Mechanical ventilation in patients with amyotrophic lateral sclerosis: literature review and reflection

Ventilação mecânica em pacientes com esclerose lateral amiotrófica: revisão de literatura e reflexão

Ventilación mecánica en pacientes con esclerosis lateral amiotrófica: revisión de la literatura y reflexión

Luiz Alberto da Silva Santos Junior[1], Débora Dadiani Dantas Cangussu[2], Camilla Alves Santana[3], Ana Luiza Ferreira Donatti[3]

How to cite: Santos Júnior LAS, Cangussu DDD, Santana CA, Donatti ALF. Mechanical ventilation in patients with amyotrophic lateral sclerosis: literature review and reflection. REVISA. 2020; 9(2): 327-43. Doi: https://doi.org/10.36239/revisa.v9.n2.p327a343

REVISA

1. Centro Universitário Estácio Brasilia, Federal District, Brazil.

2. Centro Universitário Estácio Brasilia, Federal District, Brazil.

3. Centro Universitário Estácio Brasilia, Federal District, Brazil.

Received: 15/01/2019
Accepted: 18/03/2020

RESUMO

Objetivo: avaliar a importância da fisioterapia respiratória por meio da ventilação mecânica (invasiva ou não invasiva) dada a aplicabilidade e importância dessa prática no trato com a esclerose lateral amiotrófica. **Método:** foi elaborada uma revisão da literatura através de pesquisa bibliográfica em diversas bases de dados que versam sobre o tema. **Resultados:** Na fisioterapia respiratória não-invasiva, se usam ventiladores e máscara sem necessidade de intervenção cirúrgica, já a ventilação mecânica invasiva consiste principalmente na realização da traqueostomia**. Conclusão:** não há uma regra efetiva sobre qual o tipo de ventilação mecânica é mais adequada na lida de pacientes com esclerose lateral amiotrófica. No entanto, pôde se observar que se aplica a ventilação mecânica não invasiva geralmente no estágio inicial da doença e a partir do momento em que o paciente perde a autonomia respiratória passa-se a utilizar a ventilação mecânica invasiva. É consenso também que cabe ao paciente e à família, devidamente esclarecidos, optar ou não pela ventilação mecânica, e por uma ou outra modalidade.

Descritores: Ventilação Mecânica; Fisioterapia Respiratória; Esclerose Lateral Amiotrófica.

ABSTRACT

Objective: to evaluate the importance of respiratory physiotherapy through mechanical ventilation (invasive or non-invasive), given the applicability and importance of this practice without treatment with amyotrophic lateral sclerosis**. Method:** a literature review was carried out through bibliographic research in several databases dealing with the topic. **Results:** In non-invasive respiratory physiotherapy, ventilators and a mask are used without the need for surgical intervention, whereas invasive mechanical ventilation consists mainly of performing a tracheostomy. **Conclusion:** there is no effective rule on which type of mechanical ventilation is most appropriate in dealing with patients with amyotrophic lateral sclerosis. However, it was observed that non-invasive mechanical ventilation is generally applied in the initial stage of the disease and from the moment the patient loses respiratory autonomy, invasive mechanical ventilation is used. There is also a consensus that it is up to the patient and family, duly clarified, to choose mechanical ventilation or not, and one or the other modality.

Descriptors: Mechanical Ventilation; Respiratory Fisioterapy; Amyotrophic Lateral Sclerosis.

RESUMEN

Objetivo: evaluar la importancia de la fisioterapia respiratoria a través de la ventilación mecánica (invasiva o no invasiva) dada la aplicabilidad y la importancia de esta práctica en el tratamiento de la esclerosis lateral amiotrófica. **Método:** Tres muestras de yugo de caña de azúcar de establecimientos distribuidos en diferentes regiones del Distrito Federal. Todos los experimentos se llevaron a cabo utilizando el método de sedimentación y se analizaron por triplicado. El análisis de los resultados se realizó con la ayuda de microscopía óptica, para investigar la presencia de humanos, quistes, ooquistes y otras formas evolutivas parasitarias**. Resultados:** En todos los casos, existe la presencia de algún tipo de contaminación, con levaduras registradas en el 100% de los casos, en 50% Entamoeba sp., 10% Taenia sp., 10% Giardia sp. y en 10% Ascaris sp. **Conclusión**: es posible observar una ineficiencia en las prácticas higiénico-sanitarias de los establecimientos responsables del procesamiento y del molino de vegetales, lo que permite la contaminación en la mayoría de las mujeres. Esta contaminación puede causar diferentes tipos de enfermedades en humanos, particularmente ofreciendo más riesgo a las personas inmunocomprometidas, lo que requiere un monitoreo más efectivo de este tipo de alimentos, así como su distribución.

Descriptores: Ventilación mecánica; Fisioterapia respiratoria; Esclerosis lateral amiotrófica

REVIEW

Introduction

Amyotrophic lateral sclerosis (ALS), also called Lou Gehrig's disease, or Charcot's disease, is characterized by neuron-motor degeneration, of the motor cortex, brain stem and spinal cord, which leads to progressive muscle paralysis and functional dysfunction; affects upper and lower motor neurons and affects about 0.02% of the world population.[1-3]

The term Amyotrophic Lateral Sclerosis, secondary muscle weakness and impairment of motor neurons, can be interpreted as follows: Sclerosis (hardening; scarring); Lateral Sclerosis (stiffening of the lateral portion of the spinal cord due to the death of the upper motor neurons); Amyotrophic [A (not) Myo (muscle), Atrophy (decreased, weakened)]. And the causes of this pathology can be: genetic mutation - hereditary; chemical imbalance - excess glutamate (harmful to some nerve cells); autoimmune disease - some healthy cells in the body are mistakenly affected by the individual's immune system (in this case, neurons); misused proteins - inside the nerve cells protein deviation occurs, which can cause an accumulation of abnormal forms of proteins in the cells which triggers the death of nerve cells.[4]

