Volume 1 • Issue 1 • 2016

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Scientia Ricerca

Research Article

Current Opinions in Neurological Science

ISSN: 2575-5447

Patients with Amyotrophic Lateral Sclerosis in the Intensive Care Unit

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Received: December 13, 2016; **Published:** December 20, 2016

Abstract

Background: Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disorder characterized by progressive muscular paralysis, reflecting degeneration of motor neurones in the primary motor cortex, brainstem and spinal cord. Respiratory function and nutrition are crucial symptomatic concerns for patients with ALS, with respiratory failure being the main cause of death. The aim of this work is to present our experience with patients diagnosed with ALS, hospitalized in intensive care units (ICUs) and to suggest possible solutions for medical, nursing and social problems encountered.

Methods: A retrospective study was carried on 38 patients previously diagnosed with ALS. They were admitted to eight ICUs of public hospitals and private clinics located in the Attica region between 2008 and 2016. The causes of hospitalization, the complications during hospitalization, the daily nursing care applied and the outcome were analyzed.

Results: Before admission in the ICU, all patients used non-invasive mechanical ventilation. Main causes of hospitalization of patients with ALS was respiratory failure resulting in hypercapnia (26.3%), infection of the respiratory system (73.6%) or hypoxemia (7.8%). During their ICU stay, all patients were mechanically ventilated, developed complications during hospitalization and received daily nursing care and enteral tube feeding (LEVIN). Tracheostomy and jejunostomy was performed in 100% of cases. Mortality rate was 76.3%. Main causes of death were ventilator associated pneumonia (VAP) (71.4%), heart failure (19%) and septic shock (9.6%). The average cost per day of hospitalization was approximately 1500 euro. Patients discharged from the ICUs required continuation of mechanical ventilation first in the hospital ward and then at home.

Conclusions: Long term mechanical ventilation, even though it predisposes to serious complications, should be applied outside hospital. Malnutrition is one of the most serious problems of patients with ALS. While hospitalized, treatment cost for patients with ALS is very high. Thus, treatment at home of patients in late stages of ALS -covered by the state- reduces hospitalization costs, emotional load of patients and relatives, improve their quality of life and prolong survival time due to avoidance of constant hospital infections. More so, for a close and proper monitoring of patients with mechanical ventilation at home, a national center should immediately be established.

Keywords: *Amyotrophic Lateral Sclerosis; Mechanical Ventilation; Non-invasive Mechanical Ventilation; Tracheostomy; Septic Shock*

Abbreviations: Amyotrophic Lateral Sclerosis: ALS; Intensive Care Unit: ICU; Ventilator Associated Pneumonia: VAP

Volume 1 Issue 1 December 2016

Introduction

Amyotrophic lateral sclerosis can be defined as a neurodegenerative disorder characterized by progressive muscular paralysis reflecting degeneration of motor neurones in the primary motor cortex, brainstem and spinal cord. It is one of the most common neurological diseases following multiple sclerosis but its causes are hardly known. Advances in the understanding of the glutamate neurotransmitter system and the discovery of causal genes linked to the development of ALS have stimulated research interest while survival in ALS is understood to be dependent on several factors, including clinical presentation (phenotype), rate of disease progression, early presence of respiratory failure, and the nutritional status of patients [1]. However, attempts to establish the complex genetic basis for sporadic ALS by identifying susceptibility genes have had little success.

Respiratory function and nutrition are crucial symptomatic concerns for patients with ALS, with respiratory failure being the main cause of death. Thus, expert consensus guideline recommendations have been developed for key care concerns in ALS, including respiratory management, nutrition, and palliative care.

Respiratory compromise is commonly present at diagnosis in patients with ALS. Nocturnal hypoxia, and associated symptoms of lethargy, loss of concentration, morning headaches, and unrefreshed sleep are consequences of central dysfunction. Diaphragmatic weakness can be diagnosed with spirometry, with vital capacity undergoing a progressive decline over the course of disease [2]. The damage of the central and peripheral motor neurons directly affects the functioning of the respiratory system and causes most pulmonary problems which are expressed as respiratory failure. This evolution requires immediate or long-term treatment in intensive care units [3]. The decision to ventilate a patient with ALS is complex, given the rapid evolution of the disease in most cases, as he may become completely ventilator dependent [4].

Patients' treatment in the ICUs has significantly improved in recent years both in medical and nursing terms. Therefore, patients' hospitalization time is significantly shortened while survival time is considerably prolonged.

The aim of our work is to present our experience with patients diagnosed with ALS and hospitalized in the ICU at late stages of disease and to suggest possible solutions for medical, nursing and social problems encountered.

