

Palliative care and locked-in syndrome: Brazilian case report

Abstract

The Locked-in syndrome (LIS) is characterized by tetraplegia, aphasia and preservation of the level of consciousness, and it's associated with a poor prognosis. The palliative approach to these patients is a challenge. This article aims to examine how palliative care could improve LIS patients' quality of life. It is a case report of a male patient, 29 years, married, photographer, with LIS after traumatic brain injury (TBI), assisted at a hospice in São Paulo. Palliative Care for situations like this is still a relatively unexplored field; in Brazil, there are no published studies. The identification LIS is a clinical challenge and misdiagnosis can occur; the association of serial clinical examination, imaging tests and multimodal neurophysiological evaluation is necessary. The care provided by the Palliative Care team provided symptom control and emotional support to the patient and family. However, the integration with the Neurology team was insipient. It was observed that the access to the health services and health technologies were complicated factors. The lack of early diagnosis and rehabilitation also interfered negatively in the process, causing psychological and social suffering to the patient and his family.

Keywords: locked-in syndrome, neurology, long term neurological conditions, palliative care

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Abbreviations: LTNC, long term neurological conditions; LIS, locked-in syndrome; TBI, traumatic brain injury

Introduction

The term Long Term Neurological Condition (LTNC) defines a condition resulting from illness, injury, or damage to the central or peripheral nervous system that affects the person and his family for the rest of their lives. Includes conditions of sudden onset (such as stroke), progressive conditions (such as Motor Neuron Diseases), intermittent conditions (such as epilepsy syndromes), and stable conditions with or without relation to age degeneration (such as cerebral palsy).¹

Locked-In Syndrome (LIS) is one of the conditions that can result from a LTNC. The LIS is characterized by tetraplegia, aphasia and preservation of the level of consciousness, and it is associated with a poor prognosis. The patient with LIS usually depends on certain communication patterns and use the remaining control of eye muscles to answer simple questions with the blink of an eye (yes or no).²⁻⁴ In these situations people experiences a very serious disability and major dependence, which end up leading to a condition of non-communication or an immense difficulty for this. All of that brings a lot of consequences to care planning.

In scientific literature, palliative care for LIS still is a relatively unexplored field and there aren't many articles about it, although in recent years there has been a small increase in publications about it.⁵⁻⁷ In Brazil, the

research about people with LTNC is even fewer, with a small number of relevant research in last years.⁸⁻¹¹ There is no specific mention about very severe incapacities at Brazilian official available data about

disabilities' incidence and prevalence.¹²⁻¹⁴

The palliative approach to these patients isn't easy; misdiagnosis can occur, and the models of prognosis are not accurate enough to define advanced care planning.¹³ In addition, there are complex economic, social and ethical implications.⁴ Clinical examination

should be associated with imaging tests and multimodal neurological evaluations. Symptom control, communication, ethical aspects are everyday challenges faced by the team.⁴ The lack of adequate and early diagnosis interferes negatively in the process of care (and in the prognosis of the patient), causing psychological and social suffering to the patient and his family.⁵ Therefore, from a comprehensive care perspective, the human dignity should be promoted, and this article aims to examine how palliative care could improve LIS patients' quality of life.

Case presentation: The practice to support the theory

A.C.L. is a male patient, 29 years old, married, photographer, with LIS after Traumatic Brain Injury (TBI), assisted at a hospice in São Paulo, Brazil. Detailed clinical history and his biography were collected at his hospital and hospice records and with his parents (which authorized this case report and signed the consent form).

This hospice is a private long-term care institution dedicated to non-oncology palliative care patients, with a total capacity of 70 beds. The service is provided by a multidisciplinary team, composed of doctors, nurses, physiotherapists, occupational therapists, nutritionists, psychologists, social workers, dentists, among others.

Before hospice, A.C.L. remained admitted to a general hospital for 2 years after his traumatic injury. For the first couple of months, he was in a long-term induced coma for a long period, using vasoactive drugs. After clinical stabilization, he was brought back out from induced coma and remained in the ward, receiving basic general care, such as: food by gastrostomy, hygiene, wound prevention, and respiratory physiotherapy. The transfer to the hospice was made after two years of TBI. According to the medical report from this period, presented at the transfer to the hospice, there was no motor or cognitive recovery. At the hospice, A.C.L. is attended by all team.

In addition to adequate symptom control and continuous observation, there were developed strategies to restore meaningful activities in the patient's daily life. The rehabilitation team played a

key role in recovering his life story. The main procedures undertaken were: evaluation of visual perception, motricity and stimuli response; offering sensorial stimuli (touch, smell, sounds, taste and so ever); organization of activities outside hospital bedroom, such as a tour in the garden; prescription of simple assistive technology devices for bed/wheelchair positioning; development of alternative communication.