ALS predominantly affects individuals between 40 and 60 years of age, characterized by the dominant involvement of the skeletal motor system, manifested by muscle atrophy and weakness, spasticity, anarthria (difficulty in articulating words) and respiratory problems that culminate in serious outcomes.[5] In a general overview about ALS, the average age of onset of this pathology is between 43 and 52 years old, in cases of familial ALS and, between 58 and 63 years old, in sporadic cases; that the individual risk of developing ALS over a lifetime is from 1 to 350 to 500, and is generally more frequent, 1.2 to 1.5 times, in males than in females.[2] There is a potential genetic component (especially with pathogenic genetic variants of the C9orf72, TARDBP, SOD1 or FUS genes), but the etiology and pathogenesis are unclear and ALS is probably due to several genetic and environmental factors.[6] This pathology presents a characteristic of progressive loss of strength, in the middle stage of evolution, affects the respiratory muscles[4,6]:

- Family (autosomal dominant inheritance): in patients with a family history of ALS or frontotemporal dementia (which may be associated with ALS), reported in about 5% to 20% of patients, it starts ten to fifteen years earlier than that in the case of sporadic ALS, or may have a juvenile onset; approximately 10% of patients with the adult form of familial ALS suffer from a mutation in the copper / zinc superoxide desmutase (SOD1) enzyme gene on chromosome 21. Normally, SOD performs the dismutation of the free radical superoxide ion (O2) in hydrogen peroxide (H2O2) and molecular oxygen (O2). It is believed that the decrease in SOD activity leads to the accumulation of the superoxide ion, which then binds to nitric oxide (NO) to form the free radicals peroxidonitrile (ONOO-) and hydroxyl (OH-). The accumulation of free radicals is highly damaging to the cell, as it causes peroxidation of the membranes and modification of proteins, culminating in the death of the motoneuron. More than 100 mutations have already been cataloged in SOD1, practically all inherited in the form of an autosomal dominant inheritance. The substitution of valine for alanine in position 4 (A4V) is the most common form of SOD1 mutation. Other mutations have been described in other genes: 9q34; 9q21-22; 2q33; 15q15-22.

• ALS with mutation in the VAPB gene (ALS type 8): Described in Brazil, it is clinically characterized by the onset of symptoms between 25-40 years, with variable progression. Mutation in the vesicle gene associated with the membrane of protein associated with protein B (VAPB), mapped in the 20q 13.31 region, has been implicated as a causal factor.
• West Pacific ALS: affects inhabitants of Guam Island, has a prevalence 50 to 100 times higher than in the rest of the world; possibly associated there is a neurotoxin present in food sources of the inhabitants.
• Sporadic (classic form): reported in about 80% to 95% of patients, the spread and amplification of the initial lesion occurs when the death of a motoneuron releases large amounts of nitric oxide, free radicals, glutamate, calcium and free, harmful metals to neighboring cells. The selectivity of the lesion, exclusive to the motor neuron in ALS / DNM (Motor Neuron Degeneration), is explained by the unique characteristics of this cell, such as the large size of the pericardium, the richness of dendrites and axons, the great extension of the axons, the importance of the cytoskeleton, the presence of receptors that allow selective calcium entry, the presence of receptors for androgens, the intracellular expression of choline acetyl transferase, the low concentration of calcium buffering proteins and the low affinity of receptors for the factors of neuronal growth.
• Classical - ALS spinal or bulbar. It may be the beginning of the spine: reported in about 46% of patients, initial degeneration of the motor neuron in the spinal cord. The initial presentation may involve localized symptoms and signs attributable mainly to the loss of upper motor neurons or the loss of lower motor neurons; or bulbar onset: reported in about 23% of patients, initial degeneration of the motor neuron in the brain stem.
• Beginning in the spine or bulbar with concomitant frontotemporal dementia: reported in about 5% to 15% of patients.
• Bulbar isolated: reported in about 5% of patients Characterized by isolated pseudobulbar or bulbar paralysis for years, more common in women with symptoms of dysarthria and emotional lability.
• Progressive Restricted Spinal Muscular Atrophy (lower motor involvement only) or Primary Lateral Sclerosis (upper motor neuron involvement only): reported in about 10% of patients.

ALS affects two types of motor neurons: upper motor neurons (NMS), or first neuron (Betz cells); lower motor neurons (NMI), or second neuron, which are located in the brain stem and in the anterior portion of the spinal cord. The NMS regulate the activity of the NMI, by sending chemical messages (neurotransmitters). The activation of the NMI allows the contraction of the voluntary muscles of the body. The brainstem NMI activates muscles of the face, mouth, throat and tongue. NMI in the spinal cord activates all other voluntary muscles in the body, such as those of the limbs (upper and lower), trunk, neck, as well as the diaphragm. Most patients have spinal-onset ALS (with initial degeneration of motor neurons in the spinal cord) or bulbar-onset ALS (with initial motor neuron degeneration in the brain stem, associated with dysarthria and dysphagia).[4,6]

The disease evolves with the predominant involvement of the musculature of the limbs (upper rather than lower); bulbar involvement, usually asymmetric; installation of symptoms; and lastly, vocal and respiratory functions are affected. Symptoms and signs of amyotrophic lateral sclerosis (ALS) are due to loss of

neurons and denervation of target cells. With the loss of upper motor neurons (the loss of motor neurons in the motor cortex), degeneration of the corticospinal axon occurs, which manifests itself as thinning and scarring (sclerosis) of the corticospinal tract. In a next stage, the loss of lower motor neurons (loss of motor neurons in the brain stem and spinal cord), leads to thinning of the ventral roots of the spinal cord and atrophy; denervation (amyotrophy) of the muscles of the limbs, oropharynx and tongue; loss of neurons in the frontal and temporal lobe; to symptoms of frontotemporal dementia. In an intermediate phase, there may be loss of neurons that innervate the eye muscles and bladder in ALS. In an advanced stage, the loss of neurons is accompanied by inflammatory processes (recruitment of astrocytes, microglia and oligodendroglia) that can cause additional damage.[4,6]