Materials and Methods

We retrospectively analyzed the medical files of 38 patients (30 male and 8 female) admitted to eight ICUs of public hospitals and private clinics located in the Attica region during a 9-year period, with ALS in late stages. All patients had been diagnosed with ALS in special neurological clinics before arrival. In analyzing our experience, we took into consideration the causes of hospitalization, complications during hospitalization, daily nursing care applied and the outcome.

Results and Discussion

Thirty-eight patients with ALS in late stages (30M + 8F), aged 40-88 years were hospitalized in the last 9 years (2008-2016) in our ICUs (Table 1).

Year	Number of patients	Mean duration of hospitalization (days)	Deaths
2008	2	85	3
2009	1	90	2
2010	2	47	2
2011	1	349	2
2012	2	42	2
2013	3	55	4
2014	5	169	-
2015	7	120	7
2016	15	31	7
Total	38	26	29

Table 1: *Patients' distribution.*

The mean duration of hospitalization in the ICU was 26 days. Patients were diagnosed with ALS in special neurological clinics 2-5 years and all of them used non-invasive mechanical ventilation before arrival in the ICUs.

The reason of hospitalization for 10 patients (26.3%) was respiratory failure resulting in hypercapnia, for 28 patients (73.6%) was infection of the respiratory system, for 3 patients (7.8%) was hypoxemia while for 3 patients, other causes led to their admission in the ICU (Table 2).

A/A	Cause of hospitalization	No. of patients
1	Failure resulting in hypercapnia	10
2	Infection of the respiratory system	28
3	Hypoxemia	3
4	Other causes	3

Table 2: *Causes of hospitalization.*

Once admitted to our ICUs, all patients (100%) were mechanically ventilated and all developed complications during hospitalization: hospital-acquired pneumonia (HAP) 38 patients (100%), pneumothorax 8 patients (21%), bedsores 9 patients (23.6%), GI bleeding/gastrorrhagia 16 patients (42.1 %), thrombophlebitis 8 patients (21%), sepsis 8 patients (21%), myocardial infraction 3 patients (7.8%), cardiomyopathy 5 patients (13.2%), micturition disorders 17 patients (44.7 %), acute pancreatitis 3 patients (7.8%), paralytic ileus 3 patients (7.8%) (Table 3).

A/A	Complication	No. of patients
1	Hospital-acquired pneumonia (HAP)	34 patients
2	Pneumothorax	8 patients
3	Bedsores	9 patients
4	GI bleeding/gastrorrhagia	16 patients
5	Thrombophlebitis	8 patients
6	Sepsis	8 patients
7	Myocardial infraction	3 patients
8	Cardiomyopathy	5 patients
9	Micturition disorders	7 patients
10	Acute pancreatitis	3 patients
11	Paralytic ielus	3 patients

Table 3: *Complications during hospitalization.*

Mortality rate was 76.3%. Main causes of death were VAP (71.4%), heart failure (19%), acute pancreatitis and paralytic ileus possibly due to bacteria translocation, which led to septic shock (9.6%) (Table 4).

A/A	Cause of death	No. of patients
1	Ventilator-Associated Pneumonia	19 patients
2	Heart failure	6 patients
3	Septic shock	4 patients

Table 4: *Causes of death.*

All patients received daily nursing care (Table 5) and enteral tube feeding (LEVIN) during their ICU stay while, at dismissal, underwent jejunostomy.

A/A	Type of care
1	Morning patient care
2	Bed linen change
3	Bath
4	Prevention and treatment of pressure sores
5	Wash the oral cavity
6	Eye Care
7	Change wounds
8	Routine change of central venous and arterial catheters
9	Change endotracheal tube position
10	Change tracheostomy pipes
11	Change the supporting (neck tape) and care of the area
12	Maintain aseptic rules
13	Monitoring vital signs
14	Inpatient treatment care
15	Monitor fluid balance

16	Change of patient positioning
17	Weight control, diet and stool
18	Monitoring the ventilator operation
19	Check drains operation
20	Humidification
21	Suctioning
22	Physiotherapy and Locomotor Therapy

Table 5: *Daily nursing care of patients with ALS in ICUs.*

The average cost per day of hospitalization in the ICU was approximately 1500 euros (without calculating the cost of equipment maintenance) (Table 6). Those patients who were discharged from the ICUs required continuation of mechanical ventilation first in the hospital ward and then at home.

