Perioperative Considerations for Patients with Central Hypoventilation Syndrome and Diaphragmatic Pacemakers

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Abstract

Congenital Central Hypoventilation Syndrome (CCHS), also called Ondine's curse, is a life threatening genetic disorder with autonomic nervous system abnormalities resulting in hypoventilation, especially while sleeping. While mechanical ventilation is the gold-standard treatment, an alternative option is a diaphragmic pacemaker (DP), which stimulates both phrenic nerves and causing diaphragmatic contraction to generate breaths. Conditions during surgery and general anesthesia can lead to complications in patients with diaphragmatic pacemakers. There is a paucity of literature on proper management of patients with diaphragmatic pacemakers in the perioperative period. We present the case of a 12-year-old male with CCHS with an implanted diaphragmatic pacemaker who underwent outpatient surgery under general anesthesia with a discussion of perioperative management of diaphragmatic pacemakers.

INTRODUCTION

Congenital Central Hypoventilation Syndrome (CCHS) is a genetic disorder involving autonomic nervous system dysregulation. (1). These children are intellectually intact, however, the mobility restrictions imposed by a bulky ventilator is drastically life-altering and hinders normal development and learning In the 1970s, Dr. Carl E. Hunt at Children's Memorial Hospital (Northwestern University) introduced diaphragmatic pacing to children and infants suffering from CCHS. Children with chronic respiratory failure from CCHS can utilize diaphragmatic pacing (DP), also referred to as phrenic nerve stimulation, as an alternative for mechanical positive pressure ventilator support. Given this relatively uncommon and unfamiliar device to pediatric anesthesiologists and medical professionals involved in perioperative care, it is not surprising that there is a paucity in the literature regarding perioperative risk and safe discharge criterion for such patients presenting for ambulatory surgery. After reviewing the literature and gaining manufacturer insight, we present a case report, provide recommendations and highlight important clinical considerations involving patients with diaphragmatic pacemaker presenting for ambulatory surgery.

CASE PRESENTATION:

A 12-year-old male with CCHS treated with an implanted diaphragmatic pacemaker, presented to San Diego children's hospital for left open reduction, internal fixation of a distal radius fracture that was sustained after falling on an outstretched hand. The patient was originally diagnosed with CCHS within the first days of life, leading to placement of a tracheostomy at 1 month of age. Genetic testing was positive for the PHOX2B genotype, a known mutation for CCHS. The patient was also diagnosed with Hirschsprung's disease requiring a colostomy and intestinal pull through, epilepsy, and sinus pauses requiring an epicardial cardiac pacemaker. The patient remained ventilator dependent and the family decided to undergo placement of an Avery Biomedical Device for DP at 6 years of age with hope to ultimately discontinue mechanical ventilation. The DP consisted of a Mark IV transmitter, implanted receiver and electrode for bilateral pacing. At the time of diaphragmatic stimulator placement surgery the diaphragm pacer was set at a rate of 18; amplitude 3.0 mHz on the left and 3.0 mHz on the right; pulse interval 50 msec; pulse width 150 sec. These settings were selected to achieve the results of adequate tidal volume/minute ventilation, which were confirmed to be optimal based on a formal sleep study performed by a pulmonologist. The patient was continuously paced 24 hours

a day. He also had a patent tracheostomy and a home ventilator. Despite optimal function of the diaphragmatic pacer, the patient was mechanically ventilated at night, as DP was unable to provide all his ventilatory needs while sleeping. It also provided positive pressure ventilation support during periods of acute respiratory illness.

After fasting, induction of anesthesia was accomplished by inhalation of increasing concentration of sevoflurane, titrated to unconsciousness using the tracheostomy. After loss of consciousness, an intravenous catheter was inserted and general anesthesia was maintained with a propofol infusion titrated between 150 and 200 mcg/kg/minute. Positive pressure ventilation was given through the tracheostomy tube. Due to the possibility of a conflict with the DP and unipolar cautery, the diaphragmatic and cardiac pacemakers were turned off by the manufacturer representatives before the start of the case, and were turned back on after emergence. Intravenous fentanyl (2mcg/kg), meperidine (0.5mg/kg), and acetaminophen (15 mg/kg) were administered for pain control, in addition to local anesthetic infiltration at the incision site by the surgeon. The case proceeded without complication and the patient began breathing with assistance at the end of the case, prior to leaving the operating room with normal end-tidal CO2 on pressure support of 7cm H20. The patient was brought to post anesthesia care unit (PACU), with supplemental oxygen being administered via a mapleson C circuit. On admission to the PACU, the patient was sleepy, but breathing spontaneously. DP was immediately reinitiated with the pre-operative settings. After 30 minutes of spontaneous ventilation with the pacemaker functioning, the patient was not fully conscious so an arterial blood gas (ABG) was obtained. ABG results showed a Pao2 of 65 mmHg and Paco2 of 88 mmHg while receiving supplemental oxygen at 4L/minute. To treat hypercarbia and to support ventilation while emerging from general anesthesia the patient was restarted on positive pressure ventilation through his tracheostomy tube. After 45 minutes, the patient appeared significantly more awake, with markedly improved tidal volumes (300 mL/breath or 10 mL/kg/breath as observed on the ventilator monitor) and was weaned off mechanical ventilation. The patient had total of 2.5 hour PACU stay. A repeat ABG was obtained prior to discharge and showed normocapnia and normoxia on room air (paCO2= 42 and paO2=81). After meeting standard PACU discharge criteria the patient was discharged home. The decision not to observe the patient overnight was made because due to the family's desire to return home and the family's ability to provide mechanical ventilation. The family used mechanical ventilation with end-tidal monitoring the first night following the procedure with strict precautions to return to the hospital should end-tidal remain elevated or mental status remain suppressed.

