Enfermería Global
ISSN 1695-6141 Revista electrónica trimestral de Enfermería
Nº63 Julio 2021
www.um.es/eglobal/

REVISIONES

Effects of the Covid-19 pandemic on the quality of life of patients with Amyotrophic Lateral Sclerosis

Efectos de la pandemia por Covid-19 sobre la calidad de vida de los pacientes con Esclerosis Lateral Amiotrófica

Sara Bermudo Fuenmayor[1]
Pedro Jesús Serrano Castro[2]
Pablo Quiroga Subirana[3]
Tesifón Parrón Carreño[4]

[1] Nurse, PhD candidate at the University of Almería. Nurse at Hospital de Poniente, Almería. Spain. sbf319@inlumine.ual.es
[2] Physician, University of Malaga. Neurologist at the Regional Hospital of Malaga. Spain.
[3] Physician, University of Almería. Neurologist at Torrecárdenas Hospital Complex in Almería. Spain.
[4] Doctor. Family and Community Medicine Specialist. Specialist in Preventive Medicine and Public Health. Professor at the University of Almería. Head of Service of the Territorial Delegation of Equality, Health and Social Policies. Almería. Spain.

https://doi.org/10.6018/eglobal.457891

Received: 30/11/2020
Accepted: 3/03/2021

ABSTRACT:

Introduction: In recent months, the emergence of SARS-CoV-2 as a global pandemic has caused a devastating impact on health worldwide. Health systems have been overwhelmed, and the follow-up of those patients who require continuous monitoring and evaluation, such as Amyotrophic Lateral Sclerosis (ALS), have been negatively affected.
Material and method: A search of the current literature has been carried out to determine the impact of the Covid-19 pandemic on the quality of life of patients diagnosed with Amyotrophic Lateral Sclerosis and their family.
Results: The diagnosis, treatment, follow-up and home care of these patients have been modified, adapting them as much as possible to the technology and available resources to try to minimize the loss of quality of life of patients diagnosed with Sclerosis Amyotrophic lateral and ensuring comprehensive care.
Conclusion: Despite numerous efforts and advances in research, both in the case of ALS and COVID-19, there is still a long way to go.

Keywords: Amyotrophic Lateral Sclerosis; ALS; COVID-19; Coronavirus; SARS-COV2; Quality of life.

RESUMEN:

Introducción: En los últimos meses, la irrupción del SARS-CoV-2 como pandemia mundial ha causado un impacto devastador en la sanidad a nivel mundial. Los sistemas sanitarios se han visto desbordados, y el seguimiento de aquellos pacientes que requieren un control y evaluación continuos, como es el caso de la Esclerosis Lateral Amiotrófica (ELA), se han visto afectados negativamente.

Material y Método: Se ha llevado a cabo una búsqueda de la literatura actual para determinar el impacto de la pandemia de Covid-19 sobre la calidad de vida de los pacientes diagnosticados de Esclerosis Lateral Amiotrófica y su familia.
Resultados: El diagnóstico, tratamiento, seguimiento y cuidados domiciliarios de estos pacientes se han visto modificados, adaptándolos en la medida de los posible, a la tecnología y los recursos disponibles para tratar de reducir al mínimo la pérdida de calidad de vida de los pacientes diagnosticados de Esclerosis Lateral Amiotrófica y asegurando la atención integral.
Conclusión: A pesar de los numerosos esfuerzo y avances en la investigación, tanto en el caso de la ELA como en la COVID-19, queda mucho camino por andar.

Palabras clave: Esclerosis Lateral Amiotrófica; ELA; COVID-19; Coronavirus; SARS-COV2; Calidad de vida.

INTRODUCTION

Amyotrophic Lateral Sclerosis (ALS) is a progressive, degenerative neuromuscular disease of unknown origin. It presents an affectation of the motor neurons in the cerebral cortex, brainstem, and spinal cord, with a rapid and fatal evolution [1–3].

As the disease progresses, dysphagia is common, increasing the risk of aspiration, respiratory infection, pneumonia, and suffocation. When the respiratory muscles are affected, there is a loss of the effective ability to cough and clear the throat voluntarily, limiting the ability to clear debris from the pharynx. This causes a progressive limitation of ventilation. Eventually, global respiratory failure occurs. Death usually occurs within 3-5 years from diagnosis, due to respiratory failure [3–7].

