In high cervical quadriplegia, when the phrenic nerve nuclei are injured, incomplete or complete paralysis of the diaphragm can occur. Among these cases are patients in whom one side of the diaphragm is completely paralyzed and the other side is partially paralyzed. Compromised respiratory muscles seriously limit the mobility and phonation of these patients, and they need ventilation even during the chronic disease stage. Because of the degeneration of the nerve fiber resulting from damage to the phrenic nerve cell body, rhythmic electrical stimulation treatment of the phrenic nerve is usually inappropriate for these patients. However, the SAN usually remains intact, and the trapezius and sternocleidomastoid muscles innervated by the SAN participate in breathing activities in which the muscle contractions spontaneously keep in step with inspiration. We wondered if we could restore function to the paralyzed diaphragm through phrenic nerve neurotization using the functional SAN. Here, we present a case in which we attempted to transfer the SAN and make a neurotization with the ipsilateral, completely paralyzed phrenic nerve so that the SAN could innervate and eventually drive the paralyzed side of the diaphragm to improve respiratory function. To our knowledge, this is the first report of the clinical application of this technique.

The authors report a case of functional improvement of the paralyzed diaphragm in high cervical quadriplegia via phrenic nerve neurotization using a functional spinal accessory nerve. Complete spinal cord injury at the C-2 level was diagnosed in a 44-year-old man. Left diaphragm activity was decreased, and the right diaphragm was completely paralyzed. When the level of metabolism or activity (for example, fever, sitting, or speech) slightly increased, dyspnea occurred. The patient underwent neurotization of the right phrenic nerve with the trapezius branch of the right spinal accessory nerve at 11 months postinjury. Four weeks after surgery, training of the synchronous activities of the trapezius muscle and inspiration was conducted. Six months after surgery, motion was observed in the previously paralyzed right diaphragm. The lung function evaluation indicated improvements in vital capacity and tidal volume. This patient was able to sit in a wheelchair and conduct outdoor activities without assisted ventilation 12 months after surgery. (DOI: 10.3171/2011.3.SPINE10911)

Key Words • spinal accessory nerve • phrenic nerve • paralyzed diaphragm • high cervical quadriplegia

Abbreviation used in this paper: SAN = spinal accessory nerve.
Case Report

History and Examination. A C-2 complete spinal cord injury and C2–3 ossification of the posterior longitudinal ligament were diagnosed in a 44-year-old man after a fall from a bicycle. Two days postinjury, an intubation ventilator supported his breathing as a result of dyspnea. Three weeks postinjury, he underwent “combined anterior and posterior cervical decompression and internal fixation” surgery as well as a tracheostomy. Continuous mechanical ventilation was provided after the operation. During the initial 7–9 months postinjury, the ventilator was removed tentatively and temporarily. At 10 months postinjury, the ventilator was completely removed, while tracheal intubation was retained. The patient had a weak productive cough and usually required sputum suctioning through the tracheal tube. Moreover, dyspnea occurred after falling asleep or minor physical activity or fever, requiring oxygen uptake or assisted ventilation for relief. He frequently experienced sudden arousal after choking and needed oxygen. Neurological examination showed that the lowest normal sensory level was C-2, that sensation at the C-3 level was weakened, and that sensory sensations below C-4 had disappeared. The strength of the bilateral trapezius muscles was graded as 4+, and all key muscles were graded 0 (measured by manual muscle testing). Results of the diaphragm x-ray fluoroscopy indicated no motion of the right diaphragm muscle, and the up-down motion scope of the left diaphragm muscle was approximately 1 intercostal space (between the 9th and 10th intercostals; about 2.5 cm). Breathing exercises, in which the patient was trained to contract the trapezius muscle when inhaling and relax it when exhaling, were started from the 4th postoperative week. The exercise included four 15- to 30-minute sessions per day.

The patient was evaluated at 4 time points as follows: preoperatively and at 6, 9, and 12 months postoperatively. The observed factors included physical activity and symptom improvements, trapezius muscle strength, blood oxygen level, motion of the diaphragm observed with chest x-ray, and pulmonary function.

