Case Report

Phrenic Nerve Stimulation for Diaphragmatic Pacing in a Patient with High Cervical Spinal Cord Injury

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Background: *Phrenic nerve stimulation is a therapeutic option for patients with central hypoventilation syndrome due to brain stem and high cervical spinal cord dysfunction.*
Case Report: *A 28-year old woman with high cervical spinal cord injury at the level of C2 had chronic central hypoventilation syndrome, requiring long-term use of a home ventilator. Preoperative end tidal CO_2 and tidal volume during spontaneous breathing indicated hypoventilation syndrome. Bilateral phrenic nerve stimulation for diaphragmatic pacing was performed with spinal cord stimulators used for chronic pain. The end tidal CO_2 pressure ($ETCO_2$), tidal volume, and spontaneous breathing time have improved up to 29 months of postoperative follow-up period.*
Conclusion: *Phrenic nerve stimulation for diaphragmatic pacing can reduce all-time requirement of ventilatory support in patients with high cervical spinal cord injury.*

Keywords: *Central hypoventilation syndrome, Phrenic nerve stimulation*

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Some patients with central hypoventilation syndrome caused by brain stem or high cervical spinal cord lesion require all-time ventilatory support due to insufficiently spontaneous respiration. Phrenic nerve stimulation for diaphragmatic pacing is a therapeutic option to reduce the need of a respirator[1-3].

Because a lack of the specific device of phrenic nerve stimulation for diaphragmatic pacing in Thailand, the authors adapted to use spinal cord stimulator for the operation in a patient with central hypoventilation syndrome caused by high cervical spinal cord injury[4,5].

Case Report

A 28-year-old woman had a car accident five years ago. She was diagnosed as unstable fracture of the axis with cervical spinal cord injury. An occiput-C1-C2 fusion was performed. She had respiratory insufficiency requiring long-term ventilator use, alternating with spontaneous breathing. Thereafter, the patient could breathe spontaneously and be discharged.

One year later, she developed spontaneous peritonitis caused by small bowel gangrene necessitating surgical resection and end-to-end anastomosis. After the operation, the patient could not breathe spontaneously and required all-time home ventilator probably due to weakness of the incised abdominal muscle, which is a group of accessory muscles for respiration. Subsequently, the authors decided to use phrenic nerve stimulation to restore respiration for the patient. Preoperative spontaneous tidal volume (TV) was less than 50 ml. End tidal CO_2 ($ETCO_2$) during 1-minute spontaneous breathing was progressively elevated more than 50 mmHg with symptoms of hypoventilation and hypercarbia.

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Since the specific device for phrenic nerve stimulation was not available in Thailand, the patient was operated by using the surgical technique, phrenic nerve stimulation for diaphragmatic pacing with a spinal cord stimulator[4,5], innovated by Taira T et al. The cervical and chest incisions (Fig. 1) and 1-cm nerve sheath exposure were demonstrated (Fig. 2). The phrenic nerve was identified with an intraoperative electrical nerve stimulator. Muscle relaxant was not used to observe contractions of the ipsilateral diaphragm and corresponding chest movement. A quadripolar lead-type electrode for deep brain stimulation (DBS) was placed obliquely along the site of exposed nerve and fixed to surrounding connective tissue (Fig. 3). A stimulation device (Itrel-3-Medtronic used for spinal cord stimulator in patient with chronic intractable pain) was implanted into the subcutaneous pocket performed on the anterior chest wall (Fig. 4). The electrode and stimulation device were connected together. The contralateral side was operated in the same manner.

The devices were used in cyclic mode (on 2 seconds, off 3 seconds, soft start 1 second), pulse width 150 μsec, frequency 21 Hz. The amplitude (voltage) was gradually adjusted to achieve the optimal setting (3.3 v on the right side, 3.7 v on the left side). This brought about maximal tidal volume, longer time, and absent adverse effect. The post-operative parameters were improved ($ETCO_2$ 35-45 mmHg, TV 300-350 ml). Postoperative radiographs revealed good position of the stimulation devices and electrodes (Fig. 5). Realtime fluoroscopic study of the chest during turning on the device showed movement of the ipsilateral diaphragm (Fig. 6).

Twenty-nine months postoperatively, she could breathe approximately 45-75 minutes on each side. Total spontaneous breathing time was 5-10 hours per day depending on her demand and daily activity.

