Locked-In Syndrome

A collection of research reviews on rehabilitation topics from NARIC and other information resources.

At the National Rehabilitation Information Center, Information Specialists field requests on a wide range of disability rehabilitation issues. While serving as a dissemination center for NIDRR funded research, NARIC serves persons with disability and their families by providing information and referral on a variety of rehabilitation and disability topics. This two part edition of reSearch focuses on two specific topics affecting our consumers: Locked-In and Stiff-Person Syndromes.

According to the National Institute of Neurological Disorders and Stroke Information Page, Locked-in syndrome is a rare neurological disorder characterized by complete paralysis of voluntary muscles in all parts of the body except for those that control eye movement. It may result from traumatic brain injury, diseases of the circulatory system, diseases that destroy the myelin sheath surrounding nerve cells, or medication overdose. Individuals with locked-in syndrome are conscious and can think and reason, but are unable to speak or move. The disorder leaves individuals completely mute and paralyzed. Communication may be possible with blinking eye movements. (http://www.ninds.nih.gov/disorders/lockedinsyndrome/lockedinsyndrome.htm)

Our search resulted in a range of documents from the most current through the past five years of Locked-In Syndrome research. Searches in NARIC REHABDATA and National Library of Medicine database, PubMed resulted in over 60 search terms. A sample of these terms is listed below:

- Combined Modality Therapy
- Communication/Devices
- Consciousness Disorders
- Early Intervention
- Ethics (clinical)
- Eye Movements
- Motor Skills
- Muscle Spasticity/Weakness
- Paralysis
- Post-Operative Complications
- Practice Guidelines
- Psychosocial Factors
- Quality of Life
- Rare Disorders
- Speech Impairments/Therapy
- Spinal Injuries (complications)
- Stroke
- Thrombolytic Therapy
- Treatment Outcomes

There is a limited amount of information available on Locked-In Syndrome. Our search of REHABDATA resulted in four documents ranging from 2005-2003. The PubMed database search resulted in 16 documents ranging from 2006-2001. The complete citations are included at the end of this research brief.

In addition to document searches, NARIC searched its Program database of National Institute on Disability and Rehabilitation Research (NIDRR) projects to locate grantees/projects related Locked-In Syndrome. While there are no specific grantees/projects on Locked-In Syndrome; there were several projects which addressed the symptoms and disabilities presented by this syndrome which may be useful to our patrons:

Rehabilitation Engineering Research Center on Mobile Wireless Technologies for Persons with Disabilities
Project Number: H133E010804
(Active)

Rehabilitation Engineering Research Center on Communication Enhancement
Project Number: H133E030018
(Active)

Practical Clinical Trial of Cognitive Rehabilitation in Neurologic Illness
Project Number: H133G050063
(Active)
Documents from NARIC’s REHABDATA search listed are listed below:

2005


NARIC Accession Number: J48659

**ABSTRACT:** Case study describes the cognitive functioning in a person with locked-in syndrome due to purely pontine lesions and documents the process of recovery by serial testing over a lengthy period. A previously well man in his thirties was admitted to the hospital with progressive stroke symptoms and persisting clinical features of LIS. He had minimal change during the first month, then slowly improved and was discharged home seven months after stroke. Neuropsychological testing, conducted at 6, 12, and 24 months after stroke, showed cognitive dysfunction, including mild difficulties with attention and concentration, significant reduction in speed of processing, moderate impairment of perceptual organization skills, mild inefficiencies in new learning of verbal information, and a moderate reduction in executive skills. There was progressive improvement in most areas of physical and cognitive functioning until at least two years after stroke.

2003


NARIC Accession Number: O15496

**ABSTRACT:** Paper describes the development of the prototype “aware system”, designed to assist patients with locked-in syndrome to use brain signals to communicate and control their environment. Four factors were considered important when designing the system interface: speed, accuracy, context, and the mood of the patient. This paper was presented at the 2003 annual conference of RESNA, the Rehabilitation Engineering and Assistive Technology Society of North America and is available on CD-ROM.


NARIC Accession Number: J45714

**ABSTRACT:** Study evaluated the effects of early and intensive rehabilitation on the recovery of patients with locked-in syndrome (LIS). Researchers evaluated 14 patients with LIS who received intensive nursing care and early rehabilitation, begun within 1 month of the morbid event, which included physical therapy and respiratory, swallowing, and speech training. Functional recovery during hospitalization was evaluated based on changes in motor recovery, breathing, swallowing, communication, gaze, and bowel and bladder control. After discharge, follow-up telephone calls were made to determine status. Significant motor recovery was found in 21 percent of subjects, within 3 to 6 months of onset of onset; complete swallow recovery in 42 percent; verbal communication in 28 percent; communication through devices in 42 percent effective bladder and bowel control in 35 percent; and good breathing patterns in 50 percent. At follow-up, the mortality rate was 14 percent and only two complications were reported.