Today, despite numerous efforts, there is no curative treatment for ALS- Palliative care and treatment of symptoms are key to the approach to these patients [6,7].

Coronavirus disease 2019 (COVID-19) is an infectious disease caused by a new strain of coronavirus, severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) [8].

Following the first report in China, the World Health Organization (WHO) declared SARS-CoV-2 a global pandemic on March 11, 2020, with a devastating impact on health practice around the world [8–11].

As a result of the emergence of Covid-19, health systems have been forced to reorganize their resources, as well as the spaces available for health care. Specialized care was adapted for the treatment and care of patients with COVID-19, canceling all non-urgent scheduled activities. This had a direct impact on the follow-up of those patients who require continuous monitoring and evaluation, such as Amyotrophic Lateral Sclerosis (ALS) [6].

Also, in COVID-19, respiratory distress is the most characteristic symptom. More than half of the patients infected with SARS-CoV2 with dyspnea required admission to intensive care. Admission to intensive care units increases the risk of developing a neuromuscular disorder related to prolonged immobilization, the appearance of recurrent infections, the requirement for mechanical ventilation or corticosteroids. Therefore, if we combine both diseases, ALS and COVID-19, the result can be devastating [6,8].

The objective that we propose with our work is to identify the effects that the COVID-19 pandemic has had on the health care and quality of life of patients diagnosed with Amyotrophic Lateral Sclerosis.