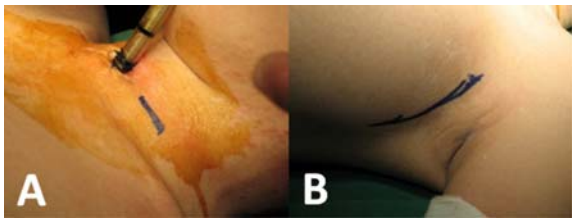

Fig. 1 (A) A transverse cervical incision was rostral to the clavicle across the posterior border of the sternocleidomas toid muscle. (B) A chest incision was located at the anterior axillary line in a curve fashion resulting in a better cosmetic outcome

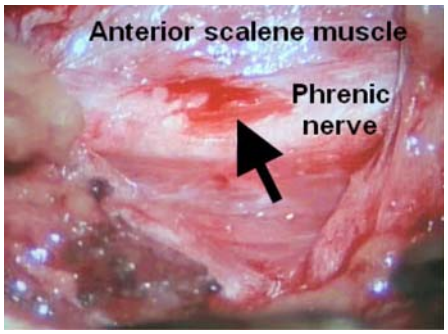

Fig. 2 A 1-cm length opening of the nerve sheath (arrow)

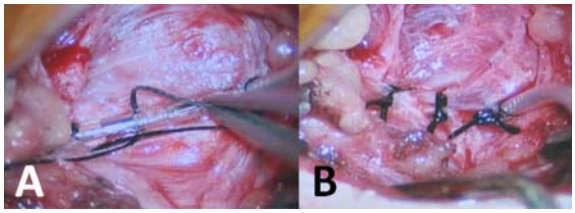

Fig. 3 (A) A quadripolar lead-type DBS electrode was placed obliquely along the phrenic nerve. (B) The electrode was fixed to the adjacent tissue

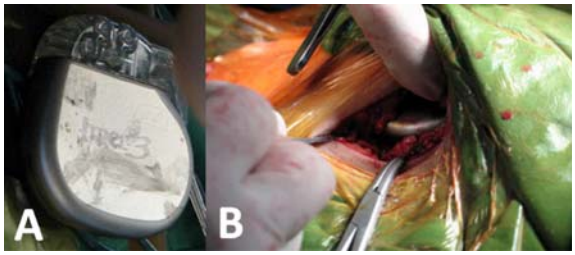

Fig. 4 (A) A stimulation device (Medtronic-Itrel 3). (B) The device was implanted into the subcutaneous pocket on the chest wall

Discussion

Phrenic nerve stimulation for diaphragmatic pacing is an alternative option to restore the respiration in patients with central hypoventilation syndrome caused by high cervical spinal cord or brain stem lesions[6,7]. An animal study of electrophrenic respiration was proposed in 1948[8]. There were many publications and worldwide uses of the operation[9]. Glenn et al used radiofrequency electrophrenic respiration in four patients with central hypoventilation. They reported an effectiveness of one-sided phrenic nerve stimulation for 8-10 hours nightly. They recommended

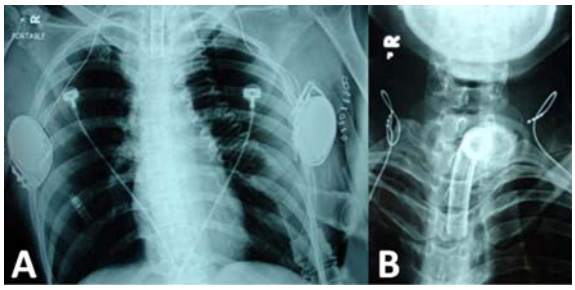

Fig. 5 Postoperative radiographs of the stimulator devices (A) and electrodes (B)

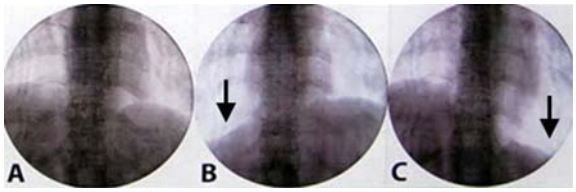

Fig. 6 (A) Normal position of the both sides of the diaphragm during the stimulation device was being turned off. (B) Downward movement of the right diaphragm during the right-sided stimulator was being turned on. (C) Turned on left-sided stimulator with concurrent downward movement of the left diaphragm

