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ORIGINAL ARTICLE

Diaphragmatic pacemaker as an alternative to mechanical ventilation in patients with cervical spinal injury

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KEYWORDS
Spinal cord injuries;
Artificial respiration;
Diaphragmatic pacemaker;
Survival analysis;
Quality of life

Abstract
Objective: To verify that the diaphragmatic pacemaker is a form of respiratory support that can be used to replace a volumetric respirator in cervical spinal injury patients with cervical spinal lesion and diaphragmatic paralysis by means of its comparison with the traditional volumetric respirator.
Design: Retrospective study of a prospective database and age-matched case control study.
Setting: Intensive Care Unit and Intermediate Care Respiratory Unit, Paraplegics National Hospital, Toledo (Spain).
Patients: We collected data on all patients discharged from the Hospital with permanent respiratory support by volumetric respirator or diaphragmatic pacemaker during a follow-up period of 25 years. Personal interviews were conducted to evaluate health-related qualify of life. Comparison and survival tests were used for statistical comparisons.
Interventions: Quality of life questionnaire.
Main variables: The main variables collected were demographic data, hospital stay, mortality, family reintegration and health-related quality of life.
Results: We evaluated the clinical records of 101 patients, 37 in the pacemaker-group and 64 in the volumetric respirator-group. Our results show that ICU admission duration and hospitalization as well as family reintegration, without significant differences, with a tendency to greater survival in pacemaker patients (18.18 versus 9.67 years by the Kaplan-Meier method, $p<0.001$). However, this difference becomes non-significant ($p=0.06$) after adjustment of the groups by age. Furthermore, better quality of life was found in the same patients with pacemakers in terms of security, communication, sociability, comfort and mobility in the patients.

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Conclusions: Diaphragmatic pacemaker ventilation is an effective alternative to mechanical ventilation with similar efficacy that improve quality of life in patients with severe respiratory failure due to cervical spinal cord injury.

PALABRAS CLAVE
Lesión medular cervical;
Ventilación mecánica;
Marcapasos diafragmático;
Supervivencia;
Calidad de vida

Marcapasos diafragmático como alternativa a la ventilación mecánica en el paciente con lesión medular cervical

Resumen

Objetivo: Comprobar que el marcapasos diafragmático es una forma de soporte respiratorio que puede usarse para facilitar la retirada del respirador volumétrico en pacientes con lesión medular cervical y parálisis diafragmática, mediante su comparación con el respirador volumétrico tradicional.

Diseño: Análisis retrospectivo de una base de datos prospectiva y de tipo caso-control apareado por edad.

Ámbito: Unidad de Cuidados Intensivos y Unidad de Cuidados Intermedios Respiratorios del Hospital Nacional de Parapléjicos de Toledo.

Pacientes: Se han recogido los datos de todos los pacientes dados de alta del hospital con soporte respiratorio permanente mediante respirador volumétrico o marcapasos diafragmático con un periodo de seguimiento de 25 años y se han realizado entrevistas personales para valorar la calidad de vida relacionada con la salud. Para las comparaciones estadísticas se han usado tests de comparaciones y de supervivencia.

Intervenciones: Cuestionario de calidad de vida.

Variables de interés: Datos demográficos y clínicos, estancia hospitalaria, mortalidad, readaptación familiar y calidad de vida relacionada con la salud.

Resultados: Hemos examinado las historias clínicas de 101 pacientes, 37 con marcapasos y 64 con un respirador volumétrico. Nuestros resultados muestran tanto una duración del ingreso en UCI y de la hospitalización como una reintegración familiar sin diferencias significativas, con tendencia a una mayor supervivencia en los pacientes con marcapasos (18,18 frente a 9,67 años por el método de Kaplan-Meier, $p < 0,001$), aunque esta diferencia deja de ser estadísticamente significativa ($p = 0,06$) tras controlar los grupos por la edad. Asimismo, muestran una mejor calidad de vida en estos mismos pacientes con marcapasos en términos de seguridad, comunicación, sociabilidad, comodidad y movilidad.