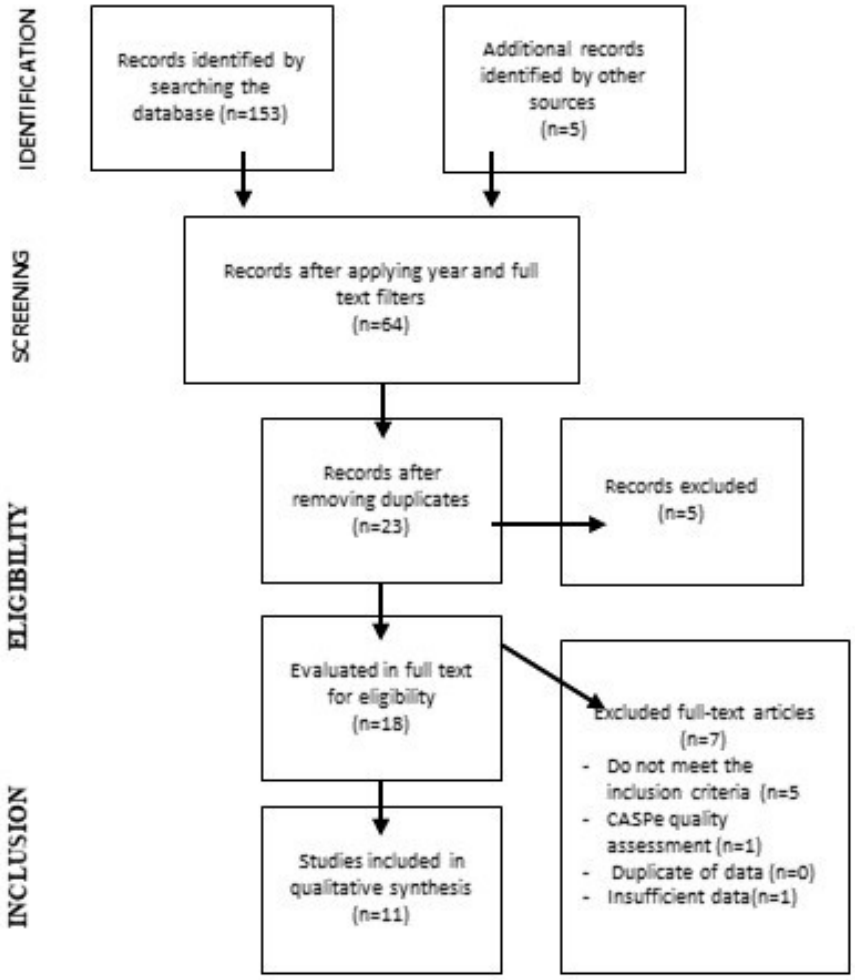
MATERIAL AND METHOD

A search of the current literature has been carried out to determine the impact of the Covid-19 pandemic on the quality of life of patients diagnosed with Amyotrophic Lateral Sclerosis and their family. This search took place in the Medline, ÍnDICEs-CSIC, Cochrane, and CUIDEN databases between September 1, 2020, and November 15, 2020. The keywords used in the search were 'Amyotrophic Lateral Sclerosis',' ALS ',' Covid-19, 'Sars-cov-2 ”in English, and“ Amyotrophic Lateral Sclerosis ”,“ ALS ”in Spanish, as well as the free term“ coronavirus ”. The operators used were "AND" and "OR". The search strategies used were (Amyotrophic Lateral Sclerosis) AND (Covid-19); (ALS) AND (Covid-19); (ELA) AND (Covdi-19); (Amyotrophic Lateral Sclerosis) AND (SARS-COV-2); (Amyotrophic Lateral Sclerosis) AND (SARS-COV-2); and (Amyotrophic Lateral Sclerosis) AND (Covid-19).

The result of the search carried out concluded with 153 articles, of which 11 were included in the present review. Duplicate articles or papers that did not comply with the CASPe methodology were excluded.

Figure 1 shows the information flow diagram followed for the methodology through the different phases of the bibliographic review (see figure 1).

Figure 1



RESULTS

The current coronavirus pandemic (COVID-19) has had an impact on global health, with great repercussions on the physical and psychological health of the general population, creating unprecedented challenges for the approach to neuromuscular disease [3,12,13].

This new health situation forced the reorganization of the public care services in all hospitals and health centers. The greatest changes and difficulties derive from the need to modify the approach and management of patients, as they are forced to postpone routine follow-up visits and cancel tests in the hospital setting. This has led to an increase in morbidity and mortality in many patients with neuromuscular diseases, including patients diagnosed with Amyotrophic Lateral Sclerosis (ALS).

Consequently, this situation also resulted in a delay in the diagnosis of new pathologies[6,10,11].

The results of a recent survey by the Northeast Amyotrophic Lateral Sclerosis Consortium (NEALS) in the United States show the modification that COVID-19 has caused in the clinical evaluation of patients with ALS, reducing face-to-face consultations and incorporating consultations via video conferencing as an alternative. This survey detected difficulties in accessing the multidisciplinary team, home care and palliative care, as well as difficulties in obtaining essential resources for these patients such as feeding tubes or wheelchairs[13].

The diagnosis

During the pandemic, more than 3.9 billion people have been confined to their homes in more than 90 countries, limiting medical assistance to emergency cases, with special attention to patients at risk, such as ALS. The overload of the health systems, and the fear of contagion by these patients, who already have a basic respiratory problem, limited the care of these patients to exceptional emergency cases [11,14].

For patients diagnosed with ALS, whose early diagnosis is key to preserving their quality of life, all this delay in follow-up and in performing diagnostic tests has a direct impact on survival[10].

In the context of the pandemic, key diagnostic techniques in assessing the prognosis of ALS, and fundamental for decision-making such as spirometry, have been discouraged by experts, since they can stimulate coughing, generating aerosols, the main route transmission of COVID-19. This delayed the introduction of tools such as non-invasive ventilation, invasive ventilation, or enteral feeding, the incorporation of which is related according to the available evidence, with an improvement in the quality of life of these patients. Furthermore, the spirometry result is a key criterion for inclusion or exclusion in clinical trials[13].

On the other hand, COVID-19 presents a great diversity of nonspecific symptoms, such as headache, dizziness, anorexia, and diarrhoea, and neurological symptoms such as loss of taste, hearing, and sight; confusion or decreased level of consciousness, there are even registries that identify cases of seizure or cerebrovascular accident, which makes its diagnosis even more difficult, raising a greater concern for the approach to the safety of the patient with ALS and their caregiver[15].

In recent years, the development of technology has allowed patients with neurological disorders to access health services from home, through standard telemedicine (with videoconference), telehealth (with clinical monitoring by the multidisciplinary team), telecoaching and telecare (to support independent living), among others, which also improve the quality of life of these patients during the Covid-19 pandemic[11].