In the diagnosis of ALS, when performing physical activities, look for signs of respiratory impairment; abnormal reflexes and other signs of loss of motor neurons. The main signs and symptoms of ALS are: progressive weakness, muscle atrophy, spontaneous muscle contractions (fasciculations), muscle cramps, spasticity, dysphagia, dyspnea, difficulty in swallowing, emotional lability, difficulty in breathing, gagging, drooling, stuttering (dysphemia) , drooping head (due to weakness in the neck muscles), difficulty raising objects, difficulty climbing stairs and difficulty walking, paralysis, diction problems (such as a slow or abnormal speech pattern), voice changes - hoarseness, jaw exaggerated and weight loss. [4,6,8]

When talking about the diagnosis of ALS, it is stated that it is based on symptoms, physical examination findings, results of electroneuromyography (ENMG) and other imaging (tomography) and laboratory tests. Thus, it is a clinical, neurophysiological and exclusion diagnosis.[2]

The evolution of ALS is progressive and death occurs due to respiratory failure, aspiration pneumonia or pulmonary embolism, due to prolonged immobility.[3] The average survival time is said to be three to five years.1 However, in exceptional cases patients die in the first year of the disease or survive for more than twenty-five years.[3]

Bed confinement occurs between two and four years after the first signs, usually manifested by limb paralysis (hands, usually asymmetrically), muscle cramps and denervation; with worsening the paralysis extends to other areas of the body, gradually increasing the individual's Respiratory activity is usually achieved late, but can be affected in the initial stage of the disease..[3] And as the disease worsens, with increased dependence, the patient's functional activities are compromised. It is a consensus that the approach and treatment of ALS must be multidisciplinary. In this context, the respiratory physiotherapist's intervention plan should provide for activities that improve the individual's quality of life and prolong life span.[7]

Few considerations regarding the respiratory physiotherapist's performance: the physiotherapist plays a fundamental role in preparation and rehabilitation; respiratory physiotherapy seeks to control the symptoms of respiratory disorders by means of aerobic exercises; improves physical fitness and cardiovascular function; uses techniques capable of improving respiratory mechanics, vital capacity, pulmonary compliance, tidal volume, promoting pulmonary reexpansion and bronchial hygiene. The methods include: exercises of ventilatory patterns-PV's (exercises that aim to increase the tidal volume,

improve the inspiratory capacity and the effectiveness of the cough, which can be performed through sustained or fractional deep inhalation followed by exhalation, associated or not with movements of the upper limbs) with encouragement for deep breathing, use of incentive spirometer and bronchial hygiene maneuvers with cough stimulus (they are techniques that favor the removal of pulmonary secretions, such as manual chest percussions, vibrations and / or vibrocompressions, cough stimulus, postural drainage, tracheal and airway aspiration), pulmonary reexpansion maneuvers (aim to expand lung volume by increasing the gradient of transpulmonary pressure and reducing pleural pressure), and respiratory incentives (act with the increase in lung volume, promoting visual feedback and, in this way, stimulating the patient to perform the re training spiratory).[9-18]

Thus, respiratory physiotherapy must proceed with the assessment, treatment and management of patients with ALS. The professional in this area must decide, for example, among the existing techniques for performing respiratory physiotherapy in patients with ALS, which ventilation method to use, between the use of invasive and non-invasive ventilation methods, according to the patient anamnesis.[19]

Mechanical ventilation aims to provide respiratory support to the patient until the underlying problem is resolved, or to maintain support for the patient with chronic ventilatory problems.[20]

Invasive mechanical ventilation (IMV) - through a prosthesis introduced into the airway (intubation) or tracheostomy - can save the lives of patients who are unable to maintain a satisfactory breathing pattern. The main objectives of VMI: to reduce overwork in the respiratory muscles, improve gas exchange and optimize respiratory function.[21]

In this context, regarding non-invasive respiratory ventilation supports, it is stated that this type of ventilation avoids complications and difficulties associated with invasive mechanical ventilation. Thus, it prevents acute pulmonary morbidity, hospitalization and intubation, enables oral feeding and speech, reduces the risk of respiratory infection, and even helps to mobilize secretions. However, the author warns that in cases where the disease presents at a late stage, invasive mechanical ventilation should be maintained. That said, noninvasive mechanical ventilation should be maintained as much as possible in order to increase the patient's survival time, delaying the use of invasive ventilation mechanisms.[22]

Thus, addressing the issue of respiratory physiotherapy, mechanical ventilation and its applications in patients with Amyotrophic Lateral Sclerosis is of great relevance in helping students, physiotherapists and other health professionals in the management of ALS. It is also important as it will draw attention to the contribution of these practices in increasing the life span and improving the quality of life of patients.

In this sense, the objective was to conduct a literature review on the use of mechanical ventilation in the treatment of patients with Amyotrophic Lateral Sclerosis (ALS) emphasizing the importance of the physical therapist's performance.

Method

Bibliographic research was used as the method, with the final need to present a descriptive literature review. Thus, searches were made for bibliographic references in books, scientific journals, theses and dissertations that deal with the applicability of respiratory physiotherapy and mechanical ventilation in patients with Amyotrophic Lateral Sclerosis (ALS). The following databases were accessed for this purpose: Capes Thesis Bank, IBICT Thesis and Dissertations Library, LA Reference, VHL Research Portal, FioCruz Periodical Portal, SciELO - Scientif Eletronic Library Online, Periodical Portal DynaMed. The relevant articles were selected, that is, those that fit the key words of the subject: Mechanical Ventilation, Invasive Mechanical Ventilation, Non-Invasive Mechanical Ventilation and Amyotrophic Lateral Sclerosis; and more current. Afterwards, the contents were recorded. Also, in consultation with the legislation on ALS, a survey was carried out on the Ministry of Health website that returned a relevant document on the topic.