DISCUSSION:

CCHS is an autosomal dominant disorder caused by a defect in the PHOX2B gene, leading to a failure in automatic control of breathing and abnormal response to hypercarbia and hypoxia. The PHOX2B gene is responsible for regulation of neural crest cell migration and development of the autonomic nervous system. Due to disordered integration of input from chemoreceptors responsible for responding to hypercarbia and hypoxemia, these patient will hypoventilate or remain apneic despite marked abnormalities in arterial carbon dioxide levels and arterial saturation. This disordered breathing typically occurs during sleep, but in severe cases is present while awake. Most patients will be diagnosed as neonates, but in a small subset of patients symptoms will not present until later in life. Due to the abnormal neural crest cell migration, CCHS patients have an increased incidence of Hirschsprung's disease, as well as, tumors of neural crest origin, such as mediastinal and adrenal ganglioneuromas. There is also increase incidence of cardiac conduction abnormalities, specifically bradyarrhythmias, which may require cardiac pacemaker placement. Due to the abnormal response to hypercarbia, which will worsen with general anesthesia, diaphragmatic pacemakers, cardiac pacemakers and multiple comorbidities, patients with CCHS present unique challenges throughout the entire perioperative period.

Pre-operative planning is essential in patients with CCHS. All patients should receive a pre-operative evaluation, with coordination amongst the multiple specialists involved in the patient's care. If scheduling permits, it is recommended that patients with DP have cases scheduled as first starts, to provide enough time to evaluate patient pre and postoperatively. In addition, there are practical concerns related to contacting Avery Biomedical Devices customer service. If the providers need technical support should device malfunction, customer support is only available weekdays from 8 to 4pm EST.

Risk for direct surgical or procedural related destruction of device components should be minimized. Familiarity with device placement and background on surgical approach undertaken for their initial placement can aid in determining

if the patient is high risk for such injury. As a basic overview, the device has external and internal equipment. Starting with the external hardware, there is a "radio" transmitter powered by 9V batteries as well as an external antenna system (Figure 1&2).

Specific settings can be dialed into the transmitter for the patient's pulmonary mechanics and ventilatory needs, such as rate /amplitude, and this information is converted in the form of radio waves, and "transmitted" to internal hardware, transcutaneously. The internal devices include a radio receiver, which "receives" the radio signals and converts them into an electrical signal sent to the phrenic nerve via an electrode. Thus, there is a controlled, stimulation of each phrenic nerve, which leads to diaphragm contraction. This is unlike an automatic implantable cardioverter-defibrillator, which will sense the patients' electrical activity and can be considered a more "active" rather than "passive" system. The external transmitter is portable and the antenna is a flat, circular structure, directly connected to the transmitter that can be taped onto the surface of the patient above the radio receiver (Figure 1 &2,3). (4)

Two surgical approaches are typically undertaken when the device is initially implanted: cervical and thoracoscopic. The cervical approach involves neck incisions with electrode placement approximately at the anterior scalene muscle on the phrenic nerve and a subcutaneous pocket approximately 5 cm below the clavicle, where the receiver is placed. This procedure is repeated on the opposite side. The thoracoscopic approach involves a subcostal, subcutaneous pocket for receiver placement, and the electrode is placed in the chest at the phrenic nerve located at the cephalad aspect of the pericardium, where it is connected to the receiver. (6) If unsure of the exact location, radiographs of the area in question can show the path of the implanted devices.

