Thoracoscopic Placement of Phrenic Nerve Electrodes for Diaphragmatic Pacing in Children

By Donald B. Shaul, Paul D. Danielson, J. Gordon McComb, and Thomas G. Keens
Los Angeles, California

Background/Purpose: Diaphragmatic pacing can provide chronic ventilatory support for children who suffer from congenital central hypoventilation syndrome (CCHS) or cervical spinal cord injury. The authors report a new thoracoscopic approach for establishing diaphragm pacing.

Methods: Between 1997 and 2000, 9 children ranging in age from 5 to 15 years and suffering from these disorders underwent thoracoscopic placement of bilateral phrenic nerve electrodes. A 3- or 4-trocar technique was used to dissect the phrenic nerve in the midchest and suture a phrenic nerve electrode (Avery Laboratories I-110A, Commack, NY) into place. The electrode was tunneled to a subcutaneous pocket in the upper abdomen and attached to an implanted pacing unit.

Results: Bilateral electrodes were placed successfully into all patients. The average procedure time was 3.3 hours (range, 2.5 to 4.6), and average hospital stay was 4.2 days (range, 3 to 5). Four patients experienced postoperative complications (pneumonia, atelectasis, bradycardia, and pneumothorax). Average follow-up has been 30 months (range, 15 to 49). Eight patients have reached their long-term pacing goals.

Conclusions: Phrenic nerve electrodes can be implanted thoracoscopically and allow the successful use of diaphragmatic pacing therapy. Avoidance of thoracotomy with its associated perioperative morbidity and scarring may encourage wider utilization of diaphragmatic pacing in children.
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INDEX WORDS: Diaphragm pacing, congenital central hypoventilation syndrome, thoracoscopy.

CONGENITAL CENTRAL hypoventilation syndrome (CCHS, Ondine's Curse) is a rare disorder of automatic controlled breathing.[1-5] CCHS patients hypoventilate or fail to breath, and their symptoms worsen during sleep. They will not outgrow the condition.[5] Therefore, they have a lifelong need for ventilatory support. Diaphragm pacing is one method of providing chronic ventilatory support for these patients.[7-12] During diaphragm pacing, diaphragmatic contraction results from direct electrical stimulation of the phrenic nerves. Candidates for this technique must have intact phrenic nerves and diaphragmatic muscles. We report the establishment of bilateral diaphragmatic pacing using thoracoscopic placement of phrenic nerve electrodes.

MATERIALS AND METHODS

Permission to perform this retrospective study was obtained from the institutional review board at Childrens Hospital Los Angeles. Nine patients were suitable candidates for diaphragmatic pacing because they were afflicted by CCHS (8 patients) and cervical spinal cord injury (1 patient). The ages ranged from 5 to 15 years with a median age of 9 years. Five were girls and 4 were boys. Many patients had a history of prior chest tube placements, one had a history of prior empyema, and one had undergone a patent ductus arteriosus (PDA) ligation as a neonate. One patient also had Hirschsprung's disease. All of the patients were consented for thoracoscopic placement of phrenic nerve electrodes with the possible need for an open thoracotomy.

Operative Technique

The operative technique evolved over the course of the 9 operations. The following describes the technique, which was used in the latter patients.

Patient preparation. The patient is prepared by establishing definitive single-lung ventilation using a dual lumen endotracheal tube in older patients or a contralateral main stem intubation using a small, cuffed endotracheal tube in younger patients. The patient is positioned on a vacuum extractable beanbag with the chest in a nearly full lateral position and the hips rotated so that the abdomen is in a semilateral position. We found it best to position both surgeons behind the patient with the scrub nurse and monitor in front of the patient. A second monitor can be placed behind and at the head of the table to allow the scrub nurse to visualize the procedure. Parenteral antibiotics (cefazolin or vancomycin) are administered in standard dosages.