Results and Discussion

The use of mechanical ventilation in patients with Amyotrophic Lateral Sclerosis

The table below lists the works that deal directly or indirectly with Mechanical Ventilation in the treatment of patients with Amyotrophic Lateral Sclerosis and some considerations that made them eligible as an aid in the elaboration of this work, either in the introduction of the topic, or in the discussion.

AUTHOR(S)/YEAR	TITLE	RELEVANT CONSIDERATIONS
MORELOT-PANZINI, C.; BRUNETEAU, G.; GONZALEZ-BERMEJO, J. /2019.[23]	NIV in Amyotrophic lateral sclerosis: The 'when' and 'how' of the matter	NIV significantly improves survival, quality of life and cognitive performance. However, the more advanced use of NIV also requires pulmonologists to master palliative care associated with the end of life, as well as the ways to stop ventilation when it becomes irrational.
FIORENTINO, G. F.; *et al.*/2018.[24]	Continuous noninvasive ventilation for respiratory failure in patients with amyotrophic lateral sclerosis: current perspectives	The decision to use or not to use non-invasive mechanical ventilation depends on factors such as health insurance coverage and varies from country to country. Based on the level of knowledge currently available, if the respiratory deficit is severe and NIV is ineffective, the alternative to NIV or death is invasive ventilation. This condition can only be effectively supported by a multidisciplinary team of specialists who make decisions together with patients and caregivers, respecting the will and dignity of the person.

FERREIRA, C. R.; METZKER, C. A. B.; ATHAYDE, F. T. S./2018.[25]	Benefits of non-invasive ventilation on chronic respiratory failure in patients with Amyotrophic Lateral Sclerosis	The investigated literature points to benefits related to the treatment of alveolar hypoventilation by the use of NIV in patients with ALS, specifically by improving survival, HRQoL (health-related quality of life), quality of sleep, GER (resting energy expenditure) and volumes pulmonary. Positive effects seem to be limited in the case of patients with the bulbar type, especially regarding survival.
DE MATTIA, E.; *et al.*/2018.[26]	Passive versus active circuit during invasive mechanical ventilation in subjects with amyotrophic lateral sclerosis.	The objective of the work was to evaluate the effectiveness and safety of a single member circuit with intentional leaks (passive circuit) compared to a circuit with an expiratory valve (active circuit). Currently, domestic ventilators can operate with a single limb circuit, even in cases of IMV, which streamlines patient management and mobilization. According to the type of exhalation port, there are 2 types of circuits for a single member: an active circuit, with a true expiratory valve that directs all the exhaled air out of the circuit; and a passive circuit, with a calibrated intentional leak placed closer to the patient. The passive circuit, with continuous flow to the circuit, proved to be as effective as the active circuit to avoid rebreathing. The passive circuit proved to be as effective and safe as the active circuit during invasive home ventilation in individuals with Amyotrophic Lateral Sclerosis.
MARKOVIC, N.; *et al.*/2018.[27]	Patterns of Non-Invasive Ventilation in Amyotrophic Lateral Sclerosis (Padrões de ventilação não invasiva na esclerose lateral amiotrófica)	A retrospective study indicates that the use of NIV in Amyotrophic Lateral Sclerosis increases over time and remains for a long period at the end of life in people living with ALS. It suggests that future prospective studies, exploring the use of NIV in the course of the disease and how NIV affects decision-making at the end of life in people with ALS are needed.
ROCHA, E.; OLIVEIRA, E. P. F.; LEMES, G. E. L./2018.[21]	Asynchrony during invasive mechanical ventilation: a literature review.	The ventilator-patient interaction can be synchronous or asynchronous and depends on how the ventilator will respond to the patient's respiratory effort and, in turn, how the patient will respond to the breath provided by the ventilator. Synchrony depends on several factors: sedation, compatibility of the machine's inspiratory time with the patient's neural, graphic observation, technique used for detection, type of pathology, ventilation mode,

		metabolic disturbance, fever, pain, delirium and confounding factors . As a result of the study, it was observed that the most common asynchrony was related to: the trigger (ineffective trigger, double trigger and self-trigger); flow (insufficient and excessive inspiratory) and cycling (early and late). Authors concluded that asynchrony makes it difficult to manage the patient, lengthens the time on IMV, which favors the appearance of harmful situations such as: greater waste of respiratory work, patient discomfort, increased need for sedation, confusion during weaning, ventilation prolonged mechanics, longer stay and probably higher mortality.
KETTEMANN, D.; *et al.*/2017.[28]	Clinical characteristics and course of dying in patients with amyotrophic lateral sclerosis withdrawing from long-term ventilation	The patient's reasons for interrupting ventilation, validating the quality of death and judging the procedure by family members cannot be explained. In ALS, with the decision to withdraw ventilation in the long term, the patient can define his own date of death. The experiences of the patient before removing ventilation, family and health professionals before and after are virtually unknown. The emotional and psychosocial experience close to death is a little explored field of research. The withdrawal of long-term ventilation (weaning) among ALS patients sets a precedent for research on this important issue.
DOMINGOS, Ana Margarida Marques./2017.[2]	Amyotrophic Lateral Sclerosis: A clinical case with inaugural Respiratory Failure.	The description of the clinical case presented and its evolution revealed an increase in survival after adaptation to NIV, although it was instituted late (24 months after the onset of symptoms) and already with signs of global respiratory failure. The duration of survival is significant compared to that described by some authors, cited in the same study, in similar cases, which can be explained by frequent monitoring, with individualized optimization of the parameterization of the ventilator.
MATOS, L. U. I.; RABAHI, M. F./ 2017.[29]	Respiratory management in neuromuscular diseases: literature review.	The institution of noninvasive mechanical ventilation at the appropriate time and coughing maneuvers during respiratory exacerbations, coupled with multidisciplinary involvement in driving patients and family members, have contributed considerably to improving quality of life and reducing mortality in neuromuscular diseases.
GUIMARÃES, V. S.; *et al.* 2017.[30]	Respiratory Complications in	This investigation shows that one of the main manifestations of ALS is muscle atrophy, loss of

