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**Türk Fizyoterapi ve Rehabilitasyon Dergisi**

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# ROLE OF PHYSIOTHERAPY AND REHABILITATION WITH COOPERATION OF FAMILY IN LOCKED-IN SYNDROME: FOUR-YEAR FOLLOW-UP OF A CASE

## CASE PRESENTATION

### ABSTRACT

**Purpose:** Locked-in syndrome (LIS) is characterized by quadriplegia, lower cranial nerve palsy, and mutism. In its classic type, patients are only able to move their eyes vertically and blink their upper eyelids. In the classic type of LIS, the patients are depended on a bed, and all their systems are affected due to immobilization. We presented a four-year follow-up of a case of LIS undergoing physiotherapy and rehabilitation

**Methods:** A 51 years old male patient with a classic type of LIS was applied preventive and supportive physiotherapy and rehabilitation, and the results of the four-year follow-up were presented. The physiotherapy and rehabilitation program consisted of the passive range of motion, positioning, passive cycling, and supported standing up with a device by the family for seven days a week, and electrotherapy application, mobilization techniques and bronchial drainage by a physiotherapist at least three days per week throughout four years.

**Results:** The patient was free from muscle shortness or joint limitations over four years. There was not any development of scar or wound on the skin due to immobilization. The patient was able to manage passive sitting and standing up position for 15 minutes, and communicate with blinks.

**Conclusion:** Preventive and supportive physiotherapy and rehabilitation practice in combination with the family and caregiver involvement are indispensable for patients in cases requiring care such as LIS.

**Key Words:** Locked-in syndrome; Physiotherapy; Rehabilitation.

# LOCKED-IN SENDROMLU BİR HASTADA AİLE KATILIMIYLA GERÇEKLEŞTİRİLEN FİZYOTERAPİ VE REHABİLİTASYONUN ROLÜ: BİR OLGUNUN DÖRT YILLIK İZLEMİ

## OLGU SUNUMU

### ÖZ

**Amaç:** Locked-in Sendromu (LIS) kuadripleji, alt kranial sinir felci ve mutizm ile karakterize bir durumdur. Klasik tipte, hastalar sadece gözlerini dikey olarak hareket ettirebilir ve üst göz kapağını kapatabilirler. Klasik LIS tipinde hasta yatağa bağımlıdır ve tüm sistemler immobilizasyondan etkilenir. Bu olgu sunumunda, LIS olan bir olguya dört yıllık süre boyunca uygulanan fizyoterapi ve rehabilitasyon programının etkileri incelendi.

**Yöntem:** Klasik tip LIS tanısı olan 51 yaşındaki erkek olguya önleyici ve destekleyici fizyoterapi ve rehabilitasyon programı uygulandı ve dört yıllık takip sonuçları sunuldu. Rehabilitasyon programı dört yıl boyunca haftada yedi gün olacak şekilde aile tarafından yaptırılan pasif eklem hareket açıklığı egzersizleri, pozisyonlama, pasif bisiklet aktivitesi, destekli ayağa kalkma aktivitesi ve haftada en az üç gün fizyoterapist tarafından yapılan elektroterapi uygulaması, mobilizasyon teknikleri ve bronşial drenaj yöntemlerinden oluştu.

**Sonuçlar:** Dört yıllık takip sonucunda, olguda herhangi bir eklem hareket kısıtlılığı veya kas kısalığı gelişmedi. Ayrıca, immobilizasyona bağlı olarak deride skar veya yara oluşmadı. Olgu 15 dakika boyunca pasif oturma ve ayakta durma pozisyonunu tolere edebiliyor ve göz hareketleri ile iletişim kurabiliyordu.

**Tartışma:** Bu sonuçlar destekleyici ve önleyici fizyoterapi ve rehabilitasyon uygulamalarının ev programı ile kombine edilerek, aile ve bakım veren katılımının, LIS gibi bakım gerektiren durumlarda tedavinin ayrılmaz bir parçası olduğunu göstermektedir.

**Anahtar Kelimeler:** Locked-in Sendromu; Fizyoterapi; Rehabilitasyon.

## INTRODUCTION

Locked-in syndrome (LIS) is a rare neurological disorder usually resulting from infarction of the ventral pons. Patients with LIS are diagnosed with quadriplegia, lower cranial nerve paralysis, and mutism. Patients are awake and conscious and also may be able to communicate with others through coded messages by blinking or moving their eyes vertically, which are often not paralyzed (1-2). The LIS may be divided into three categories: the classic form, the incomplete form (patients have other voluntary movements), and the complete form with complete paralysis of all voluntary muscles where the eyes are paralyzed as well (3). The classic form of LIS typically results from an infarct, hemorrhage, or trauma of ventral pons that causes quadriplegia, anarthria, and preservation of consciousness. Patients communicate non-verbally via vertical eye movements.