Electrode preparation. Diaphragm pacing works by transmitting a radio signal from an external antenna (Mark IV Transmitter and Antenna, Avery Laboratories, Commack, NY) to an implantable receiver, which converts the radio signal into an electrical impulse. The electrical impulse travels from the receiver through a detachable electrode (Model I-110 A, Avery Laboratories) to the phrenic nerve (Fig 1). Electrical stimulation of the phrenic nerve causes diaphragmatic contraction. The patient can remove the external antenna (Fig 2); however, the receiver and electrode are implanted permanently. Spare coils of electrode wire and the connection to the receiver are kept in a SILASTIC® (Dow Corning, Midland, MI) envelope made by folding a

From the Departments of Surgery, Neurosurgery, and Pediatrics, Childrens Hospital Los Angeles and Keck School of Medicine, University of Southern California, Los Angeles, CA.

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Address reprint requests to Donald B. Shaul, MD, 4650 Sunset Blvd, MS #100, Los Angeles, CA 90027.

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Fig 1. The phrenic nerve electrode wire is shown attached to the receiver. They are detached from one another during lead placement to facilitate passage of the male end of the electrode through the diaphragm. The long tab on the electrode can be reduced by half its length to facilitate insertion into the chest and positioning around the nerve.

sheet of SILASTIC® onto itself and suturing the ends together. Use of the envelope is important because it prevents direct scar tissue formation around the spare electrode wire permitting it to be subsequently changed or pulled into the chest during growth. The envelope can be prepared during anesthetic induction and establishment of single lung ventilation.

With single contralateral lung ventilation established, a 5-mm trocar is inserted in the seventh intercostal space in the posterior axillary line. The chest is inflated to a pressure of 5 mm Hg to speed up lung deflation. All trocar sites are infiltrated with 0.5% Bupivacaine during their placement. An additional 5-mm trocar then is inserted into the ninth intercostal space, posterior axillary line followed by a third 5-mm trocar in the fifth intercostal space, posterior axillary line. Adhesions are taken down as needed. A fourth trocar can be inserted as a lung retractor if needed. The phrenic nerve is identified, usually at the cephalad aspect of the pericardium, and 2 parallel incisions are made in the mediastinal pleura (Fig 3). Placement of the electrode on the cephalad aspect of the pericardium leaves sufficient length of distal phrenic nerve available in the event that future electrodes might be required. A small subcostal incision then is made, and a subcutaneous pocket is formed to house the receiver and SILASTIC® envelope containing the wires. A 4-inch length of Penrose drain then is placed over the male end of the electrode wire and is tied in position with a heavy suture. The opposite end of the Penrose is inserted through the lower most trocar site. The Penrose, wire, and electrode are fed sequentially into the chest. The long tab present on the electrode can be trimmed off before inserting the electrode. Great care must be taken when passing the phrenic nerve portion of the electrode into the chest because this electrode is fractured easily. A tonsil clamp then is passed from the subcutaneous pocket under the ribs and through the anterolateral and peripheral aspect of the diaphragm into the chest cavity. The free end of the Penrose drain is grasped and pulled, along with the male end of the electrode and excess wire, into this subcutaneous pocket.

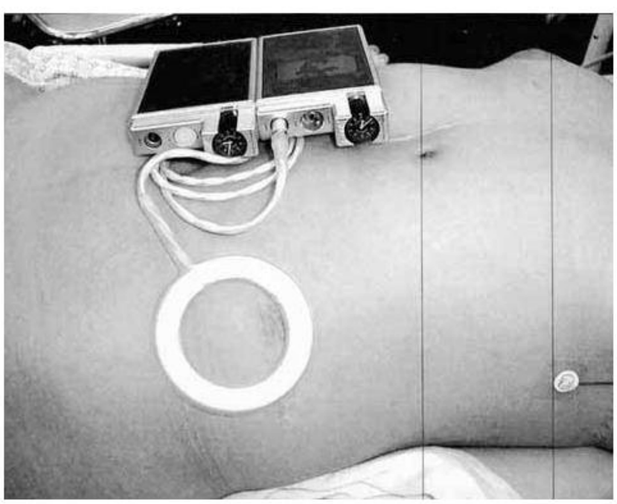

Fig 2. The Mark IV Transmitter and Antenna (white ring) are shown positioned on the patient. The ring is centered over the subcutaneous receiver to optimize transmission of the signal. The transmitter and antenna are removed when not needed.