After receiving general anesthesia, CCHS patients can often have significant decompensation of ventilatory control, which can occur without clinical signs of respiratory distress. This is not surprising as volatile anesthetics and opioids decrease the response to carbon dioxide, further impairing ventilation. Pulse oximetry and end-tidal carbon dioxide monitoring is important in the post-operative period, but monitoring devices using transthoracic impedance cardiorespiratory monitoring cannot be used because the pacemaker will cause artifact. (5) If hypoxemia is diagnosed from pulse oximetry, the likely etiology is hypoventilation. However, as CCHS is indeed a problem with hypoventilation, hypercarbia can occur in the setting of normal oxygen saturation. Hypoventilation after anesthesia can be severe, with arterial carbon dioxide levels rising to 70-80 mmHg and sometimes into the 100's if they do not receive ventilatory support with diaphragmatic pacemaker or mechanical ventilator. Patients with tracheostomies can be placed on ventilators until recovered from anesthesia. Even with the diaphragmatic pacing, without some central respiratory drive, ventilatory requirements may not be met, requiring assisted ventilation. It is possible that anesthetic agents such as volatile agents, and opioids can depress central and peripheral neuronal function depressing brainstem and phrenic nerve function resulting in hypoventilation even with a DP, as in this case. If muscle relaxant is used we recommend using a nerve stimulator to guide reversal, as inadequate reversal can lead to ventilatory compromise. If a patient were having any issues returning to baseline or requiring significant oxygen via ventilator in the PACU, and home ventilation is not available, it would be safest to admit the patient.

Several things can be considered to ensure that the device is working well postoperatively. First, confirm that the antennas are properly positioned over each receiver site, and firmly affixed to the patient's body via adhesive tape. Then, clinical evidence of the diaphragm contracting in sync with the light indicator system during inspiratory phase, along with the manual palpation of the chest, visualization of the costal margin, and respiratory excursion are important signs to consider. When the diaphragm contracts, a light on the machine will turn on. One should contact Avery customer service department and the patient's primary physician if additional questions or troubleshooting needs arise. Also, Avery provides trans-telephonic monitoring which can provide a quantitative report of the patient's physiologic responses to the implanted equipment settings. In addition, according to the manufacturers instruction manual, if you have an AM radio, you can also confirm device function by setting the radio to 1600 kHz, and placing it near the transmitter and listening for a burst of static with each stimulation when the transmitter is on. Finally, alkaline batteries, 9V rechargeable should be available for replacement. (4)

Families often receive a significant amount of education support with home care of an Avery Biomedical diaphragmatic pacemaker. In regards to adjusting the diaphragmatic pacemaker, caregivers can turn up the

amplitudes, when the patient is asleep. The tidal volumes could be adjusted via the amplitude adjustment to keep saturation > 94%, and maintain adequate chest rise at parental discretion. Parents are also instructed not to have the patient taken off the diaphragmatic pacer when they are sleeping even if they look clinically normal. Even in patients with normal saturation, the caregivers are often instructed to continue some form of ventilator support (pacer or ventilator) while asleep. When faced with acute respiratory infections, our patient utilized his portable home ventilator, the Pulmonetics LTV950, which was set at a respirator rate of 18, peak inspiratory pressure of 23 cm H2O, positive end expiratory pressure of 5 cm H2O, pressure support of 15 cm H2O, and inspiratory time 1-second, while on room air. Of note, minimally invasive methods of determining end tidal CO2 are available, including the EMMA ™ Mainstream Capnometry Device, which can digitally measure end tidal carbon dioxide, which was used for this patient.

Figure 1

Avery Biomedical Devices Mark IV diaphragmatic pacing system a. External transmitter system with antenna b. Illustrated details of control panel c. Implanted receiver and electrode *Image used with permission from Avery Biomedical Devices

