Telematic adaptation to home mechanical ventilation in patients with amyotrophic lateral sclerosis

Adaptación telemática a la ventilación mecánica domiciliaria en pacientes con esclerosis lateral amiotrófica

Respiratory involvement is a decisive factor in patient survival in amyotrophic lateral sclerosis (ALS); home mechanical ventilation (HMV) is the treatment of choice for these patients.[1,2] Although there is no consensus on the optimal moment for starting HMV,[1,3] adaptation to this treatment is always performed at the healthcare centre, most frequently on an outpatient basis and in some cases during hospitalisation.[1,3]

During the first peak of the COVID-19 pandemic, mobility and access to hospitals was greatly restricted in Spain, leading to the implementation of telemedicine in all medical specialties, including neurology.[4] In this context, from June 2020 to August 2021, we reviewed the HMV adaptation programme for patients with ALS, with a view to introducing adaptation via telemedicine. A total of 12 patients (5 men and 7 women) with a highly variable degree of functional impairment (ALSFRS-R scores of 17—43) underwent adaptation to HMV via telemedicine. During an initial in-person consultation at the multidisciplinary ALS unit, all patients were informed about the need to start HMV and had the opportunity to ask questions about the treatment.

HMV was indicated due to impaired pulmonary function in 10 patients and as an intermediate step before placement of a gastrostomy tube in the remaining 2. Ventilators were brought to the patients' homes by a company providing home respiratory care. The ventilators used were bilevel positive airway pressure (BiPAP) devices that record usage and efficacy data on a digital platform. This platform allows physicians to monitor and adjust ventilation parameters.

After the devices were delivered to patients' homes, a respiratory physiotherapist made a video call with each patient and/or their caregiver to explain the basic management of the ventilator and how to place and remove the mask. These video calls lasted 30—50 minutes (median, 42 minutes).

A second video call was made after 2 nights of use; during a structured interview, the physiotherapist enquired about any symptoms of discomfort and presence of facial pressure ulcers caused by the mask, and efficacy data were reviewed.

After the second video call, ventilation parameters had to be adjusted in 6 patients due to insufficient night-time ventilation. In these cases, additional video calls were performed every 48 hours to conduct a structured interview and review ventilation parameters, until satisfactory ventilation throughout the night was achieved. A median of 3 video calls (range, 2—5) were held per patient.

Subsequently, a month after the last video call, we reviewed usage (hours of use) and efficacy data via the digital platform. Only one patient used the ventilator for less than 4 hours/day. The efficacy of HMV was adequate in all cases. An additional video call was performed; none of the patients reported discomfort associated with HMV or facial pressure ulcers. None of the patients required an in-person consultation at the hospital for adaptation to HMV (Table 1).

Tele-monitoring of HMV is now widespread[5]; however, no official recommendations have been issued for telemedicine-based adaptation to this treatment.[6] Clinical need and the context of the pandemic led to the creation of a specific protocol. In addition to the information gathered during video calls with patients, the use of home ventilators with an integrated system for data collection and transmission is essential for obtaining objective feedback about the efficacy of ventilation. Telemedicine, defined as the provision of healthcare through communication technologies,[7] also presents ethical challenges, such as maintaining quality and safety standards, the digital divide, patient acceptance,[8] and continuity of care, all of which could weaken patient-physician relationships.[9]

Our study does not provide a sufficient level of evidence about the efficacy of telemedicine-based adaptation to HMV as compared to in-person adaptation, given the small size of our sample. However, it does serve as a starting point as it demonstrates the plausibility of adaptation via telemedicine, a particularly necessary approach in a patient group with significant mobility issues.

Funding

This study received no funding of any kind.

Conflicts of interest

The authors have no conflicts of interest to declare.

Acknowledgements

We would like to thank Anna Guillén-Solà and Montserrat Villatoro, for the care provided to all patients attended at the multidisciplinary ALS unit; Francisco Leiva, for his support in the respiratory management of patients with home mechanical ventilation; and Griselda Esteve Plantalech and Oscar García Mariñoso, for their administrative support.

DOI of original article: https://doi.org/10.1016/j.nrl.2022.01.004.

Table 1 Clinical data from patients with amyotrophic lateral sclerosis undergoing adaptation of home mechanical ventilation via telemedicine and 1-month efficacy data.

