Pediatric diaphragmatic pacing

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Abstract

Diaphragmatic pacing has been shown to play a significant role in adult patients with diaphragmatic paralysis and facilitates mechanical ventilation weaning. However, reports on its use in paediatric patients are scarce. This report is about a 4-year-old child with a spinal cord injury secondary to a motor vehicle accident that led to quadriplegia and diaphragm paralysis. The patient underwent a diaphragmatic pacing procedure, which helped start gradual weaning from mechanical ventilation. We reviewed the concept of the diaphragmatic pacer and its types depending on the site of the implantation. In addition, we reviewed who can benefit from using a diaphragmatic pacer and compared its use in adults versus paediatric patients. Our case showed that diaphragmatic pacing appears to be effective, as it facilitates mechanical ventilation weaning and improves the quality of life outcome in paediatric patients with diaphragmatic paralysis.

Introduction

Although a mechanical ventilator has beneficial outcomes over a long period of time, it can lead to recurrent respiratory infection, loss of the sense of smell, difficulties with moving or transferring patients, loss of the ability to talk, and a decrease in mobility that further negatively affects body functioning. Here, we present the concept of diaphragmatic pacing (DP) to overcome the use of a mechanical ventilator and to reduce its harmful effects in patients with diaphragmatic paralysis. DP uses electrical impulses to the phrenic nerve in order to produce artificial respiration, and it was a well-known concept in the 18th century although without clinical applications. This concept was then technically applied in the 19th century. A diaphragmatic pacer is defined as a device that uses electrical signals to stimulate the diaphragm to contract in order to restore normal breathing mechanisms and to reduce mechanical ventilation dependence in patients with diaphragmatic paralysis. A diaphragmatic pacer works by increasing tidal volume as much as possible by changing different characteristics of the breath cycle in a patient with diaphragmatic paralysis.

DP is classified into two groups depending on the implantation site. The device is either implanted exactly on the phrenic nerve or placed directly on the diaphragm muscle. Patients with respiratory insufficiency, spinal cord injury, stroke or any defect in the medulla mostly have beneficial outcomes from the diaphragmatic pacer. Other reported indications include chronic obstructive pulmonary disease (COPD), spinal muscular atrophy, idiopathic diaphragmatic paralysis, non-progressive neuromuscular diseases and chest deformities. The diaphragmatic pacer can be implanted for patients with spinal cord injury as well, but the level of injury can determine whether the patient is ready to have diaphragmatic pacer placement or not. Thus, patients with injury at the level of C3, C4 and C5 cannot be paced because of the effects on the phrenic nerve.

Diaphragmatic pacing has been shown to play a significant role in adult patients. Christopher et al. studied implantation of a diaphragm pacer in patients with different diseases and showed that they had a high survival rate and better quality of life. On the other hand, reports on paediatric patients with diaphragmatic paralysis who underwent implantation of a diaphragmatic pacer are not common. In addition, there are very few reports that compare paediatric cases with adult cases.

Case Report

Our case involves a 4-year-old child who was referred to our hospital in May 2016 for spinal surgery secondary to a motor vehicle accident 18 days before that resulted in upper cervical cord contusion with an injured apical ligament. This led to quadriplegia and diaphragm paralysis, for which the patient underwent posterior occipitocervical fixation.

After fixation surgery, the patient stayed in the Paediatric Intensive Care Unit (PICU) for a long time since he became ventilator-dependent secondary to his diaphragm paralysis. Then, he was moved to the high dependency unit with his home ventilator. He was completely quadriplegic without any signs of significant improvement; however, he was mentally intact and communicated by eye movement. Thus, a multidisciplinary medical decision had been made for diaphragmatic pacing, which was done 14 months following the primary injury. Weaning from the mechanical ventilator was not smooth due to a disconcerting sensation from pacing pulses during the conditioning phase, and so the pacing device was set at low amplitude initially, 10 mA (milliampere), and pulse width of 110 µS (microseconds), after which it was increased gradually until it reached an amplitude of 20 mA and pulse width of 150 µS. These steps were associated with weaning from a mechanical ventilator by increasing the free ventilator time gradually until it reached 8 hours in the daytime.

Currently, he is on a pacer for only eight hours in the daytime and on a home ventilator without the pacer for the remaining 16 hours. The medical team’s plan was to increase pacing time gradually.

Discussion

To our knowledge, the majority of the studies on DP in the literature review are related to adults and not paediatric patients. Regarding studies that have been reviewed,
most adult patients tolerate diaphragmatic pacing and can be easily weaned off the mechanical ventilator in a short time. In general, outcomes of DP in adults are better than those in paediatric patients. Oakes DD performed a study on 11 adult patients with implantation, of which 3 became completely independent of their ventilators in three months.11 Weaning from the mechanical ventilator in paediatric patients was not smooth, and most of the patients reached the maximum 10 hours off the ventilator, which occurred one year after the procedure.8

Looking at our case, we started with continuous pacing initially with gradual ventilator weaning by increasing the ventilator off-time. Due to intolerance of the electrical sensation produced by the pacer and sleeping disturbance, the pacer was only used during the daytime and the weaning strategy was started with 5 minutes only. Then, this time was increased gradually while only the ventilator was used for the remaining time. Week by week, this time was increased until a time of 8 hours off the ventilator with pacing was reached, and the patient shifted again to the mechanical ventilator for the rest of the day. This weaning achievement took five months after the procedure. With gradual weaning from the mechanical ventilator, we look forward to achieving our goal to liberate patients completely from ventilator dependency.

Conclusions

Our report demonstrates the safety of diaphragmatic pacing (DP) in children, which appears to be effective in patients with diaphragm paralysis. In addition, we found that DP facilitates mechanical ventilation weaning and improves quality of life in paediatric patients. Nonetheless, further studies are needed to validate the long-term safety and outcome of DP.

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