	Amyotrophic Lateral Sclerosis and Rehabilitation Methods.	strength and, consequently, respiratory damage. These damages considerably decrease the quality of life of patients, since it affects the diaphragm and the external intercostal muscles, resulting in alveolar hypoventilation. Respiratory treatments can be invasive or non-invasive. There are researchers who suggest alternative therapies so that the most invasive interventions are avoided. Thus, methods such as non-invasive mechanical ventilation and physiotherapy treatments are applied that aim at the best excretion of pulmonary secretion. Commonly, treatments consist of an attempt to improve the individual's physiological functioning.
SANTOS, Gláucia da Silva. /2015.[31]	Characterization of the evolutionary picture of respiratory function in patients with Amyotrophic Lateral Sclerosis	Patients using noninvasive ventilation showed stability or improvement in the respiratory function variable, maximum voluntary ventilation (MVV).
FERRARESSO, Amanda.2013.[7]	Clinical and functional assessment of respiratory impairment in patients with Amyotrophic Lateral Sclerosis	The clinical and functional evaluation of patients with ALS made it possible to measure the severity of respiratory impairment, and also served to monitor the evolution of these patients after the breathing exercise program and home guidelines.
ALMEIDA, Sara Regina Meira. /2009.[22]	Amyotrophic lateral sclerosis: prospective study of respiratory parameters.	Authors believe to be feasible and recommended to use the support as early as possible, despite the diversity of the literature described, regarding the ideal tests for assessing respiratory function and the controversies at the beginning of NIMV. In the absence of an ideal parameter to monitor the evolution of respiratory impairment in ALS, periodic and multiparametric monitoring of the patient is necessary, with attention highlighted to symptoms and signs of respiratory distress.
LIMA, Núbia Maria Freire Vieira. /2009.[3]	Profile of generic and physiotherapeutic home care and palliative care to a group of patients with amyotrophic lateral sclerosis.	This study cites the home environment as being the most ideal for the application of Non-Invasive Mechanical Ventilation (VMNI).

MIRANDA, J. A. R.; MIRANDA, M.J./2007.[32]	Ventilatory dysfunction in Motor Neuron disease: when and how to intervene?	Invasive ventilation by tracheostomy is reserved for situations of severe bulbar involvement. However, the comfort and safety provided by the early introduction of NIV and adjacent techniques, allows the majority of patients and their families to choose not to have a tracheostomy, even in the appearance of potentially fatal respiratory failure.
MOREIRA, S.; *et al.* /2004.[33]	Acute respiratory failure as the first manifestation of amyotrophic lateral sclerosis: two clinical cases.	In the two cases studied, after hospitalization, both patients were discharged with non-invasive ventilation, with clinical improvement and independent activities of daily living, which allows us to conclude that after resolving the cause of decompensation and reducing respiratory work, there is a significant improvement in the clinical picture.

The most diverse authors studied present both non-invasive and invasive mechanical ventilation as necessary treatments for patients with ALS, and factors such as: treatment environment, costs must be considered when deciding between one form or another (non-invasive mechanical ventilation requires more human resources), health insurance coverage, worsening of the disease, among others.

Noninvasive mechanical ventilation

The Clinical Protocol and Therapeutic Guidelines for Amyotrophic Lateral Sclerosis, approved by Ordinance No. 1151, of November 11, 2015[34], of the Health Care Secretariat, of the Ministry of Health, indicates, among the non-pharmacological measures for the treatment in ALS, the noninvasive ventilatory support, in its various modalities, as the one that most increases the survival and quality of life of the patient with ALS. It also brings inspiratory muscle training as another practice with probable benefits in increasing survival and quality of life.

Morelot-Panzini, Bruneteau and Gonzalez-Bermejoniv[23] make a series of considerations regarding the use of noninvasive mechanical ventilation in patients with ALS. They point out that the site of initiation of NIV can be in any experienced environment and that no site has proven to be superior to another: home, outpatient care and even telemonitoring; all reporting successful establishment of NIV. However, the current trend, mainly due to the great motor disability of these patients, is to avoid hospitalization as much as possible, and are associated with cost savings in health. In the same vein, it has been shown that delayed onset of NIV outpatient treatment can save time and consequently improve patient survival.[23] Still, it is stated that the beginning of NIV is a relevant step in the life of patients with ALS, it can be feared and traumatic, as many associate mechanical ventilation with tracheostomy and the end of life. Thus, patients must be very well educated on this treatment modality.[23] They also indicate the following explanatory phrases as a way to reassure the patient and family: many auxiliary devices can help your breathing a lot, which left

unattended can lower your energy levels and prevent you from sleeping at night. One of these devices is a non-invasive positive pressure ventilator. Includes an easy-to-use mask that fits your face. It should increase energy and provide better sleep. It may be added that this treatment will also relieve your shortness of breath while you are using ventilatory assistance, but probably also when you are breathing on your own. [23]

Borges et al. as an example of the effectiveness of mechanical ventilation, its use as a therapeutic resource applied in ICUs with the aim of maintaining gas exchange, relieving respiratory work, reversing or preventing respiratory muscle fatigue and reducing oxygen consumption. They also state that, currently, most patients undergoing mechanical ventilatory support can be quickly removed from the ventilator by simply waiting for the condition responsible for their referral to treatment to have ceased or stabilized.[35]

Along the same lines, the importance of applying noninvasive mechanical ventilation is an example, which meant an advance in intensive therapeutic care in specific cases of acute respiratory failure (cardiogenic pulmonary edema, for example), situations in which NIV is used with the objective of reducing respiratory work and improving pulmonary gas exchange and avoiding tracheal intubation. However, they clarify that its use should be cautious, since NIV is a safe and efficient type of ventilatory support, but requires that patients be monitored and promptly intubated if their clinical conditions worsen.[36]