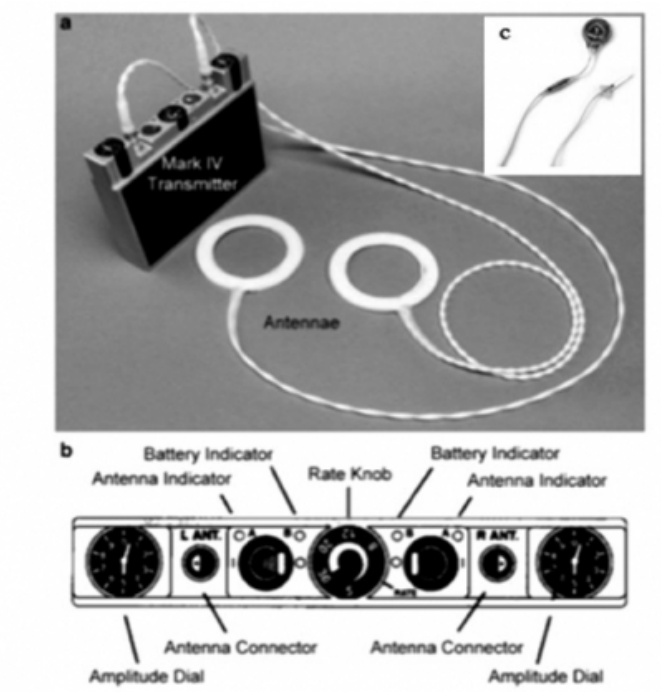
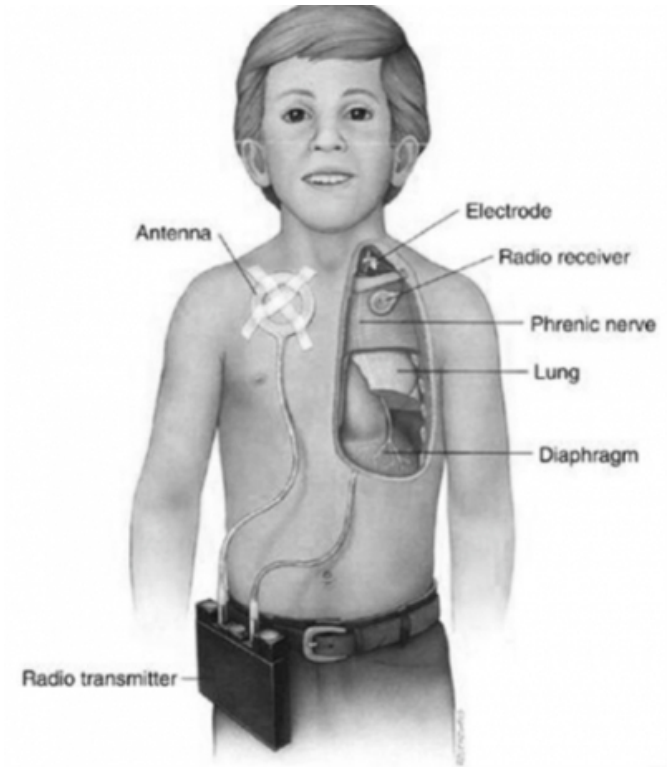


Figure 2

Avery Biomedical Devices Spirit Transmitter The Spirit Transmitter is the newest model of diaphragmatic pacer system. The transmitter is lightweight and features a backlight for nighttime viewing, clear graphic display, touch controls, and digital controls. *Image used with permission from Avery Biomedical Devices

Figure 3

Illustration of the Avery Biomedical Devices diaphragmatic pacer system in-vivo



SELECTED CAUTIONS

- Care when inserting jugular lines and performing regional nerve blocks (in particular, interscalene, supraclavicular, and infraclavicular), in those with cervical implants)

- Avoid surgery at or around device to decrease risk of direct hardware destruction
- Avoid drugs which can affect breathing or depress cell bodies in the anterior horn of the spinal cord (antispasmodics) such as baclofen (4)
- Amplitude adjustments may be needed depending on the patients position (sitting/supine) as indicated by measured tidal volumes, decreased blood gases, or patient discomfort or complaints (4)
- Failure of device could lead to respiratory arrest (4)
- Device failure can be from battery failure, broken battery connector wires, or cable failures (4)
- Component failure of the electrode wires, receivers, or external transmitter can lead to device failure (4)
- MRI, shockwave lithotripsy, and therapeutic diathermy are contraindicated (4)
- If a pacemaker is to be implanted, it must have bipolar leads and must be 10 cm from the pacemaker (4)
- The transmitter needs to be 1 meter away from oxygen enriched environments and within 1 meter away from flammable anesthetics (4)
- Defibrillators can damage internal components (4)
- Therapeutic ultrasound should be avoided (4)
- Close proximity to cell phones and Wi-Fi enabled devices should be avoided and kept more than 10 cm away (4)
- Signs of infection, anywhere in the body, including fever, redness, swelling and leukocytosis have shown diaphragmatic pacing to be ineffective and pacing at slightly higher amplitudes may be effective during treatment of infection (4)
- Electrocautery limitations: to prevent any issues during the surgery the device should be deactivated during the surgery and then re-activated once the patient has recovered from anesthesia (4)

CONCLUSION

In conclusion, knowledge of diaphragmatic pacemakers can help support safe perioperative decision making for patients with Congenital Central Hypoventilation Syndrome (CCHS) presenting for ambulatory surgery. Familiarity of the device components, being able to determine if the device is functioning normally, and minimizing risks factors that can lead to device malfunction, can reduce perioperative morbidity and mortality and aid in safe discharge planning for outpatient surgery.

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