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CLINICAL CASE

Diaphragm pacing in a pediatric patient with Acquired Central Hypoventilation syndrome

Marcapasos diafragmático en paciente pediátrico con síndrome de Hipoventilación Central Adquirido

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What do we know about the subject matter of this study?

The diaphragmatic pacemaker allows reducing or eliminate the need for mechanical ventilation in patients with central hypoventilation. Most published cases to date are related to high-level spinal cord injury and congenital central hypoventilation syndrome.

What does this study contribute to what is already known?

This study presents a detailed description of the implantation of a diaphragmatic pacemaker in a pediatric patient with acquired central hypoventilation syndrome and supports the fact that it is a feasible and potentially safe option in this group of patients.

Abstract

Diaphragmatic pacemaker is a device that reduces or eliminates the need of mechanical ventilation in patients with chronic respiratory failure who keep the phrenic nerve-diaphragm axis intact, as long as they do not present intrinsic lung disease. Although its implantation has been practiced for decades, its use is not widespread and to date, there is little published literature about it, mostly related to high spinal cord injury and congenital central hypoventilation syndrome. **Objective:** To describe an experience of diaphragmatic pacemaker implantation in a pediatric patient with acquired central hypoventilation syndrome. **Clinical Case:** Female patient with central hypoventilation syndrome secondary to ischemic brainstem lesion as a result of ventriculoperitoneal shunt malfunction. For this reason, for 5 years she was supported by inpatient mechanical ventilation. At 7 years of age, a diaphragmatic pacemaker was implanted by thoracoscopic surgery, which allowed, after a period of rehabilitation and respiratory conditioning, mechanical ventilation withdrawal, and hospital discharge. **Conclusions:** Diaphragmatic pacemaker is a feasible, potentially safe, and cost-effective option for decreasing or eliminating mechanical ventilation dependence and improve life quality in patients with acquired central hypoventilation syndrome.

Keywords:
Diaphragm pacing; thoracoscopic surgery; central hypoventilation syndrome; respiratory insufficiency

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Introduction

Central hypoventilation syndrome (CHS) is a rare condition that leads to chronic respiratory failure. Its etiology lies in the disconnection between the respiratory centers of the brainstem and the phrenic nerve, which alters the autonomic and respiratory regulation. This syndrome can be idiopathic as in the case of congenital CHS (Ondine's disease) or acquired due to brainstem infarctions, tumors, surgical trauma, hemorrhage, or meningoencephalitis. The management of these patients requires mechanical ventilation (MV), in some cases 24 hours a day and in other cases exclusively during sleep[1,2].

In 1966, Glenn was the first to describe the use of the diaphragmatic pacemaker (DP) and later Hunt developed a modified method for use in children, now allowing it implantation as early as 9 months[1,3-5]. This device generates an electrical current to the phrenic nerve, contracting the diaphragm. It consists of a radiofrequency transmitter, an antenna, a receiver, and an electrode[6] (figures 1 and 2). The phrenic nerve stimulation can eliminate or decrease the need for MV in patients with chronic respiratory failure who have an intact phrenic nerve-diaphragm axis, as long as they do not present intrinsic lung disease or obesity[1,5,7,8].

The DP electrode can be implanted through the neck or chest. When using the chest rout, it can be through open surgery or through thoracoscopy[1] which is a relatively uncommon technique. In 1998, Shaul first documented the use of this method[9] and later, he reported a series of 9 patients in which he described complications such as atelectasis, pneumonia, pneumothorax, bradycardia, and the need for reinstallation[10]. Recently, Nicholson reported a series of 18 children with congenital CHS, finding a lower rate of complications[5].

Although DP implantation has been practiced for decades, it has not been widely used and there is still much work to be done in innovation[8]. To date, there is little published literature and most of it is related to high-level spinal cord injury and congenital CHS. The objective of this study is to describe a successful experience of DP implantation in a pediatric patient with acquired CHS.