NIV is indicated as the technique that consists of applying artificial mechanical ventilation without the need for endotracheal prostheses, that the interaction between the patient and the ventilator occurs with the use of specific masks. They also indicate the Bilevel Positive Airway Pressure mode as being widely used for patients with neuromuscular diseases, consisting of two levels of pressure in the airways, one inspiratory and the other expiratory. Likewise, other ventilation types, with emphasis on volume-controlled ventilation, have also been used.[25]

Along the same line, some notes are made regarding the choice of ventilation equipment and configurations: no particular mode of ventilation proved to be superior, no difference in terms of effectiveness was identified between ventilation with assisted volume control (in which the patient receives a pre-defined volume of gas) and ventilation with pressure-assisted control (in which partial pressure assistance is provided); the disadvantage of volume-assisted ventilation is the rigid ventilation sensation and the absence of compensation for leaks, since the two main theoretical advantages are that it allows the patient to stack air to help release the airways and is able to overcome airflow obstruction. In invasive ventilation, this is the preferred mode. Some highly experienced teams effectively use this mode for NIV, probably with good effectiveness for obstructive events. The main advantages of assisted pressure control ventilation are that they are more comfortable for the patient, and support for low-level inspiratory pressure is usually sufficient when initiating NIV.[23]

In patients with neuromuscular disease[29], respiratory muscle weakness and increased pulmonary elastic retraction are responsible for rapid and shallow breathing patterns that lead to chronic CO2 retention. Thus, adequate and early ventilatory support, such as non-invasive ventilation, can reduce respiratory morbidity and mortality. The state of ventilation also worsens during sleep due to decreased function of the respiratory center and muscle relaxation, especially

in the REM sleep phase (Rapid Eye Movement, lower brain activity, however, faster; rapid eye movements and extreme muscle relaxation; phase where dreams occur). Thus, continuous and non-invasive monitoring of CO2 during the night is useful and considered for the detection of ventilatory failure. They also describe that the impairment of the respiratory musculature results in muscle atrophy and, consequently, in the limitation of the movements of the rib cage during respiratory incursions, as a consequence of this progressive chest restriction there is a continuous decrease in vital capacity. In this context, NIV aims to stabilize the decrease in vital capacity, correction of hypoxemia and hypercapnia, in addition to improving quality of life and sleep.

Adherence to noninvasive ventilation at the appropriate time and coughing maneuvers during respiratory exacerbations, combined with multidisciplinary involvement in the management of patients and family members, have been relevant factors for improving quality of life and reducing mortality in neuromuscular diseases.[29]

From 2006 to the present day[23], it demonstrated not only that non-invasive ventilation (NIV) is difficult to adjust to the management of ALS patients by pulmonologists, but also that NIV must be integrated into the multidisciplinary treatment, taking into account the progression of disease and the patient's conditions: living conditions outside the hospital. And that in this context, the family of the patient with ALS plays a predominant role in the attendance, support and daily management of treatment, including palliative care at the end of life.

Invasive mechanical ventilation

In patients with ALS[24], after acute respiratory failure and intubation, the prognosis is not always characterized by complete dependence on the ventilator. Therefore, it is necessary to analyze each case. However, the possibility of tracheostomy must be known by patients with ALS, in any case. The patient has the right to choose before critical events occur.

It is stated that invasive ventilation via tracheostomy should be considered only when there is bulbar dysfunction and risk for bronchoaspiration, and Amyotrophic Lateral Sclerosis is one of the main diseases that require this intervention at a certain stage of the disease.[29]

Invasive mechanical ventilation is one of the most common procedures in patients with acute or chronic respiratory failure in an ICU environment.[21] In addition, IMV can save the lives of patients who are unable to maintain a satisfactory breathing pattern. According to the authors, the main objectives of IMV are: to reduce overwork in the respiratory muscles, improve gas exchange and optimize respiratory function. Thus, IMV is used as a resource in an attempt to save the lives of patients who are unable to breathe spontaneously at the time and who are potentially recoverable.[21]

Tracheostomy has been shown to significantly improve survival, with quality of life considered satisfactory by patients. The technical and financial viability of this approach can be important determinants of tracheostomy placement in some countries, with the likelihood that the patient will be able to return to their place of residence after the tracheostomy, an important factor in decision making. Some countries do full reimbursement and others do partial

reimbursement, some like the UK do not do reimbursement for tracheostomy. This lack of reimbursement can lead to problematic and irreparable situations for families of ALS patients. However, some patients and their families request a tracheostomy and should be informed as completely as possible about the consequences of this decision, ideally as soon as possible, to avoid performing a tracheostomy in an acute environment when the patient and family do not have time to discuss this option.[23]

Noninvasive mechanical ventilation vs. invasive mechanical ventilation: considerations

Noninvasive ventilation (NIV) is the standard of treatment for ALS, whereas tracheostomy with invasive ventilation (TVI) is performed in a smaller number of ALS patients. However, they present data stating that in some countries, including Germany, the frequency of TVI is increasing. They indicate that the reasons underlying the constant increase in treatment that prolongs life with NIV or VI are the improvement of home ventilation, the availability of electronic communication systems and the availability of participatory activities online.[28]

Both in VMNI and prolonged IMV may be related to substantial psychosocial tension for patients, family members and caregivers, resulting in a patient's desire to withdraw ventilatory therapy. List also the typical factors that contributed to the withdrawal of long-term ventilation (withdrawal of long-term ventilation - WLTV), which are: the burden of care, social deprivation or loss of communication capacity through an ophthalmoplegia in use . ALS and the blocking syndrome.[28] Mechanical ventilatory support, regardless of whether invasive or non-invasive, must be applied in an appropriate and safe manner to avoid the injury induced by mechanical ventilation.[37]

The quality of life in continuous non-invasive ventilation is the result of objective and subjective indicators, and its quantification is not simple. In patients with ALS, quality of life is closely linked to disability, but also to individual characteristics, as well as cultural and social factors.[24] For human dignity can be presented as the ability to exercise free will and choice. Thus, it is always the patient who decides and who has the final say. In patients with ALS, noninvasive mechanical ventilation leads to an increase in survival time and also in quality of life. The authors indicate that studies concluded that, in patients with the same level of dependence, invasive mechanical ventilation does not affect quality of life. The participation of caregivers, psychologists and support to prepare the patient for the progression of the disease and to establish communication on end of life issues. They also show that current US and European guidelines recommend that patients with ALS make correct and informed choices about IMV and palliative care. This should occur from the early stages of ALS diagnosis and is especially necessary when NIV therapy is no longer tolerated or when the progression of the disease makes it ineffective.

