A 38-year-old man had been tetraplegic and ventilator-dependent after sustaining a traumatic cervical spine fracture at the C1/C2 level in 1991, at the age of 22 years. He had bedbound and mechanically ventilated since then. A multidisciplinary management team approached him in 2003 and helped him to become ambulatory and independent in his daily activities of living. We successfully implanted the diaphragm pacing stimulation system in this patient in 2004. Diaphragm pacing by phrenic nerve stimulation is well accepted in western countries, and has been in clinical application for children and adults for decades. Its use facilitates ambulation and improves the quality of life of tetraplegic individuals with chronic ventilatory failure.

In 2003, a multidisciplinary management team of doctors, nurses and therapists, including orthopaedic and rehabilitation consultants, respiratory physicians, cardiothoracic surgeons, speech therapists, physiotherapists, and occupational therapists started planning for the implantation of a breathing pacemaker to improve his independence and mobility. He was assessed by a dentist for oral hygiene, a speech therapist for swallowing and speech, a neurologist for bilateral phrenic nerve function, and a clinical psychologist for his psychological status. It is important to test phrenic nerve function preoperatively because using phrenic nerve stimulation to instigate diaphragmatic muscle contraction is only possible if both phrenic nerves are viable. They are tested using percutaneous electrical stimulation at the neck, which causes diaphragmatic contraction, recorded as a signal on electromyography. This test requires expertise for its performance and interpretation. He also had a chest X-ray (Fig a) and arterial blood gases done preoperatively (ventilator settings: controlled mechanical ventilation mode, respiratory rate 12/minute, tidal volume 440 mL, fraction of inspired oxygen 0.21), revealing a pH of 7.45, P\text{O}_2 of 186 mm Hg, P\text{CO}_2 of 24 mm Hg, bicarbonate (HCO\textsubscript{3}) of 16 mmol/L, and base excess (BE) of -6 mmol/L.

Cardiothoracic surgeons implanted the breathing pacemaker system (Avery Biomedical Devices Inc., New York, US) into his phrenic nerves bilaterally, with a thoracoscope inserted in the 7th intercostal space at the anterior axillary line, and two anterior thoracotomies over the 3rd intercostal space on the left and the 2nd intercostal...
A 38-year-old man sustained C1/C2 cervical spine injury in 1991 at the age of 22. Since then he has been tetraplegic and dependent on ventilators for life. In 2003, an interdisciplinary team planned to assist the patient to live independently. In 2004, they successfully implanted a diaphragm pacemaker. This type of diaphragm pacemaker, using the principle of stimulating the phrenic nerve, has been widely employed in children and adults in the Western world for decades. For patients with tetraplegia who have been ventilator-dependent, this technology increased their freedom of movement and quality of life.

For the Hong Kong patient, the diaphragm pacing was implanted on his right side. One electrode was sutured to each phrenic nerve and the paired radioreceivers were implanted in subcutaneous pockets in the upper chest. The chest X-ray before and after implantation is shown (Fig a). The pacing system was functioning well postoperatively, both left and right pacers worked independently but in synchrony, however, the electrical stimuli required for the diaphragmatic contraction on either side differed. He began pacing exercises 1 month after the operation, when the chest wounds had healed and the pacing wires were well stationed in the diaphragms and not so easily dislodged with muscle contraction. His arterial blood gases taken at the time were: pH 7.43, PaO\textsubscript{2} 96 mm Hg, PaCO\textsubscript{2} 26 mm Hg, HCO\textsubscript{3} 21 mmol/L, and BE -1 mmol/L. His diaphragms had not been in active use for years and failed to achieve a static tidal volume initially because of prolonged deconditioning, so the duration of pacing was increased gradually, day by day, over the first 6 months. His tidal volume was monitored with a portable respirometer on his tracheostomy while he was being paced. He was also taught to speak with a speaking valve capped on his tracheostomy, as guided by the speech therapist, while using diaphragm pacing. Occupational therapists also modified his automatic wheelchair to enable power steering using his chin mounted on the control panel, and the physiotherapists tried to increase his exercise endurance and train him in postural adaptations as he also suffered from autonomic dysfunction.

Today, he can talk and be more interactive with his environment and the people around him. He leads an independent life when using the pacers for over 10 hours during the day. This has facilitated ambulation markedly, as he can use the breathing pacemaker while sitting in his automatic wheelchair, and participate in various activities outside the hospital. His exercise endurance and autonomic adaptations are much improved, in particular, his postural-related blood pressure changes.

**Discussion**

Clinical use of diaphragm pacing was first reported in 1972, and it has been shown to reduce the incidence of pulmonary infections when compared to mechanical ventilation. It was conducted using low-frequency electrical stimulation at a slow respiratory rate to condition the diaphragm muscle against fatigue in order to maintain ventilation. There have been a number of case series and long-term follow-up studies reporting the success of its clinical application to achieve complete stable ventilation, and to improve the prognosis and life quality of patients with severe chronic respiratory failure. Most patients are able to speak while being paced. A programme consisting of careful patient selection, meticulous surgical techniques, adequate training in the use of the device, and regular follow-up will contribute to a successful outcome.