long-term electrophrenic respiration only for patients who need chronic ventilatory assistance with normal responses of phrenic nerves and diaphragm to electrical stimulation, and normal lung[2]. Prolonged stimulation of unilateral phrenic nerve resulted in fatigue of the stimulated diaphragm[12]. Therefore, the authors turned on only one side of the stimulator alternately every 60-75 minutes to avoid fatigue of the diaphragm. Continuous ventilatory support by bilateral simultaneous phrenic nerve stimulation was achieved successfully in quadriplegic patients[1,10,11]. However, most of the reports required thoracotomy to implant the electrodes in the mediastinum[1,10,12]. There was a use of thoracoscopic implantation of the phrenic nerve electrode to reduce morbidity of thoracotomy[13]. Taira et al showed good long-term efficacy of phrenic nerve stimulation for diaphragmatic pacing by using spinal cord stimulator in six patients with high cervical spinal cord or brain stem dysfunction[4,5]. Recently, intramuscular diaphragmatic pacing by laparoscopic surgery was performed in patients with cervical spinal cord injury and chronic respiratory insufficiency[14-16].

The presented patient could breathe spontaneously for many hours per day and all-time requirement of the ventilator apparently reduced. Minor adverse effects, including undesired local muscle contraction, spastic movement of the ipsilateral shoulder and upper extremity occurred sometimes. These adverse effects could be corrected by adjusting some parameters of the device setting. Good ipsilateral diaphragmatic movement while turning on the device was demonstrated with real-time fluoroscopy, as purposed by McCauley et al[17]. Of 29 months follow-up period, the device was persistently effective. Many reports also showed long-term effectiveness of the operation[2-5].

Currently, this technique is a new option for patients with central hypoventilation syndrome caused by various etiologies. Postoperative long-term follow-up and adjustment of the device setting are very crucial to achieve the maximal benefit for the patient.

Conclusion

Phrenic nerve stimulation using spinal cord stimulator is effective and can reduce all-time requirement of ventilatory support in patients with central hypoventilation syndrome caused by high cervical spinal cord injury. However, long-term follow-up and adjustment of the stimulator setting are necessary to achieve the maximal benefit.

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การผ่าตัดฝังอุปกรณ์กระตุ้นเส้นประสาทฟรีนิคเพื่อช่วยการทำงานของกะบังลมปอดในผู้ป่วยหนึ่งราย ซึ่งมีการบาดเจ็บของไขสันหลังส่วนคอระดับสูง

บรรพต สิทธินามสุวรรณ, ศรัณย์ นันทอารี

***ภูมิหลัง**: การผ่าตัดกระตุ้นเส้นประสาทฟรีนิคเป็นทางเลือกหนึ่งในการรักษาสำหรับผู้ป่วย ที่มีภาวะการหายใจไม่เพียงพอ ซึ่งเกิดจากระบบประสาทส่วนกลางอัน มีสาเหตุมาจากความผิดปกติของก้านสมอง หรือ ไขสันหลังส่วนคอระดับสูง*

***รายงานผู้ป่วย**: ผู้ป่วยหญิงอายุ 28 ปีที่มีไขสันหลังบาดเจ็บที่ระดับกระดูกสันหลังส่วนคอระดับที่ 2 มีภาวะการหายใจไม่เพียงพอเรื้อรังซึ่งมีสาเหตุจากระบบประสาทส่วนกลาง ทำให้ต้องการเครื่องช่วยหายใจตลอดเวลาในขณะอยู่บ้านมาเป็นระยะเวลานาน ค่าความดันของก๊าซคาร์บอนไดออกไซด์ในลมหายใจออกและปริมาตรของลมหายใจเข้าในขณะที่ผู้ป่วยหายใจเองก่อนผ่าตัดบ่งชี้ว่ามีภาวะการหายใจไม่เพียงพอ ผู้ป่วยได้รับการผ่าตัดฝังอุปกรณ์กระตุ้นเส้นประสาทฟรีนิคทั้งสองข้างเพื่อช่วยการทำงานของกะบังลมปอดโดยใช้อุปกรณ์กระตุ้นไขสันหลังซึ่งใช้สำหรับรักษาอาการปวดเรื้อรัง ค่าความดันของก๊าซคาร์บอนไดออกไซด์ในลมหายใจออก, ปริมาตรของลมหายใจเข้า, และระยะเวลาที่ผู้ป่วยหายใจเองมีการเปลี่ยนแปลงในทางที่ดีขึ้นจนถึง 29 เดือนในช่วงติดตามผลการรักษาหลังผ่าตัด*

***สรุป**: การผ่าตัดกระตุ้นเส้นประสาทฟรีนิคเพื่อช่วยการทำงานของกะบังลมปอดสามารถลดความต้องการในการใช้เครื่องช่วยหายใจตลอดเวลาในผู้ป่วยที่มีการบาดเจ็บของไขสันหลังส่วนคอระดับสูงได้*