The diagnosis of Amyotrophic Lateral Sclerosis requires an evaluation and physical examination to be able to detect signs and symptoms of alterations in the lower motor neurons. Although the clinical interview or anamnesis can be carried out virtually, it is not possible to assess muscle mass, muscle tone, and muscle power or tendon

reflexes, hindering the chances of an early diagnosis, which is already complicated [6,16].

The use of telemedicine allows monitoring at home, monitoring the progression of the disease, detecting signs and symptoms early, and allows us to introduce resources that improve the quality of life and independence of these patients, such as They are ankle prostheses, cane or walker to maintain posture, or wheelchair, in more advanced cases, adapted cutlery, communication, and writing support, among others[15].

In the most advanced stages of ALS, we will find a patient with respiratory failure, on many occasions, under treatment with non-invasive ventilation, so if we add to this base pathology the addiction of COVID-19, the risk complications is very high. Therefore, it is important that the multidisciplinary team make adaptations to ensure the safety and well-being of the patient and their caregiver, providing information on COVID-19 that allows early detection to prevent possible complications[7].

The treatment

With the restriction of hospital follow-up visits, the evaluation of the need to incorporate tools such as assisted ventilation or gastrostomy, which improve the quality of life of these patients, was affected. Both techniques are effective the less time elapses for its incorporation as a complementary tool, for example, in the case of gastrostomy, a forced vital capacity above 50% of the normal range is considered as the main indicator for its introduction. Early counselling of patients and their families, even in non-dysphagia stages, is essential to promote informed decision-making[13].

Regarding treatment, despite the absence of curative treatment, there are certain drugs that slow the progression of the disease, such as riluzole. During the pandemic, a reduction in the availability of many of these treatments has been detected [10,16].

This search for curative treatment continues to be carried out through clinical trials. However, in the context of the pandemic for the field of research and progress in treatment, a decrease in the enrolment of new patients in clinical trials has also been identified[10].

Other of the fundamental resources in the treatment of ALS, such as physical therapy, speech therapists, and occupational therapists, have also been affected. The implementation of the royal decree that limited mobility, access to rehabilitation therapies, work on flexibility, muscle strength, and joint resistance, as well as phonation and communication, has become difficult [3,11].

Monitoring

Due to the complexity of the symptoms of patients with ALS and their rapid evolution, the follow-up of these patients prior to the pandemic was carried out by a multidisciplinary team composed of a neurologist, pulmonologist, nutritionist, physiotherapist, nurse, and psychologist, who assess the progression of ALS every 1, 3, or 5 months. With the new situation, this monitoring is transferred to telemedicine, and many countries have been adapting so that the multidisciplinary team is available and available to the patient and their family or caregivers on a continuous basis

through the network [11,16].

In addition, the rapid progression of the disease generates great disability. This requires constant adjustment of treatment, psychological support, and close monitoring of its evolution so that a delay in health care can lead to significant functional deterioration, making it difficult to deal with complications that may arise[6].

With the suspension or reduction to a minimum of the number of visits in hospital centers, in order to reduce the risk of contagion, it becomes essential to locate a new form of communication and monitoring of these patients. The objective of the multidisciplinary team is to avoid a great deterioration of physical and psychological functions. Therefore, there is a boost in the use of emerging technologies for the relationship with the interdisciplinary team and telemedicine becomes key to facilitate communication with the patient and their family[13].

For monitoring the evolution of the disease, the neurological examination and the use of validated scales such as the revised ALS Functional Scale (ALSFRSr) are very important, key tools for the early detection of serious symptoms, such as the appearance of dysphagia or respiratory failure. , which, if diagnosed early, reduces the risk of developing aspiration pneumonia and respiratory failure, thus improving the prognosis. The ALSFRS-R scale can be obtained by phone or video call. However, we do not have objective methods that assess the progress of the disease[3,6,11].

Regardless, the evolution of technology and the advent of telemedicine also have a positive effect, as demonstrated by a study conducted by Veterans Affairs, the ALS Center in Ohio. This study says that, despite the possible problems with the audio, the difficulty of physical examination, the more impersonal tone, with a lack of privacy and emotional connection. This type of care allows obtaining information about the address and the resources available to the patient and her caregiver. In addition, the use of video calls avoided trips to the hospital, which can be a cause of fatigue, tiredness, and stress in these patients, allowing them to have a calmer and more comfortable conversation [14].