Conclusiones: La ventilación con marcapasos diafragmático es un método alternativo a la ventilación mecánica con similar eficacia que facilita una mejor calidad de vida en los pacientes con lesión medular que requieren apoyo respiratorio permanente.

Introduction

Cervical spinal cord injury above metameres C3 to C5, where the nucleus of the phrenic nerve is located, results in diaphragmatic paralysis, and therefore causes severe acute respiratory failure that can prove fatal unless immediate external respiratory support is provided.[1-3] In many cases the neurological sequelae are irreversible, and the patient is unable to recover sufficient ventilation. In such situations external respiratory support in the form of permanent mechanical ventilation (MV) is needed.[4,5]

The diaphragmatic pacemaker (DP) is a form of respiratory support that can be used to allow weaning from the volumetric respirator (VR) in patients with cervical spinal lesion (SL) who suffer severe respiratory failure of neuromuscular origin.[6-9] According to some authors, this contributes to improve patient quality of life.[10,11] Since one of the main objectives in the management of patients with SL who require artificial respiration is to increase survival and improve quality of life, the use of a DP as an alternative to MV can be considered.[12]

Diaphragmatic electrostimulation or, more aptly, electric stimulation of the phrenic nerve, also known as a phrenic pacemaker, diaphragmatic pacemaker (DP) or electrophrenic respiration, consists of the induction of diaphragmatic contractions through electric stimulation of the phrenic nerve, with the purpose of producing diaphragmatic movements similar to those occurring physiologically during breathing, with a view to compensating the absence of spontaneous diaphragmatic contraction in patients of this kind. Before using the DP, the electrophysiological study of the phrenic nerve and diaphragm must show these anatomical elements to be functional, with the absence of serious airway and lung parenchymal disease.[13] Accordingly, the DP must not be applied in patients with disorders that can adversely affect these structures (tumors, vascular diseases, multiple sclerosis, amyotrophic lateral sclerosis, diabetic neuropathy, etc.). Likewise, these devices should be avoided in the presence of

respiratory tract alterations or severely impaired consciousness precluding patient cooperation.[14,15] Since DP implantation requires major thoracic surgery, and considering the aspects commented above and the potential complications involved,[16] this technology cannot be used on a transient basis and must be reserved for those patients who are not expected to improve spontaneously over the short or middle term. Thus, in practice, DP implantation is limited to patients with respiratory paralysis secondary to high cervical SL or brainstem damage and with congenital central alveolar hypoventilation (Ondine course).[17,18] The use of a DP has been shown to be able to allow total patient weaning from the respirator, though an adaptation period is required after implantation in order to improve the diaphragmatic muscle tone, which has suffered atrophy as a result of the lack of use.[15]

The first clinical evaluations of phrenic nerve electrostimulation as a method to afford respiratory support were carried out as a consequence of the poliomyelitis epidemic in 1948 in the United States. Posteriorly, and based on the pioneering work of Glenn et al. in the 1960s, more controlled application of the technique was achieved, demonstrating its usefulness in patients with severe respiratory failure due to central alveolar hypoventilation, and in 20 tetraplegic individuals with cervical spinal lesions above C3.[19,20] Following these first experiences, improvement of the technique has made it possible to permanently ventilate patients with neuromuscular diseases using this single system.[21] In the National Paraplegics Hospital of Toledo (Toledo, Spain), we introduced this type of respiratory support 25 years ago.[22]

The present study describes our experience with this technique and compares its efficacy versus the volumetric respirator (VR), evaluating outcome endpoints such as the family reintegration rate, survival, or health-related quality of life (as assessed by means of a specific questionnaire), with a view to assessing its usefulness in weaning the patient from the respirator.

Patients and methods

The study consists of two arms: (a) a retrospective cohort survey of a prospectively compiled database for clinical and patient survival variables; and (b) an interview arm for the quality of life study. The study group in turn consisted of patients with cervical spinal lesion (SL) requiring DP respiratory support, while the control group consisted of patients remaining on VR. The study was carried out in patients enrolled in the permanent respiratory support program of the National Paraplegics Hospital of Toledo over a period of 25 years.