NARIC Accession Number: J46604
ABSTRACT: Study examined the long-term survival rate, functional status, and quality of life in 29 patients with locked-in syndrome (LIS) who were included in an initial cohort 11 years earlier. Fourteen patients were still living at the 11-year follow-up. Telephone interviews were conducted with one patient (by computer) and the caregivers of 12 others; one patient was lost to follow-up. Data collected revealed a 5- and 10-year survival rate of 83 percent and a 20-year survival rate of 40 percent. Ten patients had not been hospitalized in the previous year; eight lived with family. Improvements in communication related to technology, including computer and Internet access. Eleven left home at least monthly. Caregivers reported that seven patients expressed satisfaction with life; five were occasionally depressed. Findings indicated that individuals with locked-in syndrome can have prolonged survival, can live in the community if there is enough support, and have some measure of quality of life.

Documents from the National Library of Medicine PubMed search at http://www.pubmed.com/ are listed below:

2006

PMID: 16575301
ABSTRACT: OBJECTIVE AND IMPORTANCE: Locked-in syndrome is a state of preserved consciousness in the setting of quadriplegia, anarthria, and usually also includes lateral gaze palsy. It is most commonly associated with upper brainstem infarction variably sparing the third cranial nerve nucleus. There are likely many etiologies that contribute to this clinical syndrome. These are incompletely understood, and the syndrome remains a rare but devastating complication that can occur after neurosurgical and neurovascular interventions. Advanced magnetic resonance imaging techniques such as perfusion and diffusion tensor imaging may help to elucidate the mechanism behind locked-in syndrome. To the authors’ knowledge, there are no reports in the literature of perfusion and diffusion tensor findings in patients with this syndrome. A postprocedural case of locked-in syndrome is described with abnormalities on perfusion and diffusion tensor imaging in the absence of any changes in conventional magnetic resonance imaging.

CLINICAL PRESENTATION: A 57-year-old man who presented with acute onset headache, ataxia, and other nonspecific symptoms was found on imaging to have a giant fusiform basilar artery aneurysm. INTERVENTION: A saphenous vein bypass between the proximal right external carotid artery and P2 segment of the right posterior cerebral artery followed immediately by endovascular embolization of the aneurysm sac and distal left vertebral artery was performed. CONCLUSION: Postprocedural angiography demonstrated patency of the bypass graft, and diffusion weighted imaging showed no evidence for acute brainstem infarction. Nevertheless, despite technically successful procedures and the absence of abnormalities on conventional magnetic resonance imaging, the patient developed quadriplegia and anarthria and remained in a locked-in state until he expired. Abnormalities were, however, seen on both perfusion and diffusion tensor imaging, where hypoperfusion, increased mean diffusivity, and decreased fractional anisotropy were observed in the ventral brainstem. The findings suggested a disruption of pontine white matter tracts. Advanced imaging techniques may allow us to image important microstructural changes that were previously not discernable and assist in the evaluation of patients with complex neurological sequelae such as locked-in syndrome.

Perrin, F., Schnakers, C., Schabus, M., Degueldre, C., Goldman, S., Bredart, S., Faymonville, M.E.,

\textbf{ABSTRACT:} A major challenge in the management of severely brain-injured patients with altered states of consciousness is to estimate their residual perception of the environment. 

\textbf{OBJECTIVE:} To investigate the integrity of detection of one’s own name in patients in a behaviorally well-documented vegetative state (VS), patients in a minimally conscious state (MCS), and patients with locked-in syndrome. 

\textbf{DESIGN:} We recorded the auditory evoked potentials to the patient’s own name and to seven other equiprobable first names in 15 brain-damaged patients. 

\textbf{RESULTS:} A P3 component was observed in response to the patient’s name in all patients with locked-in syndrome, in all MCS patients, and in three of five patients in a VS. P3 latency was significantly (P greater than 0.05) delayed for MCS and VS patients compared with healthy volunteers. 

\textbf{CONCLUSIONS:} These results suggest that partially preserved semantic processing could be observed in noncommunicative brain-damaged patients, notably for the detection of salient stimuli, such as the subject’s own name. This function seems delayed in MCS and (if present) in VS patients. More important, a P3 response does not necessarily reflect conscious perception and cannot be used to differentiate VS from MCS patients.


