Arch Bronconeumol. 2014;**50(12)**:509–513

ARCHIVOS DE BRONCONEUMOLOGIA

www.archbronconeumol.org

Original Article

Amyotrophic Lateral Sclerosis: Impact of Pulmonary Follow-Up and Mechanical Ventilation on Survival. A Study of 114 Cases☆

Pilar Sanjuán-López, Paz Valiño-López, Jorge Ricoy-Gabaldón, Héctor Verea-Hernando*

Servicio de Neumología, Complexo Hospitalario Universitario A Coruña, CHUAC, A Coruña, Spain

ARTICLE INFO

Article history:
Received 15 January 2014
Accepted 7 April 2014
Available online 15 November 2014

Keywords:
Amyotrophic lateral sclerosis
Respiratory management
Ventilatory failure

ABSTRACT

Objective: To study the impact of ventilatory management and treatment on the survival of patients with amyotrophic lateral sclerosis (ALS).
Method: Retrospective analysis of 114 consecutive patients admitted to a general hospital, evaluating demographic data, type of presentation, clinical management, treatment with mechanical ventilation and survival. Statistics: descriptive and Kaplan–Meier estimator.
Results: Sixty-four patients presented initial bulbar involvement. Overall mean survival after diagnosis was 28.0 months (95% CI, 21.1–34.8). Seventy patients were referred to the pulmonary specialist (61.4%) and 43 received non-invasive ventilation (NIV) at 12.7 months (median) after diagnosis. Thirty-seven patients continued to receive NIV with no subsequent invasive ventilation. The mean survival of these patients was 23.3 months (95% CI, 16.7–28.8), higher in those without bulbar involvement, although below the range of significance. Survival in the 26 patients receiving programmed NIV was higher than in the 11 patients in whom this was indicated without prior pulmonary assessment (considered following diagnosis, $P<.012$, and in accordance with the start of ventilation, $P<.004$). A total of 7 patients were treated invasively; mean survival in this group was 72 months (95% CI, 14.36–129.6), median 49.6±17.5 (95% CI, 15.3–83.8), and despite the difficulties involved in home care, acceptance and tolerance was acceptable.
Conclusions: Long-term mechanical ventilation prolongs survival in ALS. Programmed pulmonary assessment has a positive impact on survival of ALS patients and is key to the multidisciplinary management of this disease.

Esclerosis lateral amiotrófica: impacto del seguimiento neumológico y ventilación mecánica en la supervivencia. Experiencia en 114 casos

Palabras clave:
Esclerosis lateral amiotrófica
Manejo respiratorio
Insuficiencia respiratoria

RESUMEN

Objetivo: Conocer el impacto en la supervivencia del manejo y tratamiento ventilatorio de enfermos con esclerosis lateral amiotrófica (ELA).
Método: Análisis retrospectivo de 114 pacientes con ingreso consecutivo en un hospital general, evaluando datos demográficos, tipo de presentación, manejo clínico, tratamiento con ventilación mecánica y supervivencia. Estadística: descriptiva y análisis de Kaplan-Meier.
Resultados: Sesenta y cuatro pacientes tenían afectación bulbar inicial. La supervivencia media global tras el diagnóstico fue 28,0 meses (IC 95%, 21,1-34,8). Setenta pacientes fueron derivados al neumólogo (61,4%) y 43 recibieron ventilación no invasiva (VMNI) a los 12,7 meses (mediana) del diagnóstico. Se mantuvieron con VMNI sin posterior ventilación invasiva 37 pacientes, cuya supervivencia media fue de 23,3 meses (IC 95%, 16,7-28,8), superior en los no bulbares, aunque en rango no significativo. En 26 en los que la VMNI se indicó de manera programada la supervivencia fue mayor que en 11 en que se indicó sin evaluación neumológica previa (considerando tras el diagnóstico, p < 0,012, y en función del comienzo

☆ Please cite this article as: Sanjuán-López P, Valiño-López P, Ricoy-Gabaldón J, Verea-Hernando H. Esclerosis lateral amiotrófica: impacto del seguimiento neumológico y ventilación mecánica en la supervivencia. Experiencia en 114 casos. Arch Bronconeumol. 2014;50:509–513.

* Corresponding author.
E-mail address: hector.verea.hernando@sergas.es (H. Verea-Hernando).

http://dx.doi.org/10.1016/j.arbr.2014.10.008
1579-2129/

de la ventilación, p < 0,004). Se trataron en modalidad invasiva 7 pacientes cuya supervivencia fue de 72 meses (IC 95%, 14,36-129,6), mediana de 49,6 ± 17,5 (IC 95%, 15,3-83,8), y pese a las dificultades de la atención en domicilio, la aceptación y la tolerancia fueron aceptables.
Conclusiones: La ventilación mecánica prolonga la supervivencia de la ELA. La evaluación neumológica programada tiene un impacto favorable en la supervivencia de los pacientes con ELA y constituye un elemento esencial en el manejo multidisciplinario de esta enfermedad.