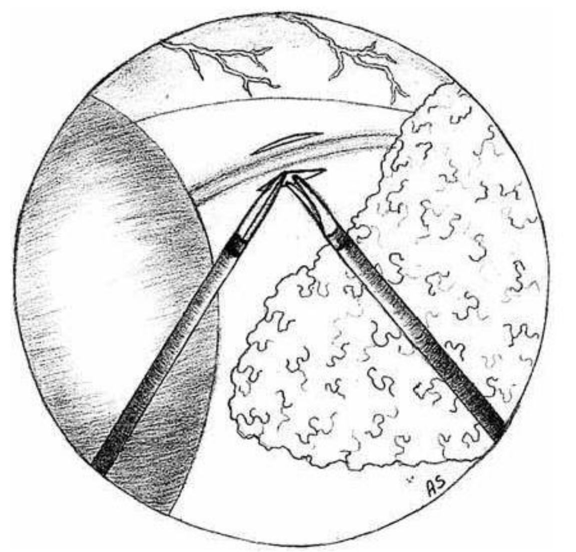

Fig 3. The surgeon's view as seen endoscopically is shown. The diaphragm is on the left, and the lower lobe, partially deflated, is shown on the right. The figure depicts dissection of the phrenic nerve on the central portion of the pericardium. Often it is easier to dissect the phrenic nerve in a more proximal location at the cephalad border of the pericardium. Great care must be taken not to damage the delicate phrenic nerve during this procedure. Electrocautery must not be used in proximity to the phrenic nerve.

The phrenic nerve electrode then is passed through the incisions in the mediastinal pleura so that the phrenic nerve lies in the groove of the electrode (Fig 4). The electrode then is sutured carefully in position using a suture on either side of the nerve using 4-0 nonabsorbable sutures on a small taper needle (Fig 5). The electrode then is attached to the receiver and tested with the help of the Avery engineer who is present at every operation. The receiver and extra coils of wire are then placed into the SILASTIC® envelope and into the subcutaneous pocket and retested. Sufficient wire is pulled into the chest cavity to prevent traction on the phrenic nerve. Care must be taken in handling the lead wire to prevent breaks in insulation, which could result in failure.

A 20F chest tube is passed through the caudal-most incision and positioned laterally in the chest cavity where it will not interfere with the wire. The lung then is inflated under vision. All incisions are closed in layers, and the chest tube is connected to suction. The patient is repositioned into the exact opposite lateral position, and the endotracheal tube is manipulated to allow for contralateral single lung ventilation. The identical procedure is performed on the contralateral side.

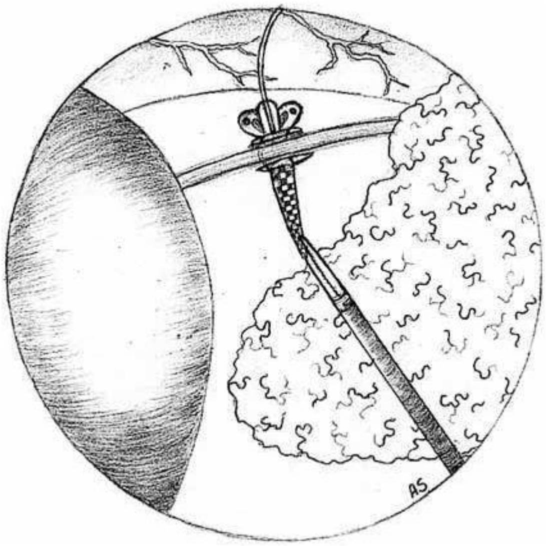

Fig 4. The electrode is shown positioned underneath the phrenic nerve. The nerve should lie in the groove on the electrode in direct contact with the exposed metal. In some cases, the electrode is best passed from an anterior to posterior direction; however, in other cases the electrode may be passed from posterior to anterior.

At the end of the procedure, the patient remains intubated, usually via their existing tracheostomy, and placed supine on the operating room table. A chest x-ray is obtained intraoperatively, and, if there is no pneumothorax, no air leaks, or other contraindication to removing the chest tubes, they are removed while the patient remains under anesthesia, and occlusive dressings are placed.

Postoperative Care

Patients always are placed in the intensive care unit after surgery, primarily for management of their ventilation. Prophylactic antibiotics are continued for 48 hours. Because this technique requires that each lung be deflated for 60 to 90 minutes, there is an increased chance of pneumonia occurring postoperatively. To combat this, aggressive pulmonary hygiene is instituted using aerosolized bronchodilators and chest physiotherapy to promote the removal of pulmonary secretions and prevent atelectasis. The patients are discharged home when stable.