\textbf{ABSTRACT:} Sildenafil citrate has been shown to enhance neurogenesis, angiogenesis, synaptogenesis, and neurological outcome by augmentation of cyclic guanosine monophosphate (cGMP) levels in animal models of ischemic stroke. Whether sildenafil citrate may be helpful for recovery in human stroke is unknown at this time.

\textbf{METHODS:} A 41-year-old woman with locked-in syndrome due to pontine infarction began receiving 150 mg of oral sildenafil citrate daily on a compassionate use basis in August 2003 and continues treatment at this time. Magnetoencephalography (MEG) was performed at 12 and 17 months after stroke. 

\textbf{RESULTS:} No serious adverse events have occurred. Significant milestone recoveries including standing, use of both arms, talking, and full return of swallowing have occurred, particularly after nine months of treatment. The MEG showed a significantly increased amplitude in the somatosensory cortex. 

\textbf{CONCLUSION:} Daily use of high dose sildenafil citrate appears to be safe in this patient with stroke resulting in locked-in syndrome. Further studies will be required to establish safety and efficacy.

\textbf{2005}


\textbf{ABSTRACT:} No abstract is available.


\textbf{ABSTRACT:} Widespread use of intravenous tissue plasminogen activator (t-PA) for acute ischemic stroke is limited by multiple contraindications to its use. CASE REPORT: This article describes a patient with stuttering symptoms of pontine ischemia caused by verteobasilar dissection who suddenly deteriorated into a locked-in state 32 hours after symptom onset. The quadriplegia was successfully reversed within three hours of onset with the combination of pharmacologically induced hypertension, anticoagulation, and intravenous t-
PA. DISCUSSION: Even in the face of numerous contraindications (including hypertension, anticoagulation, and treatment beyond three hours of symptom onset), intravenous t-PA can be used successfully in carefully selected cases.


ABSTRACT: The locked-in syndrome (pseudocoma) describes patients who are awake and conscious but selectively deafferented, i.e., have no means of producing speech, limb or facial movements. Acute ventral pontine lesions are its most common cause. People with such brainstem lesions often remain comatose for some days or weeks, needing artificial respiration and then gradually wake up, but remaining paralyzed and voiceless, superficially resembling patients in a vegetative state or akinetic mutism. In acute locked-in syndrome (LIS), eye-coded communication and evaluation of cognitive and emotional functioning is very limited because vigilance is fluctuating and eye movements may be inconsistent, very small, and easily exhausted. It has been shown that more than half of the time it is the family and not the physician who first realized that the patient was aware. Distressingly, recent studies reported that the diagnosis of LIS on average takes over two and half months. In some cases it took four to six years before aware and sensitive patients, locked in an immobile body, were recognized as being conscious. Distressingly, recent studies reported that the diagnosis of LIS on average takes over two and half months. In some cases it took four to six years before aware and sensitive patients, locked in an immobile body, were recognized as being conscious. Once a LIS patient becomes medically stable, and given appropriate medical care, life expectancy increases to several decades. Even if the chances of good motor recovery are very limited, existing eye-controlled, computer-based communication technology currently allow the patient to control his environment, use a word processor coupled to a speech synthesizer, and access the worldwide net. Healthy individuals and medical professionals sometimes assume that the quality of life of an LIS patient is so poor that it is not worth living. On the contrary, chronic LIS patients typically self-report meaningful quality of life and their demand for euthanasia is surprisingly infrequent. Biased clinicians might provide less aggressive medical treatment and influence the family in inappropriate ways. It is important to stress that only the medically stabilized, informed LIS patient is competent to consent to or refuse life-sustaining treatment. Patients suffering from LIS should not be denied the right to die - and to die with dignity - but also, and more importantly, they should not be denied the right to live - and to live with dignity and the best possible revalidation, and pain and symptom management. In our opinion, there is an urgent need for a renewed ethical and medicolegal framework for our care of locked-in patients.


ABSTRACT: The locked-in syndrome (LIS) is a severe condition originated by a ventral pons lesion causing quadriplegia and anarthria but with a preserved consciousness. LIS seems to be a well defined clinical picture, although different problems still persist, such as the diagnosis as it is usually mistaken for akinetic mutism and a vegetative state; the unclear prognosis, because of the patient’s psychological state and the lack of information and data concerning the different types of available treatment and the need for results. Rehabilitation is a challenge for physicians, new methods and techniques of specialized treatments for these patients are opening a new future that will allow us to abandon the initial pessimism. A more efficient rehabilitation of these patients depends on the intensity of the rehabilitation, the multidisciplinary approach, and duration of the treatment.