Conclusion

Mechanical ventilation is a necessary procedure in treating patients with ALS and there is no effective rule as to the type of mechanical ventilation that is more appropriate when dealing with ALS patients. When deciding on the type of ventilation to apply in the treatment, the conditions and level of aggravation of the patient, costs, mobility, coverage of the health plan, the will of the patient and the family (previously instructed) must be considered. Since noninvasive mechanical ventilation is generally used at the beginning of treatment, and with the increased dependence of the patient, it migrates to invasive ventilation. But, it is also a consensus that it is up to the patient and family, duly clarified, to choose mechanical ventilation or not, and for one or another modality, and when to interrupt the treatment, if deemed necessary.

Future updating and deepening studies are necessary given the relevance of the topic to the area of respiratory health and physiotherapy.

References

1. Prado LG, Bicalho IC, Vidigal-Lopes M, Prado VG, Gomez RS, de Souza LC, Teixeira AL. Depressão e ansiedade em uma série de casos de esclerose lateral amiotrófica: frequência e associação com aspectos clínicos. Einstein. 2017;15(1):58-60.

2. Domingos AMM. Esclerose Lateral Amiotrófica: Um caso clínico com Insuficiência Respiratória inaugural. Dissertação[Mestrado integrado em medicina]- Faculdade de Medicina de Lisboa; 2017.

3. Lima NMFV. Perfil da assistência domiciliar genérica e fisioterapêutica e cuidados paliativos a um grupo de pacientes com esclerose lateral amiotrófica. Dissertação [Mestrado]- Faculdade de Ciências Médicas, Universidade Estadual de Campinas; 2009.

4. Associação Brasileira de Esclerose Lateral Amiotrófica. Esclerose Lateral Amiotrófica. São Paulo: ABRELA; 2013.

5. Lima VCS, Mourão Junior CA, Malaguti C. Ajuste ocupacional com uso de tecnologia de comunicação em paciente com esclerose lateral amiotrófica: um autorrelato. Sci. med. (Porto Alegre, Online) ; 27(4): e27984.

6. Dynamed. Amyotrophic Lateral Sclerosis. Estados Unidos: Dynamed; 2019. Disponível em: https://www.dynamed.com/condition/amyotrophic-lateral-sclerosis-als . Acesso em 10 nov 2019.

7. Ferraresso A. Avaliação clínica e funcional do comprometimento respiratório de pacientes com Esclerose Lateral Amiotrófica. Dissertação[Mestrado]- Faculdade de Ciências Médicas, Universidade Estadual de Campinas; 2013.

8. Luchesi KF, Silveira IC. Cuidados paliativos, esclerose lateral amiotrófica e deglutição: estudo de caso. CoDAS 2018;30(5):e20170215 . Doi: https://doi.org/10.1590/2317-1782/20182017215.

9. Prochno CA. Monitoração não invasiva da pressão intracraniana e efeitos de dois protocolos de fisioterapia na força muscular respiratória em pacientes submetidos à cirurgia cardíaca. Dissertação[Mestrado]- Universidade Estadual de Ponta Grossa; 2018.
10. Coutinho MGC. Análise do efeito da fisioterapia respiratória isolada ou associada a exercícios aeróbicos, sobre a capacidade física e a qualidade de vida de pacientes bronquiectásicos. Monografia [Graduação em Fisioterapia] - Universidade Estadual do Oeste do Paraná; 2005.
11. Renault JÁ, Costa-val R, Rossetti MB. Fisioterapia respiratória na disfunção pulmonar pós- cirurgia cardíaca. Rev Bras Cir Cardiovasc 2008; 23(4): 562-9.
12. Romanini W, Muller AP, Carvalho KAT, Olandoski M, Faria-Neto JR, Mendes FL, et al. The Effects of Intermittent Positive Pressure and Incentive Spirometry in the Postoperative of Myocardial Revascularization. Arq Bras Cardiol. 2007 Aug;89(2):94-9, 105-10. doi: https://doi.org/10.1590/s0066-782x2007001400006 .
13. Lopes CR, Brandão CMA, Nozawa E, Auler Júnior JOC. Benefícios da ventilação não- invasiva após extubação no pósoperatório de cirurgia cardíaca. Rev Bras Cir Cardiovasc 2008; 23(3): 344-50.
14. EBSERH-Empresa Brasileira de Serviços Hospitalares. Ministério da Educação. POP: Técnicas de Fisioterapia Respiratória em Pacientes Adultos -- Unidade de Reabilitação do Hospital de Clínicas da Universidade Federal do Triângulo Mineiro. Uberaba: POP 025; 2015.
15. Sarmento G J V. O ABC da fisioterapia repiratória. 2ª edição. São Paulo: Manole; 2015.
16. De Oliveira M, Santos CLS, de Oliveira CF, Ribas DIR. Efeitos da técnica expansiva e incentivador respiratório na força da musculatura respiratória em idosos institucionalizados. Fisioter Mov. 2013; 26(1):133-40. Doi: https://doi.org/10.1590/S0103-51502013000100015
17. Pissinato IG, Karsten M, Neves LMT, Minatel V, Borghi-Silva A, Catai AM.Pressão expiratória positiva nas vias aéreas não reproduz as respostas de frequência cardíaca à manobra de Valsalva em homens jovens saudáveis. Fisioter Pesq. 2012;19(2):178-84. Doi: http://dx.doi.org/10.1590/S1809-29502012000200015
18. Rocha RSB, Cunha KC, Nina JC. Variáveis cardiorrespiratórias e expansibilidade torácica antes e após ouso do incentivador respiratório no pós operatório de revascularização do miocárdio. Rev Saúde. 2013; 13: 47-54. Doi: http://dx.doi.org/10.15600/2238-1244/sr.v13n33p47-54
19. Shelledy DC, Wielzali Carl P. História da terapia Respiratória. In: EGAN, Fundamentos da Terapia Respiratória. Rio de Janeiro: Elsevier; 2009.
20. Aboussouan LS. EGAN, Fundamentos da Terapia Respiratória. Rio de Janeiro: Elsevier; 2009.
21. Rocha E, Oliveira EPF, Lemes GEL. Assincronia durante a ventilação mecânica invasiva: uma revisão na literatura. Rev Inspirar Mov Saude. 2018; 18(4): 1-12.