Introduction

Amyotrophic lateral sclerosis (ALS) is characterized by muscle weakness and progressive paralysis, and is the most common form of motor neuron disease. Incidence is estimated to be around 1–2 cases per 100 000 inhabitants/year: 5%–10% of cases are the familial form, and the majority is the sporadic.[1,2] It is caused by the progressive loss of upper motor neurons in the corticospinal tract, bulbar muscles and the spinal cord, leading to weakness and paralysis of the muscles of the upper and lower limbs, face and trunk, including the diaphragm.[3] Presentation is heterogeneous, with variable involvement of the muscle groups and differences in prognosis and quality of life, probably reflecting the different mechanisms causing the disease.[4] Bulbar and respiratory muscle involvement is characterized by dysarthria, choking, ineffective cough, dyspnea, orthopnea and progressive respiratory failure, generally leading to death within 3–5 years after disease onset.[1,5]

Treatment options are very limited. One available drug is riluzole, but its action is limited and weak,[6] and the only realistic therapeutic measures are palliative. One of the best of these is non-invasive ventilation (NIV). NIV can be used at home, the equipment is user-friendly and controls respiratory failure, making it very well tolerated and accepted by patients and caregivers. As the disease advances, and when bulbar involvement is significant, NIV becomes insufficient and invasive ventilation via tracheotomy must be considered.[7]

In addition to respiratory failure, patients develop significant changes in their ability to move and communicate, along with severe nutritional disorders,[8] giving rise to delicate ethical and social issues regarding intervention and its clinical implications.[9] This is a complex situation requiring the coordination of different teams of health workers and caregivers to provide the pulmonary care necessary for the treatment of respiratory failure.[10] With aim of defining the role of the pulmonology specialist in the multidisciplinary management of ALS patients, we have reviewed the utility of programmed respiratory care appointments and the impact of mechanical ventilation on the survival of patients attending a general hospital with no dedicated ALS unit.

Materials and Methods

Patient Selection and Treatment

We performed a retrospective analysis of patients with a diagnosis of ALS consecutively hospitalized in our center between January 1, 2000 and December 31, 2010. This is a third-level general hospital with a catchment area of 550 000 inhabitants. Standard care of patients with suspected ALS in this setting did not include routine referral to a pulmonologist for evaluation of ventilatory requirements, and patients with a diagnosis of ALS (CIE-9:335.20) had to be identified from the discharge reports in the hospital database. Patients with a definitive or probable diagnosis of ALS determined by a neurologist according to the revised El Escorial criteria[3] were included. The initial list of 180 patients supplied by the hospital discharge encoders was examined and 66 patients were excluded either because they had neuromuscular processes other than ALS or because they were treated in social welfare palliative care centers not associated with our hospital. Cases with ALS seen only in outpatient clinics and never hospitalized were also excluded. Patients that, at the beginning or during the course of their disease, had obvious difficulty with phonation or swallowing, confirmed by both a neurologist and a pulmonologist, were classified as having bulbar ALS.

After respiratory assessment by the treating pulmonologist, all patients with respiratory failure, according to established criteria,[11] were offered a mechanical ventilation device. VS ultra and VS III Resmed ventilators were used in spontaneous timed (ST) and pressure support (PSV) mode with a nasal or oronasal mask. Ventilation began with the patient hospitalized or in the outpatient clinic, starting with low pressures until symptoms were relieved. Oximetry and blood gases were monitored during waking hours and oximetry during sleep. Hospitalized patients were seen by the physiotherapist attached to the hospitalization unit. In the subsequent follow-up, ambulatory oximetry monitoring was used during sleep, parameters were corrected and possible problems due to mask adjustment were checked. Invasive ventilation via tracheostomy (VS II/VS III Resmed ventilator in assist control [ACV] mode and EOLE 3 volumetric respirator) was indicated when patients presented refractory dyspnea, respiratory failure that could not be controlled with NIV, recurrent aspiration, or inability to manage secretions. If expectoration problems and a cough peak flow lower than 270 l/min were observed despite assisted cough physiotherapy, a mechanical cough assistant was indicated (Cough Assist, insufflator–exsufflator, MI-E, Emerson) and caregivers were trained in its use. Except in emergency situations, the ventilatory mode was established after conferring with the patient and caregivers. Tolerance was optimized in training sessions. Patients were monitored in face-to-face visits and telephone calls, and were seen at home by a nurse from the equipment supply company. Outpatient physiotherapy was not provided on a regular basis. Date of death was obtained from the death certificate or hospital records, but the immediate cause of death was not considered a study parameter.

Follow-Up

The date limit for follow-up of patients included in the study was May 1, 2011.

Variables Analyzed

Data were collected on demographics (age, sex, date of birth and death), clinical features (date of ALS diagnosis, area of bulbar or corticospinal onset and start dates of NIV, invasive ventilation and indication for mechanical cough assistance), pulmonary function at the start of mechanical ventilation (forced vital capacity) and percentage of time with oxygen saturation less than 90% in nocturnal oximetry readings (CT90).

Statistical Analysis

A descriptive analysis was performed of the study parameters with qualitative variables expressed as percentages and