Clinical data	Pulmonary function				Baseline nocturnal oximetry				Baseline ABG FiO_2 0.21			Ventilation parameters						Ventilation efficacy			
Sex	ALSFR-R	Onset	Age (years)	BMI (kg/m^2)	VC, % pred.	VC, mL	SNIP, cm H_2O	PCF, L/min	CT90%	ODI	SpO_2, mean	pH	$PaCO_2$, mm Hg	PaO_2, mm Hg	Ventilation mode	IPAP, cm H_2O	EPAP, cm H_2O	Usage, h/day	Leakage, L/min	AHI	MV, L/min
Man	42	Bulbar	65	23.10	69	2530	38	240	4.2	4.2	93.9	7.44	34.6	95	BiPAP	8	4	8.3	5.29	3.3	7.0
Man	34	Spinal	69	28.50	108	3580	NA	440	72.0	20.5	88.6	7.40	41.0	76	BiPAP	10	4	6.7	0.22	2.7	8.0
Man	18	Bulbar	69	18.90	NA	NA	46	160	NA	NA	NA	7.40	36.4	79	BiPAP	10	4	5.5	0.74	6.1	9.9
Man	26	Spinal	54	21.40	42	1790	32	250	0.0	4.3	93.9	7.40	33.0	93	BiPAP	8	4	2.3	2.40	1.4	7.6
Man	25	Bulbar	47	20.60	54	2090	59	320	2.4	7.3	97.6	7.45	38.9	103	BiPAP	10	4	5.1	0.40	0.5	8.5
Woman	28	Bulbar	68	28.80	34	900	63	170	13.0	6.4	92.0	7.37	39.7	122	BiPAP	8	4	6.4	4.02	9.0	6.0
Woman	27	Spinal	78	18.80	50	1010	10	160	3.7	4.2	92.4	7.42	39.7	90	BiPAP	10	4	5.4	1.20	1.1	6.4
Woman	43	Bulbar	81	25.90	72	1160	90	330	1.9	3.4	93.0	7.41	37.2	79	BiPAP	8	4	5.3	0.39	10.8	4.7
Woman	24	Bulbar	77	23.00	34	720	18	200	NA	NA	NA	7.44	45.7	71	BiPAP	16	4	6.5	5.00	5.0	6.7
Woman	29	Bulbar	58	23.50	21	680	21	150	62.5	9.0	89.0	7.42	40.1	76	BiPAP	8	4	7.4	2.00	5.2	5.0
Woman	24	Spinal	70	26.90	68	1780	44	260	38.6	11.9	89.7	7.42	40.7	87	BiPAP	12	4	4.5	0.00	1.2	7.7
Woman	17	Bulbar	59	27.89	NA	NA	NA	NA	NA	NA	NA	7.47	48.9	86	BiPAP	8	4	7.2	1.00	7.3	4.0

% pred.: % predicted value; AHI: apnoea-hypopnoea index; ALSFR-R: Amyotrophic Lateral Sclerosis Functional Rating Scale; BiPAP: bilevel positive airway pressure; BMI: body mass index; CT90%: cumulative time at SaO_2 < 90%; EPAP: expiratory positive airway pressure; FiO_2: fraction of inspired oxygen; IPAP: inspiratory positive airway pressure; MV: minute ventilation; NA: not assessed; ODI: oxygen desaturation index; $PaCO_2$: partial pressure of carbon dioxide; PaO_2: partial pressure of oxygen; PCF: peak cough flow; SNIP: sniff nasal inspiratory pressure; SpO_2: pulse oximetry; VC: vital capacity.