Diaphragm pacing generally is delayed for 6 weeks to allow for the completion of a fibrotic tissue reaction around the phrenic nerve electrode, which prevents electrode migration. Patients are then readmitted for a 3-day, 2-night period in which pacing is initiated. The amplitude, rate, and time of phrenic stimulation is adjusted to achieve the desired tidal volume and ventilation determined by noninvasive gas exchange monitoring (pulse oximetry $\geq$95% and end-tidal $P_{CO_2} \leq 35$ torr). Even though CCHS patients use their diaphragms to breathe spontaneously during wakefulness, the diaphragm pacers stimulate different nerve fibers and motor units. Therefore, patients usually will only tolerate 1 to 2 hours of diaphragm pacing in the beginning, and *endurance training* of these motor units is required. Therefore, the efficacy of pacing is monitored carefully visually and with noninvasive gas exchange monitoring (pulse oximetry and end-tidal P_{CO_2}), and pacing is stopped at the first sign of fatigue (decreased diaphragm contraction at the same electrical settings). The purpose of the hospitalization is to establish the optimal diaphragm pacer settings. The patient then is discharged home, and diaphragm pacing is increased by 1 hour each week until the pacing goal has been reached (either all night or 12 to 14 hours per day while awake).

RESULTS

Bilateral electrode implantation was completed successfully thoracoscopically in all 9 patients (18 electrodes) during the period from June 1997 through June 2000. The average procedure time was 3.3 hours (range, 2.5 to 4.6 hours) and average total anesthesia time was 5.3 hours (range, 5 to 6.5 hours). One patient had a concurrent bronchoscopy, and a second patient had a bronchoscopy and tracheostomy revision during the same anesthetic. The median intensive care unit (ICU) length of stay was 2 days (range, 1 to 4 days), and median hospital length of stay was 4 days (range, 3 to 5 days). Five of eighteen chests required a fourth trocar to be inserted for optimal exposure. Two patients had postoperative atelectasis, one of whom had a subsequent pneumonia requiring outpatient antibiotic treatment. A small pneumothorax seen on a postoperative chest x-ray developed in one patient, but this resolved without treatment. One patient early in the series had a trocar inserted through the diaphragm into the liver. This was left in place for the remainder of the procedure and then was removed without consequences. A final patient had postoperative bradycardia requiring treatment. One patient had unilateral pacing failure 28 months after the original procedure and was taken back to the operating room where it was found that the receiver had failed and was replaced.

All patients still are undergoing follow-up. The mean follow-up is 30 months (range, 15 to 49 months). Eight patients have reached their long-term pacing goals, and 3 patients do not have a tracheostomy.

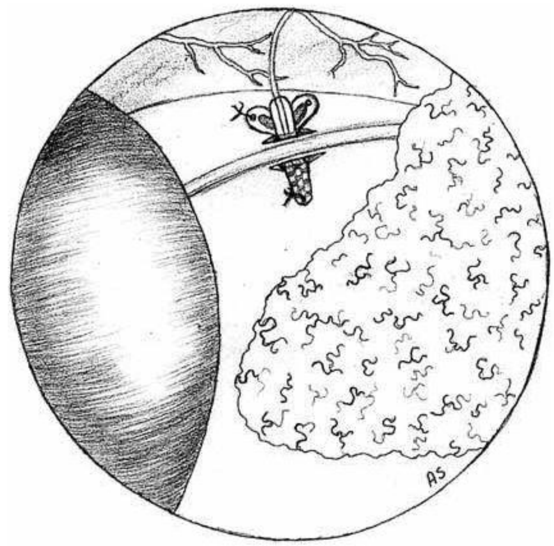

Fig 5. The electrode is shown sutured in position using 4-0 nonabsorbable sutures.

