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Bilateral Diaphragmatic Paralysis After Cardiac Surgery: Ventilatory Assistance by Nasal Mask Continuous Positive Airway Pressure

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Abstract. The case of an 8-month-old boy with bilateral diaphragmatic paralysis after surgical reoperation for congenital heart disease is presented. In order to avoid repeated intubation and long-term mechanical ventilation or tracheotomy, we used nasal mask continuous positive airway pressure (CPAP) as an alternative method for assisted ventilation. Within 24 hours the boy accepted the nasal mask and symptoms such as dyspnea and sweating disappeared. Respiratory movements became regular and oxygen saturation increased. Nasal mask CPAP may serve as an alternative treatment of bilateral diaphragmatic paralysis in infants, thereby avoiding tracheotomy or long-term mechanical ventilation.

Key words: Bilateral diaphragmatic paralysis — Respiration — Nasal mask CPAP

Unilateral phrenic nerve injury is a recognized complication of palliative or corrective cardiac surgery. The incidence of unilateral phrenic nerve paralysis in a retrospective study in children was 1.9% for open-heart and 1.3% for closed-heart operations and was more common in children requiring reoperation [9]. Spontaneous recovery of unilateral diaphragmatic function was documented in 90% of patients with postoperative paralysis of diaphragm, whereas the mean time of diaphragmatic recovery was 40.8 days and was more prolonged in patients with paradoxical as opposed to absent diaphragmatic movement [4].

Treatment of unilateral diaphragmatic dysfunction includes diaphragmatic plication [8], rocking bed [1], negative extrathoracic pressure ventilation [6], and endotracheal intubation with continuous positive airway pressure (CPAP) [5]. Because infants and small children breath mainly with the diaphragm, bilateral phrenic nerve paralysis in this age group causes severe respiratory distress and usually requires tracheotomy and prolonged mechanical ventilation [7].

The use of nasal mask CPAP instead of tracheotomy is recommended in children for the treatment of obstructive sleep apnea [3, 10], but there is no experience in children with unilateral or bilateral phrenic nerve paralysis.

This case report discusses nasal mask CPAP as an alternative treatment of bilateral diaphragmatic dysfunction after surgery for congenital heart disease in an 8-month-old boy.

Case Report

Our patient was the first-born son of a couple with an unremarkable family history; the mother was 36 years old. Ultrasound examination during pregnancy was interpreted as truncus arteriosus; amniocentesis was not performed. The child was born at 39 weeks of gestation (Apgar score of 8, 10, and 10 at 1, 5, and 10 minutes, respectively). Birth weight was 3360 g (50th percentile). Postnatal echocardiography showed a double-inlet left ventricle, L-transposition of the great arteries with subaortic right ventricular outlet chamber, and nonrestrictive foramen bulboventriculare, pulmonary atresia, and ductus-dependent pulmonary flow.

Two hours postnatally the boy was intubated because of low oxygen saturation, and a prostaglandin E_1 infusion was started immediately. A Rashkind balloon atrial septostomy followed at the age of 48 hours and at 9 days an aortopulmonary shunt to the right pulmonary artery was carried out. The postoperative course was uneventful and the boy was extubated after 4 days. Oxygen saturation without supplementation was 85%. Echocardiography showed bilateral diaphragmatic movement. Digoxin, frusemide, and spironolactone therapy was initiated and the boy, breast-fed, was discharged home at the age of 4 weeks.

Echocardiographic examination 4 weeks postoperatively showed that the ductus arteriosus had closed spontaneously and left pulmonary stenosis had developed at the insertion site with an uneven pulmonary perfusion in perfusion scintigraphy (right 86%, left 14%). The weight was 4 kg (50th percentile). Cardiac catheterization at the age of 4.5 months confirmed the former cardiac diagnosis, an intact aortopulmonary shunt and marked stenosis of the left pulmonary artery. Subsequent palliative surgery consisted of a bidirectional cavopulmonary anastomosis, take down of the aortopulmonary shunt, and patch augmentation of the stenosed left pulmonary artery.

After this operation, attempts to wean the boy from mechanical

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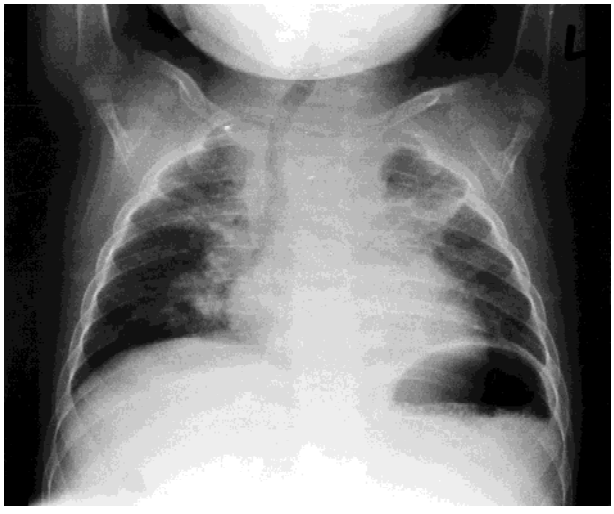

Fig. 1. Chest x-ray with collapse, consolidation, and bilateral elevated hemidiaphragm.

respiration failed. The clinical suspicion of bilateral phrenic nerve paralysis was confirmed radiologically. Electromyography exhibited a failure of diaphragmatic response to phrenic nerve stimulation on both sides. Bilateral diaphragm plication was recommended, but the parents declined.

