Diaphragmatic pacing: unusual indication with successful application

Marca-passo diafragmático: indicação incomum, aplicação bem-sucedida

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To the Editor:

Syringomyelia is the development of a fluid-filled cavity or syrinx within the spinal cord. Hydromyelia is a dilatation of the central canal by cerebrospinal fluid and can be included within the definition of syringomyelia.[1]

A 20-year-old male patient was referred to the Department of Thoracic Surgery of the *Hospital Sírio Libanês* as a candidate for diaphragmatic pacing (DP) with phrenic nerve stimulation, because he had chronic apnea due to syringomyelia and had been dependent on mechanical ventilation for the last 5 years, with repeated pulmonary infections and prolonged hospitalizations. At referral, his clinical condition was good, and his phrenic nerve conduction, as evaluated by electromyography, was normal.

The patient underwent a bilateral anterior third-space mini-thoracotomy, with double-lumen intubation. The electrodes were placed underneath the phrenic nerves (direct contact between the nerves and the exposed metal) and fixed to the pericardium using a 4-0 nonabsorbable suture.

A subcutaneous pocket was created for receiver placement near the lower costal margin, where the electrode leads entered the thorax through an intercostal space. Two identical devices were implanted, one on each side, during the same procedure. After the receivers had been connected to the electrodes, the whole system was tested, with the help of an engineer who was present in the operating room. The incisions were closed in layers and a 14F chest tube (with Heimlich valve) was placed in position.

The postoperative recovery period was uneventful, and diaphragm conditioning was initiated two weeks after implantation.

Radiofrequency signals, generated by a battery-powered transmitter (Figure 1a), were sent from an external antenna (Figure 1a), fixed to the implanted receivers (Figure 1b), which convert the radio signals in electrical impulses, causing diaphragmatic contraction.

To avoid fatigue, DP was initiated at a frequency of 15 Hz for 15 min during each waking hour in the first week, with increases of 15 min/week, as tolerated by the patient. After 30 days of DP, the patient was discharged in good clinical condition, being submitted to 60 min of continuous DP every day.

At this writing, after 90 days of DP, the patient had been submitted to continuous pacing for approximately 10 h every day, requiring mechanical ventilation especially when sleeping. No muscle fatigue had been reported. The quality of speech had improved, and the tracheal stoma had been maintained in place with a 6.0-mm diameter cannula.

In 1972, Glenn et al.[2] introduced DP with phrenic nerve stimulation for quadriplegic patients on ventilatory support. Although the procedure was successful, increasing experience identified a more physiological method of DP in quadriplegic patients using simultaneous and continuous pacing of both hemidiaphragms. In 1984, the experience of ventilation in five quadriplegic patients who had been submitted to pacing for 11 to 33 months after diaphragm conditioning was reported.[3] The authors obtained promising results, demonstrating that 24-h bilateral pacing of the conditioned diaphragm was feasible.

In quadriplegic patients and in those with congenital central hypoventilation syndrome (CCHS), DP with phrenic nerve stimulation can improve quality of life by eliminating the dependence on a ventilator. The damage to the central nervous system must be above the second or third cervical level, since DP is possible only if the nerve-cell bodies are viable (located in the anterior horns at C3-C5).[3,4] In contrast, trauma to the mid-cervical spine (at C3, C4, or C5) usually precludes the use of DP. Diseases of the lower motor neurons or of the