DISCUSSION

In 1998, we reported the first successful placement of phrenic nerve electrodes using thoracoscopy.[13] The current report includes 9 patients and describes substantial modifications to the surgical technique, primarily treating each side as a separate operative procedure under a single anesthetic. Institution of separate single lung ventilation and lateral patient positioning has facilitated the technique. The original procedure required 4.6 hours of surgical time and 5.5 hours of anesthetic time. This has been reduced to a mean of 3.3 hours of surgical time. The recognition that aggressive postoperative pulmonary hygiene is required has resulted in elimination of postoperative pneumonias. It also has been shown that thoracoscopic placement of the electrodes does not result in damage to the electrodes or injuries to the phrenic nerve, which has been seen in other patients after open thoracotomy.[7,8,11]

Patients with CCHS are very susceptible to the effects of general anesthesia, which can result in intraoperative or postoperative bradycardia. This occurred in several of our patients but only required treatment in one. They also can develop apnea and hypoventilation with atelectasis and pneumonia. Surgeons operating on these patients must be aware of these complications and be prepared to deal with their consequences. One patient in whom pacing failure developed was tested before reoperation and felt to have a break in the electrode insulation leading to speculation that the electrode may have been damaged during the original surgical procedure. At reoperation, the receiver was changed, and this solved her problem. Nonetheless, electrode fracture and failure can occur, and great care must be utilized when handling this delicate device.

The indications for diaphragm pacing include any patient who has an intact phrenic nerve and diaphragm and desires to be free from standard mechanical ventilation for a period usually less than 24 hours. CCHS patients are excellent candidates. Some CCHS patients hypoventilate only during sleep and, therefore, can utilize diaphragm pacing at night rendering them ventilator free. Spinal cord injury patients can utilize diaphragm pacing during the day to free them up from ventilator tubing while pursuing their activities of daily living. The portability of diaphragm pacing provides them with tremendous advantages. The relative contraindications to performing the procedure thoracoscopically include a recent chest infection and prior chest surgery or empyema. Two of our patients had these latter conditions and were told that thoracoscopic lead placement might not be possible. However, we found that the amount of intrathoracic adhesions was not prohibitive in either case.

Alternatives to thoracoscopic placement of electrodes have been performed. The cervical phrenic nerve has been used in some adult patients; however, the preferred location of these electrodes is on the intrathoracic phrenic nerve in children.[9-12] It is believed that attachment of the electrode to a relatively distal portion of the phrenic nerve maximizes the recruitment of motor end plates on the diaphragm. The electrode has also been placed on the inferior aspect of the diaphragm using serial diaphragm stimulation to pinpoint the most desirable locations for the lead placement.[14] The establishment of thoracoscopic placement on the visible intrathoracic phrenic nerve should obviate the need for this complex approach.

The ability to establish diaphragm pacing without the need for concurrent bilateral thoracotomy has proven to be very desirable to many of our patients. While the indications for the procedure are relatively rare, several patients opted for the technique once they learned it could be performed thoracoscopically. Surgeons performing thoracoscopy already realize its benefits in terms of decreased postoperative pain and shortened hospital stay. There are tremendous cosmetic benefits as well. Finally, it was the initial feeling of the lead author that this would be a relatively straightforward procedure to perform thoracoscopically. However, even after 9 procedures, this procedure remains technically demanding. The electrodes are quite small, and suturing them into perfect position on a beating heart has proven to be challenging. Surgeons desiring to perform this technique should be familiar with endoscopic suturing techniques. Nonetheless, the procedure can be performed successfully and safely with tremendous benefits to the patient. It is our understanding from reviewing the literature and discussing the procedure with the Avery Company that this series represents the largest series by far to date.

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Discussion

Dr Coran: (Ann Arbor, MI): There often is an association between Ondine's curse and Hirschsprung's disease, which probably would modify the way you place the subcutaneous pocket. Did any of these patients have Hirschsprung's?

P.D. Danielson (response): In fact, one patient did have Hirschsprung's disease and we were still able to locate the pocket in the upper abdomen but in an area away from any possible stoma.

Dr Caty (Buffalo, NY): Has there been lab work done? Does a pace diaphragm achieve the same title volumes as a spontaneously breathing animal or human?

P.D. Danielson (response): Yes it can. There has been extensive studies of this. This all began back at Yale University with Dr Glen, a thoracic surgeon, and the whole training process after the electrode is implanted is long because it actually converts the muscle fibers into sort of a slow twitch muscle fiber, and that is why the patients can immediately be put on to diaphragmatic pacing totally. If the transition to the slow twitch is accomplished, the patients are able to maintain similar titer volumes.