Operation. Eleven months postinjury, a neurotization of the trapezius branch of the right SAN to the right phrenic nerve was performed. Surgery was conducted under general anesthesia. The patient was placed supine with his neck extended 30° and left lateral flexion at 20°. An oblique incision 5 cm in length was made along the lower third of the posterior border of the sternocleidomastoid muscle to the lower third of the lateral border of the trapezius muscle (Fig. 1). After the trapezius branch of the SAN was exposed, pulse electrical stimulation was applied before proceeding to the next step (current intensity 2 mA, frequency 20 Hz), which showed strong contraction of the trapezius muscle. Another 4-cm incision was made within the right sternocleidomastoid region (Fig. 1), and the descending phrenic nerve was located on the surface of the anterior scalene muscle. No obvious motion of the diaphragm was observed with x-ray fluoroscopy after electrical stimulation of the phrenic nerve (current intensity 2 mA, frequency 20 Hz). The SAN was dissected at the lateral border of the trapezius muscle, and the cranial end of the dissected SAN was transposed back in an inner and upper direction through the subcutaneous tunnel to the previously prepared ipsilateral phrenic nerve and was anastomosed using the end-to-side method (Fig. 2).

Postoperative Course. Six months after surgery, all the preoperative symptoms were relieved, with no demand for supplemental oxygen and no constantly blocked tracheal tube. During deep breathing, slight breath sounds could be heard in the right lung under the armpit. The tracheal tube was removed 9 months postoperatively, and the patient could cough and dislodge sputum by himself. Twelve months after the operation, the patient went home, with wheelchair activities scheduled for at least 4 hours daily.

The strength of the trapezius muscle decreased 1 grade postoperatively, but was restored to Grade 4 three months after the operation and the breathing exercises.

The dynamic PaO2 and oxygen saturation in arterial blood were surveyed using oximetry, indicating trends toward improvements in blood oxygen pressure and oxygen saturation under conditions without oxygen uptake (Fig. 3).
The 6-month postoperative x-ray fluoroscopy study showed that the motion of the right diaphragm attained a scope of nearly 1 rib (the 8th rib, about 0.8 cm), whereas the motion of the left diaphragm increased to 3 cm (Fig. 4). There were no obvious differences in the motion of diaphragm between 6, 9, and 12 months after the operation.

Compared with the preoperative condition, obvious improvements were observed in the tidal volume rather than the vital capacity 6, 9, and 12 months after surgery. The tidal volume increased from 250 ml preoperatively to 660 ml at 6 months postoperatively and to 670 ml at 9 months postoperatively, which was close to the theoretical value of tidal volume (TVpr 660 ml). Twelve months after the operation, vital capacity was 1320 ml and tidal volume was 670 ml without the tracheal tube (Fig. 5). Although the vital capacity was far lower than the estimated theoretical value (4600 ml), the value could meet the needs of daily living.

Discussion

Anatomical and Physiological Potential

The spinal components of the SAN originate from cell bodies in the lateral portion of the anterior horn of the spinal cord (located mainly in C1–3). After they converge, it ascends posterior to the vertebral artery, enters the skull through the foramen magnum, exits the skull through the jugular foramen, and descends. Finally, it innervates the trapezius and sternocleidomastoid muscles. In the clinic we have seen that even though an injury is as high as the C-2 level, the SAN remains mostly functional. The stem of the phrenic nerve forms in the upper lateral edge of the anterior scalene muscle, descends along the surface of the anterior scalene muscle, and enters the thoracic cage between the subclavian artery and the subclavian vein, terminating at the diaphragm, which it innervates to cause movement of the diaphragm. The distance between the phrenic nerve and the SAN in the middle of the posterior cervical triangle is 3.5–5 cm. Therefore, in the cervical area, the trapezius branch of the SAN can be transposed to the phrenic nerve and directly anastomosed tension free. Furthermore, the diameters of the SAN and the phrenic nerve are very similar at 2 mm and 2.5 mm, respectively. In terms of the nerve fibers, the distal part of the SAN consists of 1328–1603 myelinated motor nerve fibers, whereas there are about 3000 of these fibers in the phrenic nerve. Thus, the diameter and type of nerve fibers match between these two nerves. While breathing at rest, respiration depends mainly on the diaphragm rather than other breathing muscles; during high-intensity breathing, especially in patients with a high cervical spinal cord injury, the trapezius and sternocleidomastoid muscles synchronize their activities with the inspiration. For patients with unilaterally paralyzed diaphragm muscles, the SAN can eventually innervate the paralyzed diaphragm after transferring the trapezius muscle nerve branch of the ipsilateral SAN to interconnect with the phrenic nerve and building up an assisted breathing model; that is to say, it is practicable to contract the trapezius muscle and synergistically activate the diaphragm.