We reviewed the case histories and respiratory management protocols of all the patients discharged from the Intensive Care Unit (ICU) with mechanical ventilation (MV) for over 90 days (294 patients). All of them came from the ICUs of other hospitals and were transferred to the ICU of our center for stabilization and inclusion in the rehabilitation program for patients with SL. Many of these patients (n=193) could be weaned from the respirator in a late phase in our Respiratory Intermediate Care Unit, reaching total respiratory autonomy, and were thus excluded from the study. Of those who required the continuation of respiratory support (n=101), some were able to abandon the respirator following the implantation of a DP (DP group, 37 patients), while others remained connected to VR indefinitely (VR group, 64 patients).

The patients in the VR group have remained connected in recent years to a portable VR (LP-10, Puritan-Bennett), with respiratory frequency, tidal volume and inspiratory time values adapted to each individual patient in order to secure blood gas parameters within normal limits. The patients in the DP group received stimuli for the simultaneous activation of both hemidiaphragms from an external generator transmitting radiofrequency (RF) energy and information to receptors implanted in the subcutaneous tissue, and which in turn were connected to transmitters adapted to both phrenic nerves through tetrapolar electrodes programmed at a respiratory frequency designed to keep the blood gas values within normal limits - with the exception of pCO_2, where values of up to 30 mmHg were tolerated as lower limit. The decision to implant a DP or leave the patient on MV depended more on the presence or absence of criteria corresponding to the protocol used in our center (cited above) than on the severity of the neurological lesion or its sequelae.

All the patients were able to communicate through short phrases - this being essential for the evaluation of quality of life. The patients on VR were able to talk on deviating the air column during the expiratory phase through the glottis, occluding the air outlet from the tracheotomy cannula without pneumoplugging using a plug or a speaking valve (Passy-Muir). The subjects in the DP group were able to talk on deviating the air column during both the inspiratory and the expiratory phase through the glottis in a more physiological manner, using a cannula without pneumoplugging or a hemi-cannula with occlusion of the orifice to facilitate air passage towards the glottis.

The following variables were recorded: patient age at the time of neurological damage, gender, cause of the spinal lesion, metameric level and degree of severity of SL as assessed using the ASIA scale.[23] Comorbidity in turn was evaluated according to the Charlson comorbidity index,[24] while the duration of stay in the ICU and the duration of stay in the hospital ward were referred to the first hospital admission episode after neurological damage until death or until 31 October 2008 (censored date in the survival study according to the Kaplan-Meier method and Cox regression), in the case of those subjects who were still alive.

Quality of life was assessed using a questionnaire in application to the patients who were still alive. The questionnaire evaluated specific aspects of health-related quality of life among patients with great disabilities and respiratory support, and is referred to as the GDRF (great disability with respiratory failure) questionnaire, based on a previous proposal[25] (annex 1). The evaluations were made by personal interview or telephone interview when the former was not possible, and involved all live patients who agreed to participate. The items were scored using a Likert scale with 5 possible answers (always; almost always; often; sometimes; never), scored from 5 (best quality of life) to 1 (poorest quality of life). A global score is thus obtained, together with a score for each of the dimensions.

The qualitative data were expressed as frequencies and percentages, while the quantitative variables were reported

as the mean ± standard deviation. The comparison of variables was carried out using nonparametric tests in the case of those parameters that did not show a normal distribution as established by the Kolmogorov-Smirnov test. Associations between qualitative variables were evaluated using the Pearson chi-squared test or the Fisher exact test. Quantitative variables were compared using the Mann-Whitney U-test or Wilcoxon test. Survival in the DP and VR groups was evaluated with the Kaplan-Meier method and multivariate Cox regression analysis, taking mortality to represent the dependent variable. In addition, in a second step, to confirm the results and after controlling for the variable "age", we repeated the univariate analysis with the Kaplan-Meier method, based on an age-matched case-control study in which the cases were drawn from the DP group and the controls from the VR group, with the aim of confirming that the difference in age - which undoubtedly introduces important bias in the interpretation of the results - is the factor that explains the difference in survival between the two groups of patients, rather than the type of respiratory support involved. The SPSS® version 15.0 statistical package for Microsoft Windows was used for the statistical analysis. In the contrast of hypotheses, the null hypothesis was rejected with an alpha error<0.05 (a p-value of under 0.05 being considered statistically significant).