ABSTRACT: No neuropsychologic studies have been reported that assess cognitive functioning in survivors of locked-in syndrome (LIS) due to purely pontine lesions and then document the process of recovery by serial testing over a lengthy period. A previously well man in his early thirties was admitted to the hospital with progressive stroke symptoms and signs. Investigations showed occlusion of the basilar artery and acute infarction of the pons, including basis and tegmentum. Despite thrombolysis, he had persisting clinical features of the LIS. He had minimal change during the first month but then slowly improved. Recovery continued gradually, and he was discharged home seven months after stroke; at this time he was ambulating with a cane, was mildly dysarthric, was able to swallow foods of modified consistency, and was independent in all self-care activities. Neuropsychologic testing, done six months after stroke, showed notable cognitive impairments. These included mild difficulties with attention and concentration, significant reduction in speed of processing, moderate impairment of perceptual organization skills, mild inefficiencies in new learning of verbal information, and a moderate reduction in executive skills. Pathologic laughing and crying were also noted. There was progressive improvement in most areas of physical and cognitive functioning until at least two years after stroke. Neuropsychologic testing in this patient suggests that the LIS may be associated with impairments of higher-level cognitive functioning.


ABSTRACT: Review, no abstract available.


ABSTRACT: Today, thanks to intensive care treatment and modern diagnostic tools, increasingly more patients with severe brain and spinal cord lesions, mainly secondary to accidents, stroke, tumours, and congenital malformations survive the acute impact on the central nervous system (CNS). Complicated operative procedures and concomitant complication may also lead to severe impairment of the sensory motor and cognitive behavioural functioning as it can be described according to the WHO-ICF criteria. New developments of functional neurorehabilitation in neurosurgery can significantly improve patients’ quality of life (QoL) in terms of both brain and body functioning and certain health-related components of well-being (such as social activities and leisure). Rehabilitation starts with assessment of the functional impairment and the underlying pathophysiology by using all modern diagnostic tools. Our concept of postoperative neurorehabilitation is exemplarily demonstrated in one patient who suffered from acute postoperative locked-in syndrome. Surgical decompression and fusion were required for post traumatic and recurrent congenital craniovertebral instability at C0-C1. Subsequent functional neurorehabilitation is based on careful planning in accordance with our concept of a holistic Spectrum of functional early Neurorehabilitation.


ABSTRACT: We describe the locked-in syndrome in a 31-year-old patient after right upper lobectomy for suspected metastasis. After surgery, vertical eye movement was her only means of communication. She remained in a ‘locked-in’ state for seven months before dying. Post-mortem examination showed
extensive metastatic tumour deposition in the ventral pons. We discuss the clinical features of this syndrome and factors that may affect onset, diagnosis and management.

ABSTRACT: We report a rare case of posttraumatic locked-in syndrome (LIS) that appeared after an unusual three-day delay. LIS was diagnosed according to clinical status and diagnostic methods (roentgenograms, computerized tomography, transcranial Doppler, electroencephalography, magnetic resonance imaging). A 14-year-old girl had a cervical spine injury during floor exercises that provoked LIS 72 hours after trauma. A rapid diagnosis of basilar thrombosis followed by antioedema and continuous anticoagulant therapy significantly improved the neurological status. In conclusion, posttraumatic locked-in syndrome can have sometimes a prolonged three-day delay in the clinical appearance.

ABSTRACT: Locked-in syndrome is a dramatic clinical condition, the patient is awake, can listen and breathe, but is unable to move any muscle, conserving only the vertical eye movements. The most common cause of locked-in syndrome is the thrombosis of the basilar artery and commonly leads to death, frequently due to pneumonia. Intravenous and intra arterial thrombolysis have been used successfully in a selective group of patients with ischemic stroke. There is only one report of two patients with locked-in syndrome who were treated successfully with intra arterial thrombolysis. Other authors, based in their experiences, do not recommend this treatment. We report two female patients aged 63 and 26 years, with Locked-in syndrome due to a basilar thrombosis who were treated successfully with intra arterial thrombolysis using recombinant tissue plasminogen activator (r-TPA). The lapses between the onset of the symptoms and thrombolysis were 5 and 8 hours respectively. A complete recanalization was obtained in both patients during the thrombolysis. One year after, the first patient has only a moderate ataxia, walking with assistance and the other has a normal neurological examination.