Clinical Case

Female patient, currently 9 years old, with congenital hydrocephalus and corpus callosum hypoplasia, who underwent ventriculoperitoneal shunt placement in the neonatal period. At the age of 2, she was admitted to our hospital with clinical signs of brain herniation due to shunt malfunction. After this event, she presented with loss of automatic control of breathing, MV dependence and swallowing dysfunction, which lead to the performance of tracheostomy and gastrostomy. During the course of the condition, we found dysmetria and partial involvement of cranial nerves from V to XII, mild developmental delay, and epilepsy.

Brain magnetic resonance imaging showed atrophy of both cerebellar hemispheres, the upper vermis, and the brainstem, mainly in the dorsal region of the medulla oblongata. The patient was managed with in-hospital MV for 5 years, tolerating periods of rest of variable duration (3 to 6 hours) while awake, situation where she maintained voluntary control of breathing, which allowed clinical evaluation of the phrenic nerve-diaphragm axis integrity.

At the age of 7, DP was placed through thoracoscopy and under general anesthesia with single-lung ventilation technique. We accessed to both sides of the chest cavity at different times. The phrenic nerve was dissected in the area adjacent to the pulmonary hilum when the right side was operated on (between T5 and T6) and over the upper edge of the pericardium when the left phrenic nerve was operated on. The electrode was then inserted into the chest cavity, where the receiver was also finally placed, and passed under each nerve attaching it to the surrounding tissues. Subsequently, we created a tunnel in the subcutaneous tissue for the electrode cable, a pocket to place the receiver (figure 3) and then we checked that the stimulation was correct. There were no intraoperative or postoperative complications.

Before the surgery, the patient participated in a 3-month respiratory rehabilitation program that included conventional respiratory kinesiotherapy, respiratory muscle training, and cardiorespiratory training. Respiratory muscle training was performed with a threshold inspiratory trainer adapted to the tracheostomy tube. Once the maximum inspiratory pressure was assessed, training was performed with 30% inspiratory load for 15 minutes daily, in intervals of 3-minute workout and 2-minute rest, 5 days a week. At the same time, cardiopulmonary training was performed using a stationary bicycle, twice a week for 30 minutes a day, in intervals of 10-minute workout and 5-minute rest.

After installing the DP, the patient was exclusively on MV for 30 days. The pacemaker setting was based on the thoracic expansion visualization, ventilometry, arterial blood gases, capnography, and SpO2. First, the working amplitude for each hemidiaphragm was established between the lower threshold, corresponding to the minimum stimulus that generated thoracic expansion, and the upper threshold, corresponding to the stimulus that did not generate a significant change

in expansion. Subsequently, the amplitude was adjusted between these values, establishing as target tidal volume, the one the patient moved in MV. To set the respiratory frequency, the one used in MV was used initially and then adjusted according to arterial gases and capnography.

In parallel, a protocol was established for MV weaning, starting the use of the device for 1 hour per day the first week and 2 hours per day the second week. Subsequently, it was increased in ranges of 2 hours per week until reaching 14 hours per day. This objective was achieved at 16 weeks post-surgery and remained so for 6 weeks. After this period, the use of nocturnal MV was discontinued and the pacemaker was used only.

After 2 years of follow-up, the patient was discharged from hospital, maintained with a tracheostomy tube, phonation valve, and without complications in her evolution.

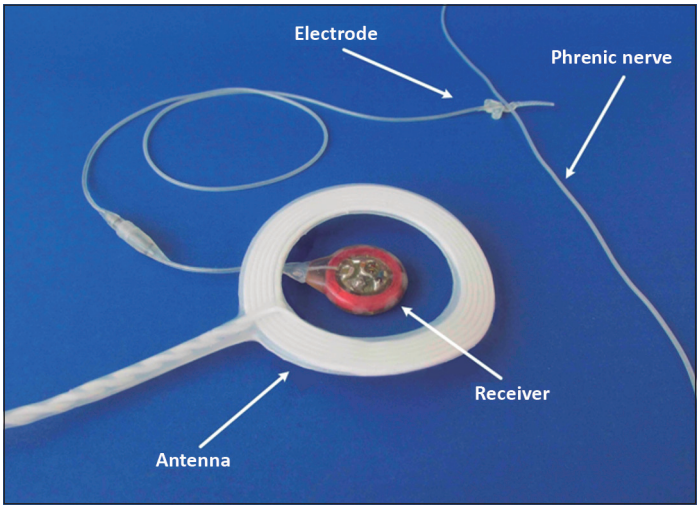

Figure 1. Components of the diaphragmatic pacemaker and schematization of the contact of the electrode with phrenic nerve.