Results

We evaluated the case histories of 101 patients requiring indefinite respiratory support: 37 with a diaphragmatic pacemaker (DP group) and 64 with a volumetric respirator (VR group). There were 64 males (63.37%) and 37 females (36.63%). The mean patient age at the time of the lesion was 31.13 years (26.77-35.49), with a range of 3-69 years. The cause of SL was trauma in 66 cases (65.4%) and medical

Table 1 Demographic, clinical and evolutive data according to the type of respiratory support provided

	DP	VR	p
Patients (total, 101)	37	64	
Gender			
Male	24 (64.9%)	40 (62.5%)	0.81
Female	13 (35.1%)	24 (37.5%)	
Age, mean (years)	16.22±10.64	39.76±22.15	0.0002
Cause of spinal lesion			
Trauma	25 (67.6%)	41 (64.1%)	0.72
Medical	12 (32.4%)	23 (35.9%)	
Level of spinal lesion			
B-C1	17 (45.8%)	25 (39.1%)	0.009
C2	19 (51.4%)	20 (31.2%)	
C3	1 (2.7%)	14 (21.9%)	
C4	0	4 (6.2%)	
C5	0	1 (1.6%)	
ASIA grade			
A	25 (67.6%)	45 (70.3%)	0.26
B	6 (16.2%)	5 (7.8%)	
C	4 (10.8%)	13 (20.3%)	
D	2 (5.4%)	1 (1.6%)	
Destination at first discharge			
Home	29 (78.38%)	33 (51.56%)	0.008
Other center or sociosanitary residency	8 (21.62)	31 (48.44%)	
Caregiver (data only available on 90 patients)			
Parents	26 (70.3%)	18 33.9%	0.006
Offspring	3 (8.1%)	13 (24.5%)	
Couple	1 (2.7%)	1 (1.9%)	
Others	7 (18.9%)	21 (39.7%)	
Survival			
Live	25 (67.6%)	22 (34.4%)	0.0013
Deceased	12 (32.4%)	42 (65.6%)	
Mean duration of stay in ICU (days)	109.19±61.75	84±57.54	0.021
Total duration of first admission to hospital (days)	635.3±595.8	665.8±796.3	0.51
Comorbidity (Charlson index)	0.11±0.39	0.36±0.65	0.026
Mean life expectancy (years) (Kaplan-Meier)	18.18±1.88	9.67±1.34	0.0003

ASIA grade: spinal lesion severity grade according to the American Spinal Injury Association; ICU: Intensive Care Unit.

Table 2 Multivariate Cox regression analysis to evaluate the effects of the different covariables upon patient survival

Variable	HR	95%CI	p
Type of respiratory support (DP vs VR)	0.89	0.38-2.05	0.78
Age	1.07	1.04-1.10	0.0003
Gender	1.34	0.67-2.69	0.41
Level of spinal lesion	0.83	0.53-1.29	0.41
Destination after first discharge	1.68	0.86-2.33	0.33
Charlson index	0.85	0.46-1.85	0.61

95%CI: 95% confidence interval; HR: hazard ratio.

in 35 (34.6%) The level of SL was bulbo-medullary and/or C1 in 42 patients (41.58%), C2 in 39 patients (38.61%), C3 in 15 patients (14.85%), C4 in 4 patients (3.96%), and C5 in a single patient (0.99%). The SL grade was ASIA A (complete sensory and motor SL) in 70 patients (69.31%), ASIA B (complete motor, incomplete sensory SL) in 11 patients (10.89%), ASIA C (incomplete sensory and motor without functionality SL) in 17 patients (16.83%), and ASIA D (LM complete sensory and motor with some functionality) in 3 patients (2.97%). The patient destination at discharge was home in 62 cases (68.89%) and to some other hospital or sociosanitary residency in 39 cases (31.11%). In the study of survival and on the censored date (31 October 2008), 47 patients were still alive (46.53%) while 54 had died (53.47%). Death occurred within the first 5 years after the spinal lesion in 38 patients (7 DP and 31 VR), between the sixth and tenth year in 10 (2 DP and 8 VR), and after 10 years in 6 patients (3 DP and 3 VR).

