Case report

Diaphragmatic paralysis after pediatric heart surgery: Usefulness of non-invasive ventilation

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1. Introduction

Diaphragmatic paralysis (DP) due to phrenic nerve injury after congenital cardiac surgery is an important respiratory complication resulting in respiratory insufficiency, lung infections, and death [1–3]. It should be considered as a severe and life-threatening respiratory complication especially in younger age. DP causes respiratory distress because of paradoxical motion of the affected diaphragm and contralateral shift of the mediastinum [1,2]. The common feature of DP after cardiac surgery in children is inability in weaning from mechanical ventilation or requirement of reintubation. This situation is the major cause of lung infections and related complications such as atelectasis and recurrent pneumonia [1,3].

Recently, non-invasive positive pressure ventilation (NPPV) has been applied to various types of respiratory failure, but data on the use of NPPV in pediatric patients with DP are limited [4–6]. This report illustrates the usefulness of invasive and non-invasive ventilatory support for a pediatric patient with diaphragmatic paralysis.

2. Case

A 5-year-old girl had an operation for repair of total anomalous pulmonary venous return when she was 9.5 months old. The patient had hypoxemia and respiratory failure in 1 h after extubation and she needed ventilatory support. She had a tracheostomy in order to avoid complications of prolonged tracheal intubation on postoperative day 20. Since the patient needed ventilatory support at the end of 3 months, she was referred to our hospital. The patient was diagnosed with ventilator associated pneumonia once during the postoperative period until the application of diaphragmatic plication. Chest radiography and computed tomography showed a raised right diaphragm, right middle and right lower lobe atelectasis (Fig. 1). Paradoxical diaphragmatic movement was seen during fluoroscopy and DP was diagnosed. Diaphragmatic plication was performed 3.5 months after the cardiac surgery. Although patient's oxygen requirement and pressure support decreased after the plication, right middle and right lower lobe atelectasis persisted and the patient could not tolerate weaning from ventilator. Thus, Bilevel Positive Airway Pressure (BIPAP, Respironics, Pittsburgh, PA, USA) via tracheostomy was started (inspiratory support pressure: 14 cmH2O and expiratory support pressure: 4 cmH2O). In order to avoid possible complications related with tracheostomy, NPPV was initiated with BIPAP via nasal mask. Patient had agitation during the first hours of the NPPV but no sedation was required. To optimize the patient's...
cooperation, the mask was initially applied manually, on to the patient's nose. After a short adaptation period; it was firmly applied on the nose by a pediatric head cap (respironics). Respiratory rate, oxygen saturation, capillary blood gas and oxygen requirement were recorded immediately before the initiation of NPPV and 1, 6, 12, and 24 h after NPPV to evaluate the effectiveness of ventilation. Since NPPV was applied intermittently, the patient could eat by herself during the off periods of NPPV. Since she had persistent atelectasis in the right middle and right lower lobe, flexible bronchoscopy was performed at the 17th day of NPPV application, which revealed mild bronchomalasia in the right main bronchus. She needed invasive ventilatory support for 6.5 months and NPPV for ten months. The total duration of ventilatory support was 16.5 months. NPPV via nasal mask was continued for another 10 months after the surgery. During follow-up, atelectasis regressed, she did not have a major respiratory problem and tolerated weaning at the end of the 10th month. She was followed up in our clinic with control chest radiographies in the last two years without any complications.

3. Discussion

Extubation failure is one of the problems in children following cardiac surgery. Respiratory system is a major system contributing to this postoperative morbidity, and four major groups could be identified in this setting: diaphragmatic dysfunction, airway obstruction, pulmonary hypertension, and pleural effusion [7–9]. The incidence of unilateral DP after pediatric cardiac surgery is reported to be 1.9% [3]. Infants and children younger than 2 years of age are at particular risk for development of respiratory failure because of the high compliance of the thoracic cage, the relatively weak intercostal musculature, and the mediastinal shifts induced by paradoxical motion of the paralyzed diaphragm [10]. Thus, DP in this age group causes severe respiratory failure and sometimes requires tracheotomy and prolonged mechanical ventilation [3,10]. Optimal management of DP in children who have undergone cardiac surgery remains controversial and consists of prolonged ventilation or diaphragmatic plication. While early plication may reduce ventilation time and hospital stay long-term follow-up of these patients demonstrates that only a small proportion shows normal diaphragmatic movement [3,8,11,12].

In this case we report a child who developed DP and respiratory failure following cardiac surgery. Since the child had bronchomalasia and atelectasis in addition to DP, plication alone did not solve the problem and she needed long-term ventilatory support. Our patient continued to receive ventilatory support for 10 months after the surgery. Patients who need a long-term ventilatory support can be ventilated invasively through tracheostomy or noninvasively via nasal or oronasal masks [13]. NPPV offers some advantages in comparison to invasive ventilation. The major advantages of NPPV, compared to invasive ventilation with tracheostomy, include greater patient comfort, easier application, use, and care, reduced incidence of complications, decreased hospital cost and decreased sedation requirement. NIV provides a technique more amenable to domiciliary use. However, there are conditions where NPPV neither safe nor practical as swallowing disorders, history of pulmonary aspiration secondary to gastroesophageal reflux and or vocal cord paralysis, inability to tolerate NPPV, failure to adequately ventilate with NPPV, high level of dependence on assisted ventilation (16–20 h/day) [14,15].

Our patient was at an age when she could learn to talk we also wanted to avoid possible complications related with tracheostomy such as infections or swallowing difficulties and decided to continue ventilation via nasal mask.

In conclusion; in young children undergoing surgical repair of cardiac lesions, increased postoperative morbidity is clearly associated with pulmonary involvement, especially when phrenic nerve dysfunction and/or central airway compression are present. Thus, early evaluation for these pulmonary conditions should be considered in any postoperative patient who fails extubation. NPPV can be preferred in patients who need long-term ventilatory support.

References