Type of expense	Amount (euro)
Staff cost (doctors, nurses, support staff)	630
Drugs, Fluids, Nutrition	550
Consumables	220
Laboratory Tests	100
Total cost	1.500

** Personalization of costs couldn't be accurately estimated due to lack of uniform protocols.*

Table 6: *ICU average cost per patient per day.*

Handling patients with serious diseases is a complex problem for health professionals and relatives both in terms of everyday care provision and emotional load. Especially in patients with ALS, whose mental function allows them to be aware of their state of health, the problem becomes more severe and requires a special approach [5]. Best care for patients with ALS is provided within a multidisciplinary environment where physiotherapists, occupational therapists, speech therapists, respiratory physicians, gastroenterologists, and social workers collaborate to guide symptomatic management through the course of disease [6].

ALS is an idiopathic fatal disease, but, fortunately, there is extensive research underway on the pathogenesis of ALS and there might be a cure in the future [7]. ALS is characterized by muscle weakness, muscle atrophy, swallowing and speech disorders and ultimately respiratory failure. Thus, until a cure is found for ALS, respiratory care providers have a crucial role in evaluating, educating, and treating patients.

Respiratory interventions such as non-invasive ventilation improve both quality of life in patients with ALS and their survival [8]. Guidelines for instituting non-invasive ventilation rely on a combination of symptoms that signify respiratory muscle weakness (dyspnoea and orthopnoea), along with signs of respiratory muscles weakness, including substantial desaturation on overnight oximetry, increased partial pressure of carbon dioxide (PCO_2) of less than 65 mm Hg and reduced forced vital capacity of less than 80% or sniff nasal inspiratory pressure of less than 40 cm H_2O [3,9]. Patients with substantial bulbar impairment and sialorrhoea might not tolerate non-invasive ventilation, and appropriate management of secretions is crucial [3]. In patients with ALS who are intolerant of non-invasive ventilation, when this form of ventilation is no longer sufficient because of progressive respiratory muscle weakness, invasive ventilation via tracheostomy seems to be the only option [9].

Respiratory failure accompanied by reduction of voluntary control over emotional reactions with paroxysms of laugh and cry but which do not correspond to the emotional state of the patient, create a vicious cycle of emotional reactions of both patients and their

environment. More so, research reveals that avoiding isolation as well as changing patients' psychological and social environment have contributed to the prolongation of patients' survival. At this stage of disease, psychological support of the patient and of his/her environment is necessary not only throughout the duration of hospitalization, but also after hospital release, while at home.

For patients diagnosed with ALS, the state insurance should assure special home-care provided by experienced medical and nursing personnel as well as medication, besides the basic treatment for the disease (Rilutek tabl. 1 x 2 daily, vit. B12 amp/15 days and Eviol tabl. 1 x 3 daily). Physiotherapy should also be provided through the insurance system. Thus, in order to have a close and proper monitoring of patients with home mechanical ventilation, as it is the case of our patients, a national center should immediately be established [10,11]. However, our experience reveals that economic crisis affecting Greece in the last years has had a negative impact on the health and quality of care of people with disability, while raising considerably the unmet needs for care. Precisely, reduction of healthcare expenditure, increases in medicines co-payments and reduction of disability pensions/benefits negatively affected the possibility to provide the necessary care for people with ALS [12,13]. Thus, the ability to provide minimum necessary therapy and care for patients discharged from the ICU depends on the financial status of the patients and of his/her relatives and is mainly provided through private medical and nursing support.

Conclusion

From our experience with patients suffering from ALS and hospitalized in our ICUs, several important issues arose. *First*, the overall hospitalization period of patients with ALS is very large. *Second*, while hospitalized, treatment cost for patients with ALS is very high. Expenses for technological equipment and nursing care at home covered by the state reduce hospitalization costs, emotional load of patients and relatives, improve their quality of life and prolong survival time due to avoidance of constant hospital infections. *Third*, long term mechanical ventilation predisposes to serious complications. However, it is necessary to ensure life continuation of patients with ALS and thus, it should be applied outside hospital. On the other hand, it also prevents complications that may occur due to long term hospitalization (especially hospital-acquired pneumonia). *Fourth*, only 25% of our patients survived and, after discharge, they required advanced palliative care at home. *Fifth*, malnutrition is one of the most serious problems of patients with ALS. Emphasis should be given to patients' feeding due to their inability of swallowing and tracheostomy that predispose to the occurrence of aspiration pneumonia. Frequent monitoring from doctors, social workers, nurses and physiotherapists is essential. *Last*, for a close and proper monitoring of patients with mechanical ventilation at home, a national center should immediately be established.

Conflict of interest

The authors declare that they have no competing interests.

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