Comparison of the DP and VR groups (table 1) revealed no significant differences between them in terms of gender distribution, cause of SL, or lesion grade as determined from the ASIA scale. In contrast, there was a significant difference in relation to age (mean 16.22 vs 39.76 years; median 15 [interquartile range 25-75, 7-25] vs 39 [interquartile range 25-75, 21.5-59]). Likewise, the patients in the VR group showed more accompanying disorders as assessed with the Charlson comorbidity index (0.11 vs 0.36), greater mortality measured in percentage deaths with respect to the total in each group (32.4% vs 65.6%), destination after hospital discharge and survival (18.18 vs 9.67 years, p<0.001). On analyzing these variables by Cox regression (table 2), it was seen that after controlling for age, the difference in survival according to the type of respiratory support involved (DP vs VR) failed to reach statistical significance (p=0.78), in the same way as the Charlson comorbidity index (p=0.61) or the family reintegration rate at discharge (p=0.13).

Since the age difference between the two groups was notorious (39.76±22.15 vs 16.22±10.64 years, p<0.001) - a situation that introduces very important bias on attempting to interpret comparative survival between them - we performed a second univariate analysis according to the Kaplan-Meier method in a selection of our patients after adjusting for age. This was done by grouping pairs of patients according to age in the two groups (DP and VR), allowing us to compare 19 patients in each series grouped by pairs with the same age. Following such adjustment we found that there were no significant differences between the two groups in relation to gender (males 73.7% vs 68.4%, p=0.72), cause of SL (trauma 57.9% vs 63.2%, p=0.74), lesion level (above C2 42% vs 57.9%, p=0.71) or SL grade (complete lesion 57.9% vs 84.2%, p=0.07), destination at discharge (home 63.2% vs 57.9%, p=0.80), the type of caregiver (direct relative 73.7% vs 52.6%, p=0.35), the duration of stay in the ICU (101.05 vs 90.79 day, p=0.59) or in the hospital (88.7 vs 90.5 weeks, p=0.95), or comorbidity (Charlson index 0.14 vs 0.18, p=0.69)(for groups DP versus VR, respectively). In this series of subjects the Kaplan-Meier univariate analysis confirmed that although mean survival was longer in the DP group (19.24±4.93 years vs 12.64±4.98 years in the VR group), the difference was not statistically significant (OR=0.33, with 95%CI 0.10-1.07; p=0.06) (figs. 1 and 2).

In relation to patient quality of life, the GDRF questionnaire was completed by 41 patients (23 with DP and 18 with VR). Multiple logistic regression analysis revealed no significant differences between the two groups (DP and VR) in terms of age (p=0.57), gender (p=0.13), lesion level (p=0.15) or grade (p=0.08), the cause of SL (p=0.18), or survival (p=0.23). Only lesser comorbidity as assessed by the Charlson index was observed in the patients with DP (0.26 vs. 0.35, p=0.02). In order to avoid interpretation differences, all interviews were administered by the same author. According to the data obtained, the patients with DP reported comparatively greater quality of life in the dimensions safety (p<0.01), communication (p<0.005), sociability (p<0.001), comfort (p<0.001) and mobility (p<0.001) (table 3).

Discussion

To date, diaphragmatic pacemakers (DP) have been implanted in patients with high cervical spinal lesions (SL) in a number of countries, involving small case series.[21] The best results are obtained in children and young individuals.[14] The poor survival of patients dependent upon mechanical ventilation (MV)[26] has improved as the respiratory support techniques and care measures have developed,[27] since the need for MV is an independent factor for poor survival in patients with SL, due to the associated respiratory complications.[28,29] Gender, race or the etiology of SL exert no influence upon the survival of these patients, in contrast to age, and lesion level and grade.[26-29] According to our results, the mean life expectancy of patients with DP is greater than in those on volumetric respirators (VR), though