The disorder leaves the individual wholly mute and paralyzed. Patients may not breathe spontaneously (4), and the patient may need invasive mechanical ventilation. This condition makes the patient immobile. All systems, especially the musculoskeletal system and the pulmonary system, are affected (5,6). Having a rest in bed is common practice worldwide, especially for mechanically ventilated patients. Pneumonia is the most common cause of death in LIS (7). The average ten-year survival rate for people with LIS have been reported as high as 80% which highlights the importance of improving quality of life and allowing patients to return to live at home with their family. Therefore, early referral to a specific rehabilitation program including specialist care and technology must be considered a cornerstone (8,9).

Intensive and early referral to rehabilitation improves functional recovery, quality of life, and reduce mortality rate (10,11). When initiated shortly after the start of mechanical ventilation, mobilization and pulmonary rehabilitation can play an essential role in decreasing the adverse effect of mechanical ventilation.

In this case report, the benefits of a preventive and supportive physical therapy program of the mechanically ventilated patient who was diagnosed with the classic type of LIS that was followed up for four years at home were presented. This case report may be relevant regarding a long-term follow-up on a rare case situation and may set an example for clinicians and physiotherapists working with LIS patients to establish a home rehabilitation program after discharge.

## CASE REPORT

The case was 51 years old male patients with LIS. The patient was hospitalized at a university hospital four years ago with a complaint of mechanical talk while trying to fall asleep during the night. The patient was diagnosed with the classic type of LIS due to basilar artery aneurysm and treated with a coiling aneurysm, implanting a stent to basilar artery. There was a previous hypertension history, complaint of numbness in hands, impaired balance while walking, and stumbling before a couple of days after the incident. After having medical treatment specific to LIS, the patient was referred to physiotherapy and rehabilitation program. The patient was included in the rehabilitation program for five days per week at the hospital in acute stage by a physiotherapist. Then, the patient was discharged from the hospital to home. After discharge, the patient's family wanted to continue physiotherapy at home. The family was informed about the goals of physiotherapy. The physiotherapy and rehabilitation program was continued for four years at home.

All procedures followed were in accordance with the Helsinki Decleration. Because the patient could not do any voluntary movement except eyes movement, an eyes approval from the patient and a written informed consent was obtained from his family.

Although, the patient was observed and evaluated every treatment session, baseline, first and fourth years' status was recorded. At baseline, the patient was able to maintain respiratory pattern voluntarily. The range of motion was measured using goniometer passively, and no limitation was detected. The patient's body was observed for skin status. There was not any scar tissue or wound formation. The patient could not contract any of the muscles voluntarily. The patient could not control his head. He could not to rotate in the bed, sit and stand. The patient did not tolerate the passive

standing upright position because of the presence of postural hypotension. The patient could not talk, and he was communicating with blinks. He had no swallowing reflex. The patient's saliva was being aspirated through nose and mouth by the caregiver using an aspirator.

Initially, the physiotherapist informed the family to design the home based on the needs of the patient. Besides, she identified the tools and machines that would be needed at home. It was advised to take a hospital bed, wheelchair standing up a device for the optimal care at home. Additionally, two electrotherapy devices, an aspirator device, a humectation device, and a mechanical ventilator were needed over time. The family provided these machines through their resources.

The goal of the physiotherapy and rehabilitation was to preserve the current status and to prevent the development of possible complications such as pneumonia, muscle shortness, joint limitation, and scar tissue or wound formation. The treatment program was modified based on the specific needs of the patient (e.g., if the patient is not feeling well, the standing up or the cycling activity time is changed from the morning to the afternoon or vice versa). The goals and content of the treatment were to preserve the range of joint motion and muscle extensibility, to preserve muscle structure and improve muscle strength, and to clean airways and preserve thoracic expansion. Proprioceptive neuromuscular facilitation techniques, connective tissue mobilization, cervical, lumbar and sacral mobilizations joint mobilization provided by physiotherapist at least three days per week to preserve range of joint motion and muscle extensibility. To improve secretion clearance and to maintain thoracic expansion, positioning with postural drainage positions (including head down positions) and manual techniques (percussion, shaking, vibration) were used. Neuromuscular electric stimulation (NMES) was applied to extensor and flexor muscle groups of the extremities for 30 minutes each with the electrotherapy device (Itelect Advanced Chattonoga, Guildford Surrey, UK) to maintain muscle structure and to improve muscle strength.

The treatment program consisted of normal range of motion exercise twice daily, positioning for 30 minutes twice a day (started three years ago after pneumonia), cycling activity in sitting position using leg and arm ergometer (MOTOmed viva 2, Reck Tecnic GmbH&Co. KH, Germany) (Figure 1) once a day, standing upright with assistance (Squod, Vermeiren, Kalmhout, Belgium) (Figure 2) for 15 minutes once a day was taught to family as a home exercise program and were performed by the family and caregiver for seven days per week for four years, and the physiotherapist revised the program once a week.