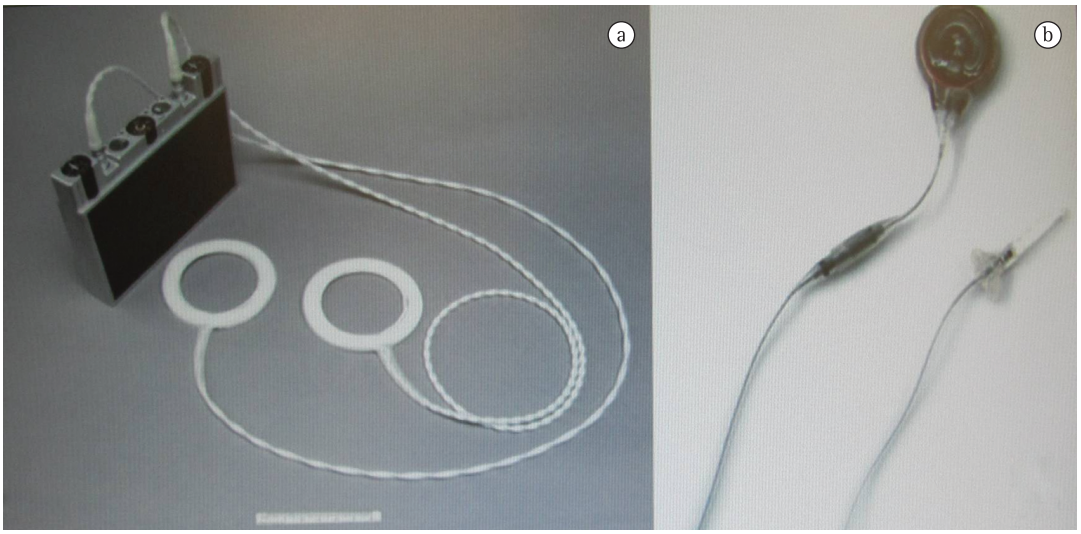

Figure 1 - Device for phrenic nerve stimulation: external parts (in a) and internal parts (in b).

anterior horn cells, such as polio or amyotrophic lateral sclerosis, have been considered contraindications to DP for the same reason.[4] Patients with CCHS are excellent candidates. Some of them hypoventilate only during sleep and can therefore utilize DP at night, rendering them ventilator-free. The ideal candidates for DP with phrenic nerve stimulation are those with complete upper cervical spine injuries (at C1 or C2) that lead to apnea, those with certain types of central sleep apnea or CCHS, and those with brainstem tumors, infarction, or strokes associated with CCHS.[5,6]

The electrodes used in DP can be placed adjacent to the phrenic nerve, either in the neck or in the chest. The cervical approach is easier, although some problems might occur, such as local infections due to previous tracheostomy adjacent to the incision, loss of conduction because of the phrenic branch at the cervical level, and rhythmic motions of upper extremities by functioning portions of the brachial plexus. Thoracic approaches include thoracotomy, thoracoscopy, or robotics. In our patient, we performed a bilateral mini-thoracotomy, because he presented with recurrent pneumonias, and decortication was necessary for phrenic nerve dissection. In this setting, the phrenic nerve is located along the mediastinum, lying on both sides laterally to the pericardium. It is important to keep a 2-3 mm margin of intact perineural tissue surrounding the nerve to preserve its blood supply.[7]

Conditioning of the diaphragm requires a gradually increasing duration of pacing, and the patient should be carefully monitored during conditioning.[7-9] Fluoroscopy can be used to determine the maximal diaphragm excursion.[4,6,7]

The selection of the appropriate DP parameters is dependent on the underlying reason for requiring ventilatory support. Patients with quadriplegia usually have significant disuse atrophy of the diaphragm, and the duration of DP should therefore be increased by only 3-5 min per day, in order to avoid fatigue.[10] In one study, 12 patients with quadriplegia and spinal cord injury were submitted to DP electrode implantation, and 6 of those patients continued DP over the long-term, with a mean duration of 13.7 years.[6]

The use of DP can provide various advantages to patients with respiratory insufficiency. Such advantages include a significant reduction in the number of upper airway infections and episodes of pneumonia; lower individual costs for airway management equipment; improvement in quality of life and speech capability; avoidance of tracheostomy decannulation, and ventilator weaning; and improved educational and employment opportunities.

The long-term success of DP depends on good selection criteria, based on pre-operative evaluations, as well as on the education of family members/caregivers and careful monitoring by the medical team.

We conclude that, in carefully selected patients, DP is clinically feasible with no apparent clinical or physiological detriment. Patients in whom long-term DP is successful demonstrate social independence and better survival. Although the potential patient pool is limited, DP offers numerous advantages when compared with ventilator systems, primarily better quality of life.

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