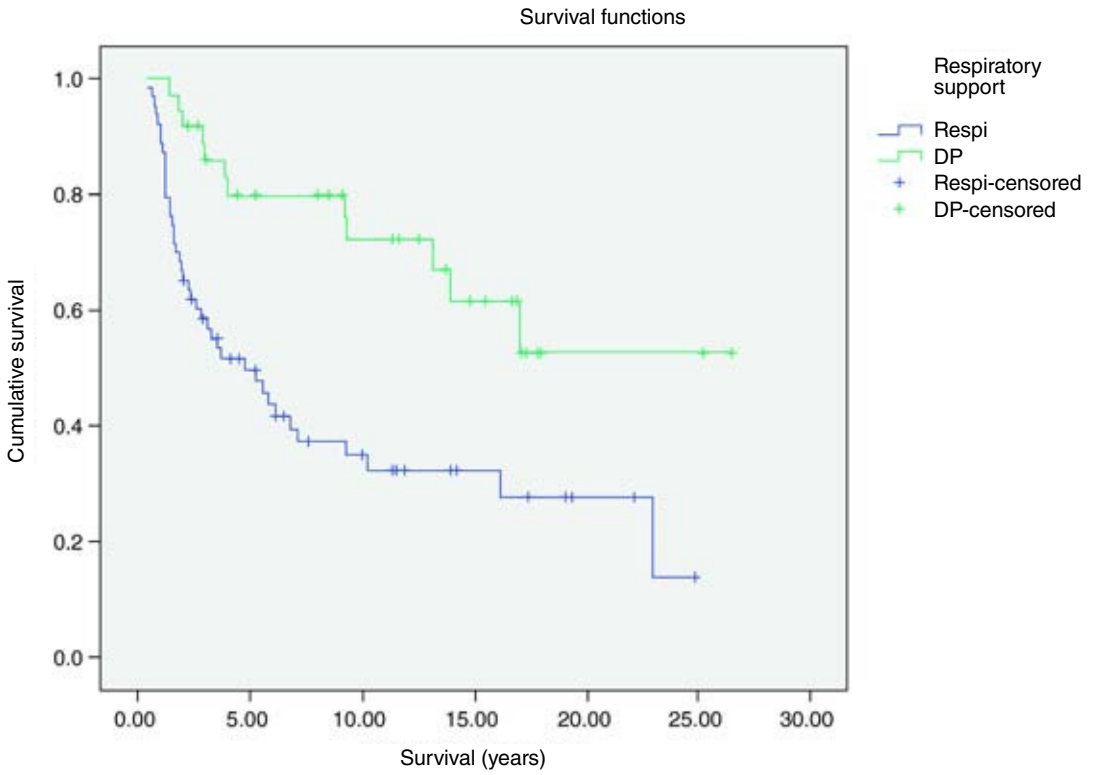

Figure 1 Comparison of the survival curves between the patients with DP and those receiving VR in the total study sample.