There were perceived signs of emotion, and he shows flashes of perception and comprehension, with episodes of crying, laughter, obedience to simple commands, attempts to keep an eye on the movement around the room and intense expressive reaction to music.

Simple communication with opening and closing of the eyes was developed and team stated that

A.C.L. has LIS. The care planning included many family conferences, that have been made to offer emotional support and guidance on new findings about A.C.L. level of awareness. All clinical decisions have been made after family meetings.

A.C.L. is still at the hospice because the family cannot provide effective care at home. Recurring infections are the only existing interurrences. He is improving his communication skills and the relatives are being cared by the team as well. Although many attempts were made to compose a coordinate care, there was no return of previous neurology team.

Discussion: The theory to support the practice

The identification of post traumatic LIS is not easy and is subject to misdiagnosis, which could be avoided through the association of continuous clinical examination, imaging tests and multimodal neurophysiological evaluation. In the reported case, the lack of early diagnosis and rehabilitation interfered negatively in the process, causing suffering to the patient and his family. It became evident that there were no previous concern for a careful evaluation and for family comprehensive care.

The need for diagnosis and early neurological prognosis was perceived, and the demand of creation basic communication channel with the patient as soon as possible. In Brazilian context, such situations are even more complex, due to our health system and social and economic situation.¹⁴

The impacts of LTNC (and notably of LIS) are many: cognitive and psychosocial changes, severe physical disabilities, communication problems, etc. The suffering can be pushed to a limit. There are repercussions on the person's identity, in a kind of biographical disruption, meaning that is an experience in which daily life structures are changed and broken; many internal and external resources are needed to face it.¹⁵

In these cases, early decision-making, shared with the family, in which the medical team establishes the rational use of advanced life support, can avoid the disproportionate use of these resources in patients with a poor prognosis, minimizing suffering to patients and their families and directing care to measures of comfort, quality of life and dignity until the end of life.⁴

It is quite common for health professionals to infer that a LIS person quality of life is poor and, therefore, consider their lives worthless. Research with LIS patients report significant levels of self-perceived quality of life and their demand for euthanasia is surprisingly infrequent.³

These data are correlated with the day-to-day observations at the service described in this article. The palliative approach is shown to produce a better quality of life for LIS patients and this case report is an example of it. The care provided by the Palliative Care team also can adequately provide emotional support to the patient and family, as well as symptom control and better communication techniques. There are four fundamental aspects that underlie the assistance for A.C.L. and other LTNC patients at the hospice, as shown at Figure 1: continuous observation, symptom control, propose of meaningful stimuli and family guidance.

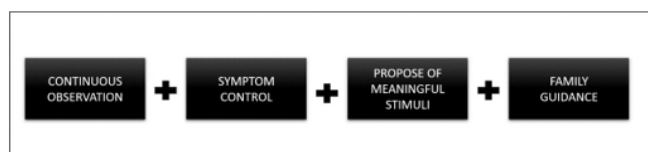


Figure 1 First step to Palliative Care approach

The rehabilitation team (occupational therapy, physiotherapy, speech therapy and psychology) had a major relevance in A.C.L. care planning, providing physical comfort, motor and cognitive stimulation, and alternative communication. Sensory stimuli related to prior patient's occupational repertoire are they are the main strategy developed and a great differential of the care provided in the presented institution.¹⁰ Along with the detailed evaluation, these professionals developed clinical strategies such as continuous monitoring, proposing activities, giving special attention to their reactions, being able to discriminate what pleases or not, and expanding the possibilities of encounter of people with LTNC and the environment which surround them.^{10,16}

Care for the family is also a major role of the hospice team in those situations.^{4,10} Family meetings about the chances of neurological recovery (Uncertainty Principle), the delimitation of advanced life support in poor prognosis and the option for treatment of acute events (infection, hydro electrolytic and metabolic) with a focus on comfort measures, seems to be an option more adequate and well accepted by family members in the clinical management of these patients.¹⁷

One major challenge lies on building an innovative communication to establish the professional-patient relationship, particularly because assistive technology tools (such as alternative communication by computer programs) are not available in Brazilian health care system.

Some lessons can be learned by this case report. It was possible to change the previous focus of care and, even with a poor functional prognosis, A.C.L. and his family perceived quality of life improvements. Although LIS patients' daily lives are very limited, without possibilities of choices and actions, life could not lose its meaning, from an ethical perspective. Hospice care should emphasize memory, identity, and dignity of a person with LIS, since, as proposed by the palliative care principles: life must be meaningful until last moment, providing dignity to patients with LTNC.

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Conflict of Interest

The authors declared that there are no conflicts of interest.

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