The results of the one- and four-year follow up are shown in Table 1. The patient began to be mechanically ventilated after having pneumonia. There was no muscle shortness or joint limitations based on the range of motion assessment. No scar tissue or wound formation had been developed on the skin. The patient could not contract any muscle and did not have an active sitting ability. However, the patient was able to manage passive sitting and passive standing upright position for 15 minutes. He can communicate with blinks. There was no active swallowing. Although the patient's physical status was not improved significantly, the patient's condition did not deteriorate, and complications did not develop.

## DISCUSSION

In this study, we presented a four-year physiotherapy and rehabilitation practice along with a home program of a case with LIS due to basilar artery aneurysm. By following up with four years, it was found that respiratory status improved and there was no muscle shortness or joint limitations. It was also found that no scar or wound was developed on the skin depending on immobilization. The patient was able to manage passive sitting and standing up position for 15 minutes and communicate with blinks.

The management of patients with LIS can be quite challenging and frustrating. Despite early surgery and physiotherapy and rehabilitation protocol, there was not any physical recovery due to the characteristic of the syndrome. Physiotherapy and rehabilitation of such conditions require palliative care which includes supportive, preventive, and protective treatment goals and the involvement

**Table 1:** The Results of One-Year and Four-Year Follow-Up.

| Status | Baseline | 1-Year Follow-up | 4-Year Follow-up |
|---|---|---|---|
| **Respiratory Status** | Tracheostomy | Mechanical ventilation with a tracheostomy | Mechanical ventilation with a tracheostomy |
| **Skin and Range of Motion** | No muscle shortness<br>No joint limitation<br>No scar tissue | No muscle shortness<br>No joint limitation<br>No scar tissue | No muscle shortness<br>No joint limitation<br>No scar tissue |
| **Functional Status** | Cannot sit or stand actively or passively | Passive sitting and passive standing at least 15 minutes | Passive sitting and passive standing at least 15 minutes |
| **Communication** | With blinks | With blinks | With blinks |

**Figure 1:** Cycling Activity in Sitting Position with Leg and Arm Ergometer.

of the family in treatment progress to prevent possible complications (10,11). We explained our aim to the patient and his family, and we developed a special daily activity program.

Literature suggests that rehabilitation of patients with LIS has different strategies depending on the phases and patient needs. During acute phase in the intensive care unit, the patient was provided intensive nursing care, such as positioning (lying prone and supine) in bed for comfortable sitting, bronchial drainage, and elevation of the extremities to prevent edema of the paralyzed hands and feet, aspiration of bronchial secretions, monitoring of fluid balance and vital parameters for possible complication of immobilization. After intensive care unit discharge, the rehabilitation program consists of muscle strengthening, limb mobilization to prevent secondary complications

**Figure 2:** Standing-Up with Life Stand Assistance.

(tendon retractions, periarticular calcifications), and attaining head and trunk control in the physiotherapy and rehabilitation clinic. The respiratory approach involved positioning for postural drainage and the use of manual techniques to improve airway clearance. Swallowing training emphasized stimulation of swallowing reflexes and buccal-lingual facial movements and introducing increasing amounts of semisolid food and thin fluids.

Before discharge from the center, family members or caregivers should have an idea of the life at home, and possible complications or necessary precautions (10). A case study has demonstrated that patient may benefit from physiotherapy and rehabilitation even if implemented 16 years after the development of LIS (12). As in this case, the application of physiotherapy and rehabilitation and involvement of the family and caregiver may have positive results in a bedridden patient without any active movement for four years, and the patient had no bedsore.

The most common cause of mortality in patients with LIS is lung infection (13,14). Therefore, we included respiratory physiotherapy to prevent lung infection. However, application of postural drainage and manual techniques, applied only one hour per day, for three days per week, were unable to prevent the development of pneumonia and the use of invasive mechanical ventilation. We thought that positioning and mobilization of the patient by the family or caregiver should be part of the daily routine of the patients with LIS. The patient was free from pneumonia despite mechanical ventilation probably due to the effects of respiratory physiotherapy which was applied by the family as well as the physiotherapist.

The NMES represents a practical and feasible interventional strategy to prevent skeletal muscle atrophy in critically ill patients (15). The NMES represents an effective method to alleviate muscle disuse atrophy in healthy subjects, and improves muscle performance and exercise tolerance, and stimulates active movement, maintains muscle mass, and prevents changes in hemodynamic response (15,16). In this case, we applied NMES to the flexor and extensor muscle for all of these aims.

In conclusion, the results illustrate an example of the importance of the involvement of the family and caregivers into physiotherapy and rehabilitation of a case with LIS.

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***Ethical Approval:*** A case study. All procedures followed were in accordance with the Helsinki Declaration

***Informed Consent:*** Because the patient could not do any voluntary movement except eyes movement, an eyes approval from the patient and a written informed consent was obtained from his family.

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