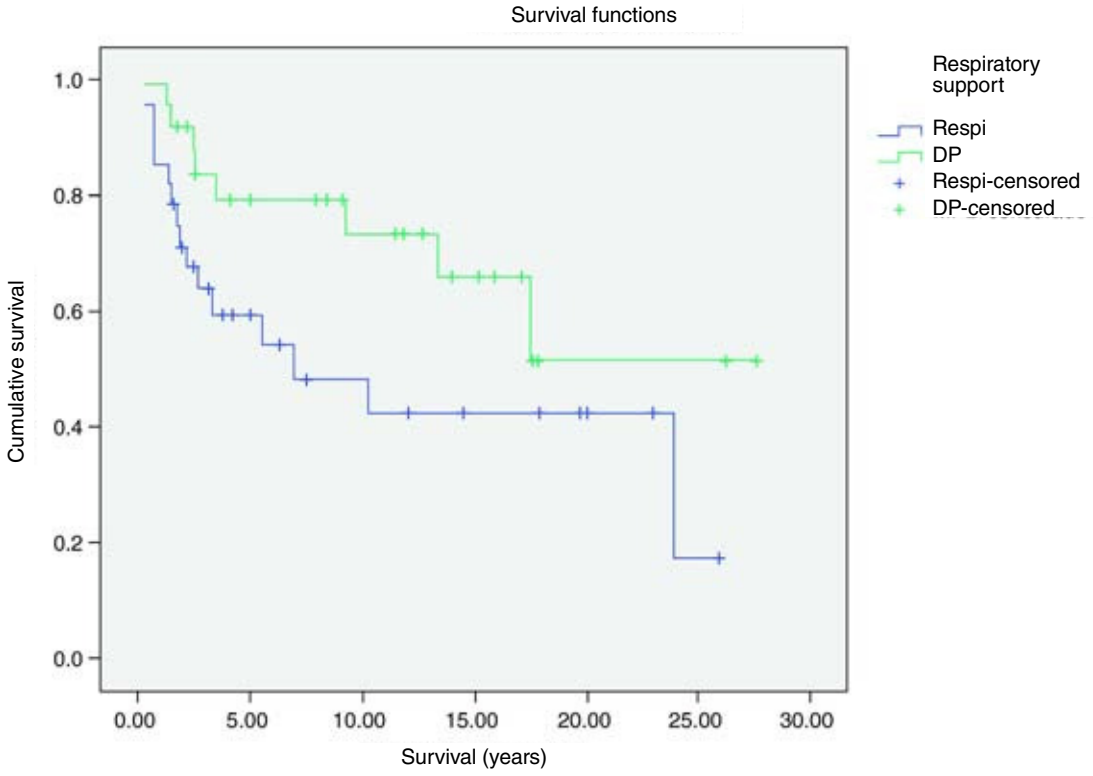

Figure 2 Comparison of the survival curves between the patients with DP and those receiving VR in the age-matched case-control group.

Table 3 Quality of life. Result of the great disability with respiratory failure (GDRF) questionnaire

	DP	VR	p
Patients (total, 41)	23 (62%)	18 (28%)	
Safety	4.13±1.14	3±1.33	< 0.01
Communication	9.17±1.15	6.89±2.82	< 0.005
External dependency	10.22±3.74	8.50±3.52	0.14
Sociability	12.17±2.87	8.39±2.61	< 0.001
Comfort	4.91±0.29	3.11±1.18	< 0.001
Mobility	3.52±1.99	14.11±6.43	< 0.001
Total criteria	64.13±7.24	44.01±11.03	< 0.001

after controlling the groups for age, the difference loses statistical significance as a result of the low statistical power of the resulting small sample size (figs. 1 and 2). In any case, it may be postulated that in view of the magnitude of the difference in mean life expectancy (18.9 versus 13.23 years), and the borderline p-value (0.06), the existence of such a difference will be confirmed in future as the number of patients enrolled in the program increases.

The patients receiving DP required a longer stay in the ICU, undoubtedly because implantation of this device requires thoracic surgery - with the resulting postoperative care and necessary diaphragm conditioning period (the diaphragm having suffered atrophy as a result of the lack of use) until optimum function is reached.[15] This conditioning period is defined as the number of days from surgical implantation of the DP to total patient weaning from VR; in our series, the mean period was 98.43±48.82 days, though we were unable to identify significant differences with respect to the total hospital stay on occasion of the first admission. Undoubtedly, the prolonged stay is also influenced by the many respiratory, hemodynamic, digestive, urological and infectious complications of the patients prior to stabilization, and the prolonged nature of the integral rehabilitation program - the ultimate objective of which is to ensure social, family and even occupational reinsertion of the patient. Regarding patient reintegration within the family setting after discharge, the global results show that the patients with DP are discharged home more often than those receiving VR (78.4% vs 51.6%, p=0.008). However, since this difference also lost statistical significance upon controlling for age, the mentioned superior family adaptability may be more attributable to the lesser age of these patients than to the type of respiratory support involved.