References

1. Miller RG, Jackson CE, Kasarskis EJ, England JD, Forshew D, Johnston W, et al. Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: drug, nutritional, and respiratory therapies (an evidence-based review): report of the quality standards subcommittee of the American academy of neurology. Neurology. 2009;73:1218—26.
2. Andersen PM, Abrahams S, Borasio GD, de Carvalho M, Chio A, Van Damme P, et al. EFNS guidelines on the Clinical Management of Amyotrophic Lateral Sclerosis (MALS) — revised report of an EFNS task force. Eur J Neurol. 2012;19:360—75.
3. Farrero E, Antón A, Egea CJ, Almaraz MJ, Masa JF, Utrabo I, et al. Normativa sobre el manejo de las complicaciones respiratorias de los pacientes con enfermedad neuromuscular. Arch Bronconeumol. 2013;49:306—13.
4. Bokolo AJ. Exploring the adoption of telemedicine and virtual software for care of outpatients during and after COVID-19 pandemic. Ir J Med Sci. 2021;190:1—10.
5. Ambrosino N, Vitacca M, Dreher M, Isetta V, Montserrat JM, Tonia T, et al. Tele-monitoring of ventilator-dependent patients: a European Respiratory Society Statement. Eur Respir J. 2016;48:648—63.
6. Guía SEPAR para la teleconsulta de pacientes respiratorios. https://www.separ.es/node/1974.
7. Jiang W, Wang L, Song YL. Titration and follow-up for home noninvasive positive pressure ventilation in chronic obstructive pulmonary disease: the potential role of tele-monitoring and the Internet of things. Clin Respir J. 2021;15:705—15.
8. Ambrosino N, Fracchia C. The role of tele-medicine in patients with respiratory diseases. Expert Rev Respir Med. 2017;11:893—900.
9. Chaet D, Clearfield R, Sabin JE, Skimming K. Ethical practice in Telehealth and Telemedicine. J Gen Intern Med. 2017;32:1136—40.

A. Balañá[a,b], M.Á. Rubio[a,b], B. Bertran[a,b], J. Martínez Llorens[a,b,c,d,*]

[a] *Servicio de Neumología, Hospital del Mar, Cataluña, Spain*
[b] *Unidad Multidisciplinar de ELA, Cataluña, Spain*
[c] *CEXS, Universitat Pompeu Fabra, Cataluña, Spain*
[d] *CIBER de Enfermedades Respiratorias (CIBERES), ISC III. Madrid, Spain*

* Corresponding author.
E-mail address: JMartinezL@parcdesalutmar.cat (J. Martínez Llorens).

https://doi.org/10.1016/j.nrleng.2022.01.006

Vitamin D and myasthenia gravis

Vitamina D y miastenia gravis

Dear Editor:

I read with great interest the article published in *Neurología* under the title ''Epidemiology of myasthenia gravis in the province of Ourense (Galicia, Spain),'' which reports that 85.1% of patients diagnosed with myasthenia gravis (MG) present vitamin D deficiency (< 30 ng/mL).[1] In recent years, growing emphasis has been placed on the importance of vitamin D due to its influence in several processes, such as immunity. Vitamin D is known to play a significant role in the homeostasis of calcium and phosphorus, in the regulation of functional hormones, and in the activation of regulatory T cells and B cells.[2]

Kang et al.[3] observed that serum vitamin D levels were significantly lower in patients with MG than in healthy controls. Furthermore, an improvement in muscle weakness has been observed after cholecalciferol supplementation in patients with low vitamin D levels.[4] However, the most remarkable finding is the report of a patient showing remission of recurrent MG after administration of megadoses of vitamin D (80 000—120 000 IU/day).[5] Nevertheless, megadoses are reported to be harmful to health due to an increase in the risk of falls, and consequently the rate of fractures.[6]

A recent randomised controlled trial did not observe a clinically significant improvement in patients treated with vitamin D with respect to those receiving placebo; however, the dose administered was 800 IU/day, which is much lower than that reported in previous studies.[7]

It is currently unclear whether low serum vitamin D levels are associated with a higher risk of MG, as has been described in other such diseases as multiple sclerosis.[8] Therefore, further studies are needed on the influence of vitamin D on the onset and progression of MG. We should also mention that between 42% and 82% of patients with MG experience central fatigue; even patients in remission or with mild symptoms show mild fatigue.[9,10] As central fatigue seems not to improve with immunosuppressant treatment, it is important to establish the role of vitamin D in the fatigue perceived by these patients.[9] Therefore, I agree with Kang et al.[3] in recommending that healthcare professionals monitor vitamin D levels in patients with MG in order to maintain optimal serum concentrations.

Funding

This study received no funding of any kind.

Conflicts of interest

The authors have no conflicts of interest to declare.

DOI of original article: https://doi.org/10.1016/j.nrl.2021.11.007.