Home care

The provision of information and training in-home care have also been affected by COVID-19, adding the doubts and uncertainties related to the disease (appointments, management of pharmacological treatment, non-invasive mechanical ventilation, and nutrition), with that related to Covid-19 (risk factors, contagion mechanism, preventive measures, effects)[15].

Frequently, the ALS patient uses airway cleansing devices, nebulization, aspiration, and use of assistive cough devices. When non-invasive ventilation is used, the spread of SARS-CoV-2 virus particles increases, due to the use of masks for ventilation, or to a poor fit of these. For this reason, in this situation, it is important that modifications are made to the mechanical ventilation equipment to reduce the transmission of the virus and therefore the possibility of contagion to their caregivers[17].

Mental health

Another aspect that we must not forget is the impact that the pandemic produces at a psychological level since the anxiety and fear that are already linked to the diagnosis of the disease are added to those related to the appearance of Covid -19. There are several studies that show the intensification of emotional distress, anxiety, and depression when concern about contracting the infection, loneliness due to isolation, and difficulty in accessing resources is added. Likewise, an increase in the burden on the caregivers of these patients has been identified, due to an increase in the needs of these patients, which are aggravated by the pandemic and confinement [12].

A study indicates that, although the long-term impact of the COVID-19 pandemic is not known, a higher level of anxiety, anguish, and feelings of loneliness has been identified in families with patients with ALS [15].

DISCUSSION

The coronavirus pandemic has forced the reorganization of health systems around the world, creating the need to introduce changes in the approach to the face-to-face therapeutic relationship that has been developing to date.

These changes have had a direct impact on the diagnosis, treatment, and follow-up of the disease until the end of life of patients diagnosed with Amyotrophic Lateral Sclerosis, as well as indirectly on the informal care at the home of these patients, affecting the overload of the caregiver's role and the mental health of both the patient and his family.

ALS, despite being a disease under study, with great advances in research, continues to be considered one of the rare diseases. The appearance of Covid-19 has also had a direct impact on the advancement of ALS research, being forced to paralyze clinical trials on many occasions.

The continuous effort to provide the best quality of care by the multidisciplinary team to these patients, introduces an opportunity for adaptation in the face of adversity, emerging new paradigms of interaction between healthcare personnel and the patient and their family that will improve the efficiency of the attention.

CONCLUSION

COVID-19 may open a door to the need to introduce modifications in the diagnosis and follow-up protocols in patients with neuromuscular disorders from home, adapting physical assessment methods, such as functional tests, by alternatives such as questionnaires or assessment scales validated, such as the use of the functional assessment scale ALSFRS-R.

The diagnosis of ALS has a great biopsychosocial impact. Generally, both the patient and her family know the progression of the disease, as well as the result from the diagnosis. Therefore, the psychological approach and support at all times are important, a fact that is very limited in the current situation due to the pandemic, the limitation of face-to-face visits, and home confinement.

In Spain, due to the main route of airborne transmission, one of the measures implemented to combat transmission was the mandatory use of masks. In patients with ALS, with a baseline respiratory limitation, this can be a source of anxiety and worsening of their quality of life, as many require non-invasive mechanical ventilation and oxygen therapy. Unfortunately, to date, there are no studies evaluating the effect of the use of masks in these patients.

There are not enough studies to demonstrate the impact of SARS-COV2 treatment in patients diagnosed with ALS.

Despite numerous efforts and advances in research, both in the case of ALS and in the case of COVID-19 much remains to be studied and investigated.