Health-related quality of life (HRQoL) can be defined as the subjective assessment, influenced by current health condition, of personal capacity to carry out important functions. It is the personal perception of each individual of the impact of health condition upon physical, emotional or social wellbeing.[30] Improving quality of life is one of the main objectives of the treatment of patients with SL, and as such it represents an excellent indicator of the effectiveness of a rehabilitation program.[31,32] Eliminating dependency upon the respirator is considered to improve quality of life;[12,33] as a result, it should be one of the goals of the management of our patients. It has been commented that DP allows more physiological breathing, with a lesser need for care, greater simplicity of respiratory management, less anxiety for the patients and those around them, and improved verbal communication.[5] In addition, this type of device affords greater quality of life as a result of improved portability and the absence of tubes and connections,[34] facilitating mobility, improving patient comfort and reducing costs.[21,35] In view of the above, it is considered that patient quality of life improves after weaning from MV due to the implantation of DP, though no criteria have been established for measuring such improvement and for comparing it with standard support in the form of a respirator.[8,10,18] We consider that our results offer more objective support of the above conclusions than previous studies, which only yielded estimations based on the subjective opinion of the authors.[15,36,37]

In evaluating health-related quality of life we did not administer an already validated questionnaire such as the very widely used SF-36, or the more specific instrument for home mechanical ventilation in non-tracheotomized patients,[38] since such questionnaires do not adapt to our patients (both assess mobility and walking capacity, which our patients do not have). We therefore used a questionnaire developed from previous experiences (shown in annex 1).

The limitations of this study are inherent to its design, though the study is obligatorily of a retrospective nature given the low incidence of patients with severe respiratory failure requiring respiratory support due to SL. This also explains the small sample size and the prolonged study time. Nevertheless, we feel our conclusions to be valid once the confounding factors are controlled - particularly the large age difference between the two groups, resulting from the necessary patient selection due to the clinical characteristics involved. Regarding the possible variations of the technique and technologies as a consequence of the prolonged assessment period, it must be underscored that basically neither the electrical stimulator nor the artificial ventilation mode have changed. In this context, the main modifications correspond to aesthetic and safety improvements (alarms, batteries, resistance of the materials used) rather than to basic parameters that could introduce bias in our results. On the other hand, the GDRF (great disability with respiratory failure) questionnaire has not been validated, though it may be in the future, once its feasibility has been confirmed.

Conclusions

In patients with severe respiratory failure due to high cervical spinal lesions, prolonged survival can be achieved with external respiratory support (both VR and DP). With the latter device there is a tendency towards longer patient survival, though in our series statistical significance could not be established due to the scant power of the comparisons, attributable to the small sample size involved. Likewise, a tendency towards easier family reintegration is seen, though in this case the younger age of the patients is clearly the reason. In effect, in such cases the parents are the main caregivers, with a greater capacity to attend the patients than when other types of relatives are in charge of care. Hospital resource utilization shows no differences with one support system versus the other. Our study indicates that the advantage of DP over VR consists of improved health-related quality of life for the patients with high cervical SL dependent upon external respiratory support - particularly as regards safety, communication, sociability, comfort and mobility. We therefore consider that diaphragmatic pacemakers may be the respiratory support technique of choice in selected patients.

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Annex 1. Quality of life perception in patients presenting great disability with respiratory failure (GDRF questionnaire)

	Always	Almost always	Often	Sometimes	Never
Having others see me connected to a respirator/ DP bothers me a lot	1	2	3	4	5
The noise of the respirator/ DP bothers me even for sleeping	1	2	3	4	5
The respirator/ DP gives me confidence that it will never fail	5	4	3	2	1
With the respirator/ DP I can speak and communicate perfectly	5	4	3	2	1
I can be alone for at least an hour	5	4	3	2	1
I need someone (nurse, assistant or others) to care for me	1	2	3	4	5
I can speak by telephone without help	5	4	3	2	1
I can use the computer without help	5	4	3	2	1
I can leave the room	5	4	3	2	1
I can leave the home / residency	5	4	3	2	1
I can leave to go on a trip	5	4	3	2	1
I can take part in social events: meals, weddings, parties, etc.	5	4	3	2	1
I can go out to the cinema, see a basketball game, etc.	5	4	3	2	1
I can study official courses: high school, university, etc.	5	4	3	2	1
I can do some kind of paid work	5	4	3	2	1

Conflict of interest

The authors declare no conflict of interest.

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