Discussion

In this report, we highlight that the use of a DP allowed a pediatric patient with acquired CHS to be weaned from MV and her subsequent discharge, which has improved her quality of life, decreased MV-associated infections, and reduced costs of care.

One of the main goals of DP placement is to improve the quality of life. Romero-Ganuza applied questionnaires to assess the quality of life in patients with spinal cord injury, finding that those with a DP compared with those who remained on MV, perceived significantly better quality of life in the areas of safety, communication, sociability, comfort, and mobility. The same study reports that 78.4% of patients who received a DP were discharged from hospital, as opposed to 51.6% of the MV group[4]. In our case, no instruments were applied to evaluate the quality of life due to the age of the patient, however, after 5 years of hospital stay, she was discharged and included into family life, also she improved her mobility once she was weaned from MV. All of which suggest a better quality of life.

The DP can decrease the incidence of MV-associated respiratory infections, as demonstrated in the Hirschfeld study, which found that in patients with DP, the frequency of respiratory tract infections significantly decreases[12]. The patient in this case following device placement and hospital discharge has remained infection-free.

Several studies suggest that using DP reduces long-term costs[1,2,12]. Hirschfeld found that the decrease in respiratory infections and the reduced need for human and technological resources compared with MV reduces costs significantly[12]. It is difficult to say at this point that in our case the DP placement reduced costs,

Figure 2. Radiofrequency transmitter.

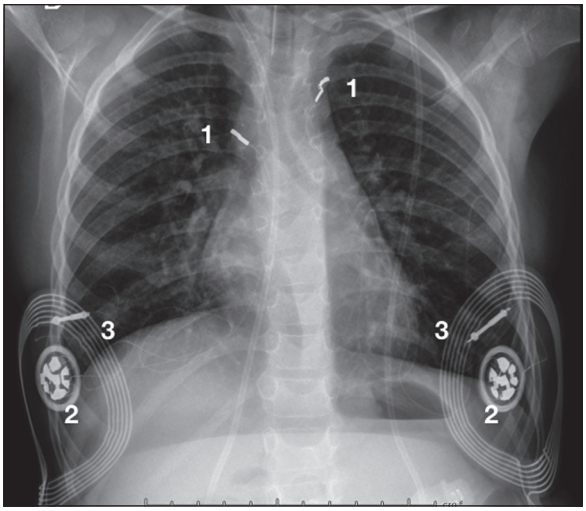

Figure 3. Postero-anterior chest x ray view: (1) Electrode in contact with bilateral phrenic nerve (2) Receiver placed subcutaneously on both sides. (3) Pacemaker antenna attached to the skin on both sides.

however, hospital discharge means a significant reduction in future costs.

Romero reports that after the DP implantation, the average time to wean from MV is 47.33 days (31-96)[11] and Hirschfeld, 51.06 days (30 - 196)[12]. Both studies were conducted in patients with quadriplegia due to high spinal cord injury. Nicholson, in his study of patients with congenital CHS, reports that the average time was 5.3 months (159 days), with an interval of 2.7-9.7 months[5] which is very similar to that of our patient (154 days). It is worth to mention that in patients who can maintain part of their work of breathing, the adaptation time is longer, as is the case with CHS. This difference is probably related to age.

It has been reported that patients with DP frequently develop upper airway obstruction, so the removal of the tracheostomy tube may mean complications such as obstructive sleep apnea[13]. Diep reviewed 15 patients with congenital CHS and tracheostomy, finding that 11 patients had their tube successfully removed after pacemaker placement, in 12.2 months on average (0.6 - 40.6)[14]. In our case, after 24 months of post-surgery follow-up, the tracheostomy tube has not yet been removed due to developmental delay and swallowing dysfunction, among other factors.