ABSTRACT: No abstract is available.

2002

ABSTRACT: We describe a patient with locked-in syndrome who had minimal volitional motor function and severe spasticity in all four extremities. The patient showed a significant improvement in volitional motor function following intrathecal baclofen pump therapy to control spasticity. This case study suggests that intrathecal baclofen pump therapy might improve motor function in select patients with locked-in syndrome.

ABSTRACT: The locked-in syndrome (LIS) is a
very severe condition caused by a primary vascular or traumatic injury to the brainstem, normally corresponding to a ventral pons lesion due to an obstruction of the basilar artery, and characterized by upper motor neuron quadriplegia, paralysis of lower cranial nerves, bilateral paresis of horizontal gaze and anarthria, and with preserved consciousness. Patients who have suffered this pontine lesion generally have preserved vertical eye movements and movement of the eyelids (blinking), this being their only means of responding to the outside world. A survey was conducted of 44 people diagnosed with LIS, all of them belonging to the Association of Locked-in Syndrome of France. Results of this survey showed that LIS was equally frequent in men and women (51.2 percent vs. 48.1 percent) and had occurred at any age between 22-77 years of age (normally between 41-52 years, the mean age being 46.79 years). The average time that transpired post-insult was 71.35 months. The principal cause of LIS was stroke (86.4 percent), with traumatic brain injury (TBI) being a distant second cause with an incidence of only 13.6 percent. The diagnosis of LIS was usually made around the middle of the second month after onset (mean of 78.76 days). The principal treatments, when present, were pharmacological and physiotherapy. However, 47.1 percent of the patients were not receiving treatment of any kind at the time of the survey. Neuropsychologically, 86 percent had a good attentional level, 97.6 percent were temporally oriented and 76.7 percent could read; 18.6 percent reported memory problems and 24 percent showed visual deficit (found mainly in patients with LIS originated by TBI); 47.5 percent reported a good mood state and 12.5 percent reported feeling depressed; 61.1 percent reported having sexual desire, but only 30 percent maintained sexual relations; 78 percent were capable of emitting sounds and 65.8 percent could communicate without technical aid; 73.2 percent enjoyed going out and 81 percent met with friends at least twice a month. Only 14.3 percent participated in social activities and 23.8 percent watched television regularly. Nearly 100 percent of the patients reported being sensitive to touch to any part of their bodies. This survey suggests diagnostics and rehabilitation procedures.

ABSTRACT: This paper describes various approaches of treatment to a male client suffering from the locked-in syndrome following closed head injury. During the course of therapy, which started more than five years after onset and lasted for as much as 11 years, the client progressed from the so-called classical to the incomplete locked-in state. The different approaches as well as the outcomes are presented in detail, followed by a discussion about interdisciplinary issues, duration of treatment and the impact of a patient’s personality on the course and goals of the treatment as well as on the mode of communication.

2001

ABSTRACT: No abstract is available.

ABSTRACT: Locked-in syndrome is a neurological condition due to a brain disease or an injury affecting the brainstem. The symptoms are tetraplegia, double-sided facial paresis, anarthria/dysarthrophonia, dysphagia and reactive involuntary laughing and crying. Vertical eye movements are the only commonly remaining voluntary motor function. Although the linguistic
abilities as well as intellectual and emotional functions as a whole remain intact, all the motor abilities of self expression are lost. Seventeen chronic locked-in syndrome patients referred to Kapyla Rehabilitation Centre between 1979-2000 are reported. The multidisciplinary rehabilitation team developed an individual alternative communication method for all patients and trained them to use it by minor movements of e.g. thumb, chin or head. An alternative communication method enabled most of the patients to interact with other people using practical as well as theoretical thinking and decision making.

About reSearch:

reSearch is a new information product from the National Rehabilitation Information Center (NARIC). Each issue is based on real-world queries received by our information specialists from researchers, educators, and rehabilitation professionals around the world.

We search several sources both in-house and online, to fill these requests including:

- REHABDATA and the NIDRR Program database
- Education Resources Information Center
- National Clearinghouse of Rehabilitation Training Materials
- Campbell and Cochrane Collaborations
- PubMed and other National Library of Medicine databases
- Agency for Health Care Policy and Research databases
- Center for International Rehabilitation Research Information and Exchange

and other reputable, scholarly information resources.

We hope you find these reSearch briefs informative in your own research.

- NARIC Information and Media Team