The diaphragm pacing system used on our patient is currently approved by the Food and Drug Administration (FDA) in the United States. It has both internal and external components (Fig b). The system consists of internal electrodes sutured to the right. One electrode was sutured to each phrenic nerve and the paired radioreceivers were implanted in subcutaneous pockets in the upper chest. The chest X-ray before and after implantation of the diaphragm pacing stimulation system. The arrows indicate the positions of the radioreceivers. (b) A schematic diagram of the internal and external components of the diaphragm pacing stimulation system (with permission from Avery Biomedical Devices Inc., New York, US).
the phrenic nerves on both sides and connected to radioreceivers placed under the skin in the chest bilaterally. An external transmitting box is connected to an antenna that is taped over the surface of the skin, just above the subcutaneous receiver on either side. The transmitting box is battery powered, sending stimuli via the antennae to the receiver implants which translate radio waves into stimulating pulses that are delivered to phrenic nerves by the electrodes. This initiates the ventilatory cycle: the diaphragm muscles contract and produce the inhalation phase of breathing. The transmitter then stops generating signals, allows the diaphragms to relax, and the exhalation phase occurs, producing a normal breathing pattern. This is an expensive device, and the implantation requires an invasive procedure, therefore, detailed pre-implantation evaluation is mandatory. Good surgical candidates should have normal cognitive function, complete respiratory failure without recovery for more than 3 months but have good lung function (assessed with chest X-rays and arterial blood gases), and intact phrenic nerves. This portable breathing system has been proven useful in both children and adults. Specific indications for consideration of the implantation of a breathing pacemaker system include patients who have high cervical spine injuries and congenital central hypoventilation syndrome, the former being the more common cause reported in the literature. Congenital central hypoventilation syndrome is a rare syndrome present from birth, and is defined as the failure of automatic control of breathing. People with this condition require lifelong ventilatory support during sleep, and approximately one third require ventilatory support 24 hours a day. Assisted ventilation with diaphragm pacing has been shown to improve quality of life as it helps increase mobility tremendously. Its use also optimises normal neurodevelopmental changes in children, thus enhancing their ability to achieve independent living.

There are problems associated with the implantation of this pacing system, such as infection and pulmonary complications following a thoracic surgical approach, dislodgement of the pacer electrode, and malfunction of the hardware. Patients who have not had a functioning diaphragm for 6 months, especially those who have had paralysed diaphragms for 2 or more years, require a period of diaphragm conditioning that may last up to 9 months before achieving optimal diaphragm function with pacing. Continuous stimulation of the phrenic nerves may induce diaphragmatic fatigue and, occasionally, irreversible damage to the lower motor neuron. Conditioning requires gradually increasing the duration of pacing time per hour during the day, using low-frequency stimulation and a slow respiratory rate, and monitoring the tidal volume with a respirometer, as well as the abdominal excursion, clinically. Patients should continue to receive mechanical ventilatory support while the pacing frequency is increased until they can breathe with the pacers alone.

Diaphragm pacing has evolved as an important therapeutic modality in a small group of carefully selected patients with severe chronic respiratory failure. These subjects can become substantially ventilator-independent if using the pacer 24 hours a day, and some may work or study during the day, enabling a much better quality of life. Recent advancements in technology have led to the development of the ‘NeuRx diaphragm pacing stimulation system’, recently approved by the FDA in the US. Its electrodes have been designed for implantation into the diaphragm muscles rather than the phrenic nerves. This procedure can be done on an out-patient basis, and it takes approximately 2 hours using a laparoscopic approach. This permits avoidance of a thoracotomy with its associated peri-operative morbidity and scarring, and thus encourages wider utilisation of diaphragm pacing in suitable candidates, but expertise in this field is required.

**Conclusion**

Use of a diaphragm pacing stimulation system is a viable alternative to mechanical ventilation in tetraplegic patients with chronic respiratory failure.

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**TABLE. Comparisons between the use of mechanical ventilation and the diaphragm pacing stimulation system for tetraplegic patients**

<table>
<thead>
<tr>
<th></th>
<th>Mechanical ventilator</th>
<th>Diaphragm pacing stimulation system</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Cost (HK$)</strong></td>
<td>130 000</td>
<td>600 000</td>
</tr>
<tr>
<td><strong>Power supply</strong></td>
<td>Major household electrical supply</td>
<td>Disposable 9V external battery</td>
</tr>
<tr>
<td><strong>Battery life</strong></td>
<td>Backup internal battery: 8 hours</td>
<td>400 hours</td>
</tr>
<tr>
<td><strong>Surgery involved</strong></td>
<td>Tracheostomy under local anaesthesia</td>
<td>Placement of electrodes and radioreceivers in chest under general anaesthesia</td>
</tr>
<tr>
<td><strong>Duration of surgery</strong></td>
<td>30 minutes</td>
<td>2-4 hours</td>
</tr>
<tr>
<td><strong>Weight</strong></td>
<td>10 kg</td>
<td>1 kg</td>
</tr>
<tr>
<td><strong>Lifespan</strong></td>
<td>10-15 years</td>
<td>Life</td>
</tr>
<tr>
<td><strong>Speech</strong></td>
<td>No</td>
<td>Yes</td>
</tr>
</tbody>
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insufficiency (Table). Implantation of the diaphragm pacers in appropriate subjects can lead to independent living, enhanced mobility, better quality of life, and ease their integration into society. A multidisciplinary team approach is crucial for achieving a successful outcome.

References