22. Almeida SRM. Esclerose lateral amiotrófica: estudo prospectivo de parâmetros respiratórios. Dissertação[Mestrado]- Faculdade de Ciências Médicas, Universidade Estadual de Campinas; 2009.
23. Morelot-Panzini C, Bruneteau G, Gonzalez-Bermejo J. NIV in amyotrophic lateral sclerosis: The 'when' and 'how' of the matter. Respirology. 2019;24(6):521-530. Doi: http://dx.doi.org/10.1111/resp.13525
24. Fiorentino G, Annunziata A, Gaeta AM, Lanza M, Esquinas A. Continuous Noninvasive Ventilation for Respiratory Failure in Patients With Amyotrophic Lateral Sclerosis: Current Perspectives. Degener Neurol Neuromuscul Dis. 2018 Sep 4;8:55-61. doi: http://dx.doi.org/10.2147/DNND.S170771 .
25. Ferreira CR, Metzker CAB, Arhayde FTS. Benefícios da ventilação não invasiva sobre a insuficiência respiratória crônica em pacientes com Esclerose Lateral Amiotrófica. Rev Ciên Saúde. 2018;3(2):1-6.
26. De Mattia E, Falcier E, Lizio A, et al. Passive Versus Active Circuit During Invasive Mechanical Ventilation in Subjects With Amyotrophic Lateral Sclerosis. Respir Care. 2018;63(9):1132-1138. doi: http://dx.doi.org/10.4187/respcare.05866
27. Markovic N, Povitz M, Smith J, Leasa D, Shoesmith C, Gofton TE. Patterns of Non-Invasive Ventilation in Amyotrophic Lateral Sclerosis. Can J Neurol Sci. 2018;45(4):445-450. doi: http://dx.doi.org/10.1017/cjn.2018.25
28. Kettemann D, Funke A, Maier A, et al. Clinical characteristics and course of dying in patients with amyotrophic lateral sclerosis withdrawing from long-term ventilation. Amyotroph Lateral Scler Frontotemporal Degener. 2017;18(1-2):53-59. doi: http://dx.doi.org/10.1080/21678421.2016.1214734
29. Matos LUI, Rabahi MF. Manejo respiratório em doenças neuromusculares: revisão de literatura. Rev Educ Saúde. 2017; 5 (2): 121-9. Doi: https://doi.org/10.29237/2358-9868.2017v5i2.p121-129
30. de Souza Guimarães V, Henriques da Silva C, Freitas Neves Y, Lopes Dutra T. Complicações respiratórias na esclerose lateral amiotrófica e métodos de reabilitação. REIPEC. 2017;3(2): 269-77.
31. Santos GS. Caracterização do quadro evolutivo da função respiratória dos pacientes com Esclerose Lateral Amiotrófica. Dissertação[Mestrado]- Escola Bahiana de Medicina e Saúde Pública; 2015.
32. Miranda JAR.; Miranda MJ. Disfunção ventilatória na doença do Neurônio Motor: quando e como intervir? Acta Med Port 2007; 20: 157-65.
33. Moreira S, Tátá M, Carvalho L, Mata JP. Insuficiência respiratória aguda como primeira manifestação de esclerose lateral amiotrófica: dois casos clínicos. Rev Port Pneumol . 2004. 10 (6): 499-504.
34. Brasil. Ministério da Saúde, Secretaria de Atenção à Saúde. Portaria nº 1151, de 11 de novembro de 2015. Protocolo Clínico e Diretrizes

Terapêuticas da Esclerose Lateral Amiotrófica. Brasília: Ministério da Saúde; 2015.
35. Borges DL, Arruda LA, Rosa TRP, Costa MAG, Baldez TEP, Silva GJP. Influência da atuação fisioterapêutica no processo de ventilação mecânica de pacientes admitidos em UTI no período noturno após cirurgia cardíaca não complicada. Fisioter Pesqui 2016;23(2):129-35. Doi: http://dx.doi.org/10.1590/1809-2950/14133523022016
36. Godoy ACF, Calderan TRA, Fraga GP. Ventilação mecânica não invasiva em uma paciente com pancreatite aguda e insuficiência respiratória. J Bras Pneumol. 2012;38(5):677-678.
37. Barbas CS, Ísola AM, Farias AM, Cavalcanti AB, Gama AM, Duarte AC, et al. Recomendações brasileiras de ventilação mecânica 2013- Parte I. Rev Bras Ter Intensiva. 2014;26(2):89-121 doi: https://doi.org/10.5935/0103-507X.20140017

Correspondent Author
Débora Dadiani Dantas Cangussu
G South 9 Q CS 11/12/15/16. ZIP: 72035-509.
Taguatinga, Federal District, Brazil.
deboradadiani@gmail.com