Neural Anastomosis

An end-to-side anastomosis of the SAN and phrenic
nerve was conducted in this study for two reasons. Firstly, the diameter of the phrenic nerve is larger than that of the SAN, which included more nerve fibers. Secondly, the end-to-side anastomosis could cause less damage to the residual axons in the phrenic nerve.

Clinical Effects

Restoration of the diaphragm by anastomosing the SAN with the phrenic nerve has been reported in animal and cadaver studies. In the animal models, the transferred SAN could effectively pace the diaphragm; however, no clinical application has been reported. Based on our case study, the procedure led to satisfactory results: 6 months after the operation, partial improvements in the motion of the diaphragm were observed, and the patient’s lung function, especially the tidal volume, could fulfill the demands of his daily life.

However, it is possible that the breathing exercises and natural recovery of the spinal cord injury, rather than the transfer of the SAN to the phrenic nerve, mainly led to the reinforcement of the left diaphragm, resulting in pacing of the right paralyzed diaphragm and improvements in pulmonary function. Identifying which factor is responsible is difficult. But the natural recovery of motor function in patients with complete spinal cord injury rarely occurs after 1 year postinjury (our patient, over 11 months). In addition, before the operation, the patient had performed the breathing exercises for 3 months and did not get perceivable improvement in pulmonary function. Hence, the most likely explanation is that reinnervation of the right paralyzed diaphragm by the SAN caused itself and natural recovery of the spinal cord injury, rather than the demands of his daily life.

Indications for the Technique

Two points should be considered in the clinical application of this new technique. First, it depends on the type of diaphragm paralysis. For patients with totally paralyzed bilateral diaphragms, even if transferring the SAN to the phrenic nerve could pace the diaphragm, they also need long-term ventilation. Most patients who are able to activate one side of the diaphragm generally have enough function to meet the demands of their daily lives. For patients with partially paralyzed bilateral diaphragms, the operation itself might cause damage to the phrenic nerve and the side effects could be worse than the positive effects of this intervention. Therefore, indications for this new technique should be restricted to patients who have a weak diaphragm on one side and total or severe paralysis on the other. In these patients, dyspnea usually occurs after physical activity or slight increases in metabolism. As a result, outdoor activities by these patients are seriously limited, and they may even need intermittent assisted ventilation or oxygen uptake. Small improvements in pulmonary function can lead to large improvements in their daily lives. Second, the time window for performing the operation should be considered. Three factors should be taken into consideration: the generation velocity of the peripheral nerve, the degradation time of the motor endplate of the diaphragm, and the natural restoration of the injured spinal cord. In terms of nerve reinnervation, the best effects should be achieved when the operation is conducted within 3–6 months postinjury. Nevertheless, in view of the natural restoration of the injured spinal cord, 6–12 months postinjury might be the optimal window of time for performing this new technique. Recent studies indicate that in the unilateral spinal cord hemisection injury model, the ipsilateral motor endplate of the paralyzed diaphragm showed adaption and delay of degradation. Therefore, our suggested window of time is within 6–12 months postinjury.

Conclusions

Transfer of a functional SAN to reinnervate the paralyzed phrenic nerve in high cervical quadriplegia could improve pulmonary functioning. However, further clinical studies of this technique are needed before advocating its use.

Disclosure

The authors report no conflict of interest concerning materials and methods used in the present report and findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: JJ Li, Yang. Acquisition of data: Yang, Zhang, Du, J Li, Wang, Gong, Cheng. Analysis and interpretation of data: JJ Li, Yang, Zhang, Gao. Drafting the article: JJ Li, Yang, Gao. Critically revising the article: JJ Li, Yang, Zhang. Approved the final version of the paper on behalf of all authors: JJ Li. Statistical analysis: Yang. Administrative/technical/material support: Yang, Zhang, Du, Gao, J Li, Wang, Gong, Cheng. Study supervision: JJ Li, Yang.

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References


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Address correspondence to: Jian-jun Li, M.M., Department of Spinal and Neural Function Reconstruction, China Rehabilitation Research Center, Beijing 100068, China. email: crrclij2010@sohu.com.