve the likelihood of returning home with family.[8] In the long-term follow up study by Doble et al of 13 patients, seven survivors were satisfied with life in general, five were occasionally depressed, and for one the caregiver was unable to assess mood. As reported by caregivers, seven patients had never considered or discussed euthanasia, one patient wished to die, and notably no survivors had a Do Not Resuscitate order (Figure 7).[35] Critically, these interviews were conducted only with patients living at home with caregiver support. In a study by Bruno et al of 65 LIS members of the French Association for LIS, 72% professed happiness and 28% professed unhappiness. Unhappiness was associated with shorter duration of LIS, dissatisfaction with community mobility, recreational activities, capacity to deal with life events, nonrecovery of speech, and anxiety.[40] Happiness in LIS was associated with recovery of speech production, absence of anxiety, and greater time spent with LIS.

Conclusion

LIS is often reported to be a devastating diagnosis with poor prognosis for long-term recovery. Indeed, outcomes for patients with classical LIS are limited, and yet meaningful functional gains can be achieved in the long term, especially for those who have supportive family and caregivers at home. For those with incomplete LIS, the prognosis for independence in many activities is better, but caregiver assistance is often still required. The majority of those living at home report good life satisfaction; however, return home requires significant personal and financial commitment from the patient and family. Attentive and knowledgeable rehabilitation care of these medically and neurologically complex patients is critical for facilitating optimal functional recovery, building confidence in both patient and caregivers and for increasing the likelihood of returning home. The primary role of the physiatrist and the interdisciplinary team, as always, is to provide knowledgeable care, proficient medical management, thorough education, and continued encouragement throughout the process of recovery.

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Disclosure

E.F., K.A. and R.L.H. Shirley Ryan AbilityLab, Northwestern University Feinberg School of Medicine, Chicago, IL, USA. Address correspondence to: R.H; email: rharvey@sralab.org

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