Conclusions

This report, and the review of the literature, suggest that DP implantation is a feasible, potentially safe, and cost-effective option to decrease or eliminate dependence on mechanical ventilation and improve quality of life in patients with acquired CHS.

Ethical Responsibilities

Human Beings and animals protection: Disclosure the authors state that the procedures were followed according to the Declaration of Helsinki and the World Medical Association regarding human experimentation developed for the medical community.

Data confidentiality: The authors state that they have followed the protocols of their Center and Local regulations on the publication of patient data.

Rights to privacy and informed consent: The authors have obtained the informed consent of the patients and/or subjects referred to in the article. This document is in the possession of the correspondence author.

Conflicts of Interest

Authors declare no conflict of interest regarding the present study.

Financial Disclosure

Authors state that no economic support has been associated with the present study.

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References

1. Ducko CT. Clinical advances in diaphragm pacing. Innovations (Phila). 2011;6(5):289-97.
2. Sieg EP, Payne RA, Hazard S, Rizk E. Evaluating the evidence: is phrenic nerve stimulation a safe and effective tool for decreasing ventilator dependence in patients with high cervical spinal cord injuries and central hypoventilation? Childs Nerv Syst. 2016;32(6):1033-8.
3. Ali A, Flageole H. Diaphragmatic pacing for the treatment of congenital central alveolar hypoventilation syndrome. J Pediatr Surg. 2008;43(5):792-6.
4. Romero-Ganuza FJ, Gambarrutta-Malfatti C, Diez de la Lastra-Buigues E, et al. Diaphragmatic pacemaker as an alternative to mechanical ventilation in patients with cervical spinal injury. Med Intensiva. 2011;35(1):13-21.
5. Nicholson KJ, Nosanov LB, Bowen KA, et al. Thoracoscopic placement of phrenic nerve pacers for diaphragm pacing in congenital central hypoventilation syndrome. J Pediatr Surg. 2015;50(1):78-81.
6. Abdunnur SV, Kim DH. Phrenic Nerve Stimulation: Technology and Clinical Applications. Prog Neurol Surg. 2016;29:64-75.
7. Le Pimpec-Barthes F, Legras A, Arame A, et al. Diaphragm pacing: the state of the art. J Thorac Dis. 2016 Apr;8(Suppl 4):S376-86.
8. Khong P, Lazzaro A, Mobbs R. Phrenic nerve stimulation: the Australian experience. J Clin Neurosci. 2010;17(2):205-8.
9. Shaul DB, McComb JG, Keens TG. Thoracoscopic placement of phrenic nerve electrodes for diaphragm pacing. Pediatr Endosurg Innov Tech. 1998;2:101-5.
10. Shaul DB, Danielson PD, McComb JG, Keens TG Thoracoscopic placement of phrenic nerve electrodes for diaphragmatic pacing in children. J Pediatr Surg. 2002;37(7):974-8.
11. Romero FJ, Gambarrutta C, Garcia-Forcada A, et al. Long-term evaluation of phrenic nerve pacing for respiratory failure due to high cervical spinal cord injury. Spinal Cord. 2012;50(12):895-8.
12. Hirschfeld S, Exner G, Luukkaala T, Baer GA. Mechanical ventilation or phrenic nerve stimulation for treatment of spinal cord injury induced respiratory insufficiency. Spinal Cord. 2008;46(11):738-42.
13. Wang A, Kun S, Diep B, et al. Obstructive sleep apnea in patients with congenital central hypoventilation syndrome ventilated by diaphragm pacing without tracheostomy. J Clin Sleep Med. 2018;14(2):261-4.
14. Diep B, Wang A, Kun S, et al. Diaphragm pacing without tracheostomy in congenital central hypoventilation syndrome patients. Respiration. 2015;89(6):534-8