REFERENCES

1. Cassmeyer V, Long B, Phipps W. Médico-quirúrgica. 3ª. Madrid: Harcourt Brace; 1997.
2. Lemone P, Burke K. Enfermería médico-quirúrgica. Pensamiento crítico en la asistencia del paciente. Volumen I. 4ª. Madrid: Pearson Educación; 2009. 1122.
3. Pinto S, Quintarelli S, Silani V. New technologies and Amyotrophic Lateral Sclerosis – Which step forward rushed by the COVID-19 pandemic? Vol. 418, Journal of the Neurological Sciences. Elsevier B.V.; 2020.
4. Estrategia en Enfermedades Neurodegenerativas del Sistema Nacional de Salud [Internet]. Ministerio de Sanidad, Servicios Sociales e Igualdad. 2016. p. 1–150. Available from: http://www.mscbs.gob.es/organizacion/sns/planCalidadSNS/pdf/Est_Neurodegenerativas_APROBADA_C_INTERTERRITORIAL.pdf
5. Wijesekera LC, Leigh PN. Amyotrophic lateral sclerosis. Orphanet Journal of Rare Diseases. 2009;4(1):1–22.
6. Bombaci A, Abbadessa G, Trojsi F, Leocani L, Bonavita S, Lavorgna L. Telemedicine for management of patients with amyotrophic lateral sclerosis through COVID-19 tail. Neurological Sciences [Internet]. 2020 Oct 6; Available from: http://link.springer.com/10.1007/s10072-020-04783-x
7. Steinbach R, Prell T, Gaur N, Stubendorff B, Roediger A, Ilse B, et al. Triage of Amyotrophic Lateral Sclerosis Patients during the COVID-19 Pandemic: An Application of the D50 Model. Journal of Clinical Medicine. 2020 Sep 5;9(9):2873.
8. Nasuello NA, Pettinaroli R, Godi L, Savoini C, de Marchi F, Mazzini L, et al. Critical illness neuro-myopathy (CINM) and focal amyotrophy in intensive care unit (ICU) patients with SARS-CoV-2_ a case series. Neurological Sciences. 2020 Oct 13;1–3.
9. Rajabally YA, Goedee HS, Attarian S, Hartung HP. Management challenges for chronic dysimmune neuropathies during the COVID-19 pandemic. Muscle and Nerve. 2020 Jul 1;62(1):34–40.
10. Solé G, Salort-Campana E, Pereon Y, Stojkovic T, Wahbi K, Cintas P, et al. Guidance for the care of neuromuscular patients during the COVID-19 pandemic outbreak from the French Rare Health Care for Neuromuscular Diseases Network. Revue Neurologique. 2020 Jun 1;176(6):507–15.
11. de Marchi F, Cantello R, Ambrosini S, Mazzini L. Telemedicine and technological devices for amyotrophic lateral sclerosis in the era of COVID-19. Neurological Sciences. 2020;41:1365–7.

12. Consonni M, Telesca A, Bersano E, Lauria G. Amyotrophic lateral sclerosis patients' and caregivers' distress and loneliness during COVID-19 lockdown. Journal of Neurology [Internet]. 2020 Jul 21 [cited 2020 Nov 2];1–4. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC7372539/
13. Andrews JA, Berry JD, Baloh RH, Carberry N, Cudkowicz ME, Dedi B, et al. Amyotrophic lateral sclerosis care and research in the United States during the COVID-19 pandemic: Challenges and opportunities. Muscle and Nerve. 2020 Aug 1;62(2):182–6.
14. de Almeida JPL, Pinto AC, Pereira J, Pinto S, de Carvalho M. Implementation of a wireless device for real-time telemedical assistance of home-ventilated amyotrophic lateral sclerosis patients: A feasibility study. Telemedicine and e-Health. 2010 Oct 1;16(8):883–8.
15. Young EI, Manta D, Sah BP, Brooks BR. Modification of non-invasive ventilation for the advanced amyotrophic lateral sclerosis patient during the COVID-19 pandemic - do it now. Vol. 414, Journal of the Neurological Sciences. Elsevier B.V.; 2020.
16. Andrews J. Amyotrophic Lateral Sclerosis: Clinical Management and Research Update (Jinsy Andrews). 2009; Available from: http://download.springer.com.ezproxy1.lib.asu.edu/static/pdf/930/art%253A10.1007%252Fs11910-009-0010-0.pdf?originUrl=http%3A%2F%2Flink.springer.com%2Farticle%2F10.1007%2Fs11910-009-0010-0&token2=exp=1455166184~acl=%2Fstatic%2Fpdf%2F930%2Fart%25253A10.1007
17. de Lucía N, Pio Ausiello F, Spisto M, Manganelli F, Salvatore E, Dubbioso R. The emotional impact of COVID-19 outbreak in amyotrophic lateral sclerosis patients: evaluation of depression, anxiety and interoceptive awareness. Neurological Sciences. 2020; 41:2339–41.

ISSN 1695-6141