Mechanical ventilation was carried out for 11 weeks. After this period diaphragmatic movement on the left side was observed ultrasonographically. Because of continuous dysfunction of the right diaphragm a plication was carried out. Four days after diaphragm plication the boy was extubated. Oxygen saturation without supplementation was 80% and the boy was discharged home.

At the age of 8 months (6.3 kg, 0.5 kg < 3rd percentile) cyanosis, sweating, and dyspnea with use of accessory respiratory muscles increased and the boy was admitted to our intensive care unit. Ultrasound showed diaphragm paralysis on both sides. Chest x-ray showed collapse, consolidation, and bilaterally elevated hemidiaphragm (Fig. 1).

In order to analyze thoracic and abdominal respiratory movement, cardiorespiratory polygraphy was performed. We recorded thoracic and abdominal motion by means of inductive plethysmography and oxygen saturation and heart rate by pulse-oxymetry, thus confirming paradoxical breathing, increased thoracic effort, and an oxygen saturation of 70% (Fig. 2A).

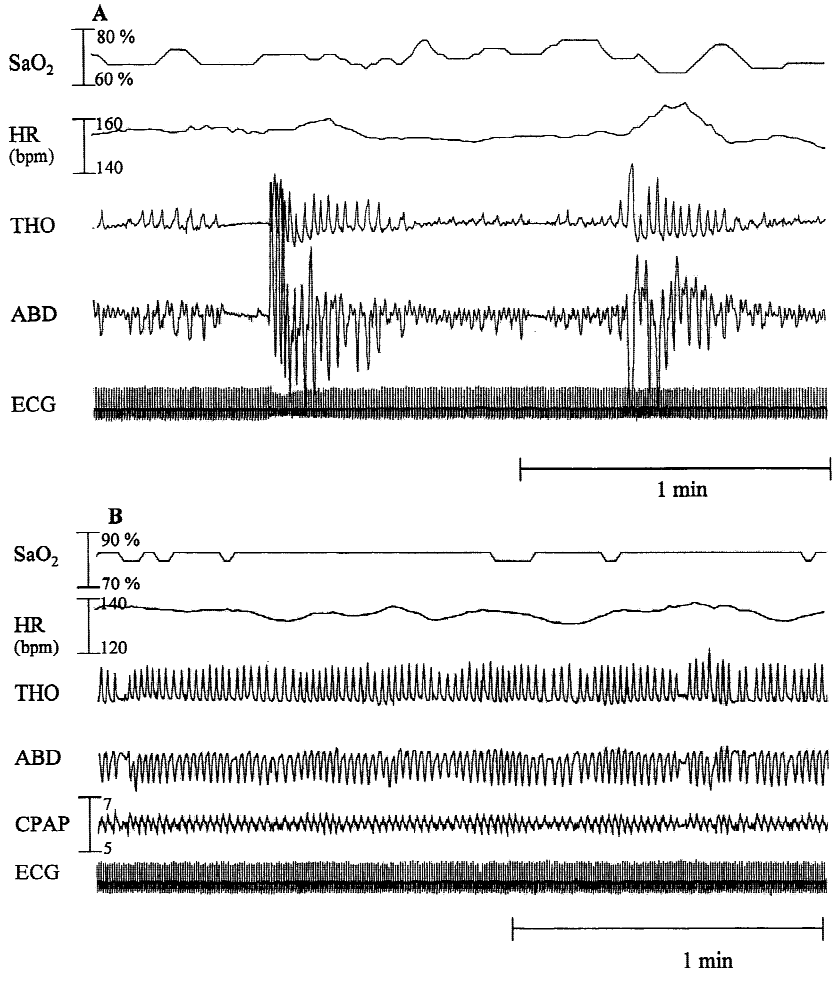

Fig. 2. A section of a cardiorespiratory polygram recorded in our patient before and during nasal mask CPAP. (**A**) Irregular paradoxical inward movement of the chest and abdomen. Apneic pauses, hyperventilation with high breathing amplitudes and oxygen saturation between 60% and 80%, and heart rate above 150 bpm. (**B**) Breathing movements with nasal mask CPAP, PEEP (positive end-expiratory pressure) 6.5 cm H_2O. Regular paradoxical breathing movements, oxygen saturation between 80% and 90%, and heart rate between 130 and 140 bpm. *ABD*, abdominal motion; *bpm*, beats per minutes; *CPAP*, continuous positive airway pressure; *HR*, heart rate; *SaO_2*, oxygen saturation; *THO*, thoracic motion.

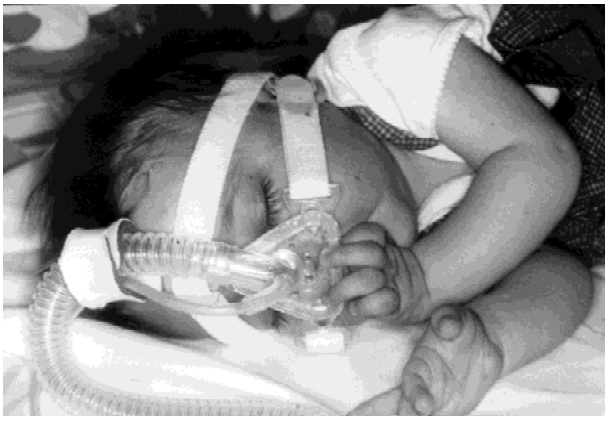

Fig. 3. Nasal mask CPAP as assistance ventilatory therapy in our patient during sleep.

Together with the parents we decided against renewed intubation and commenced ventilatory assistance via a nasal mask CPAP day and night. Within 24 hours the boy accepted the nasal mask with a PEEP of 6.5 cm H_2O (Fig. 3).

Dyspnea and pulmonary symptoms disappeared. Polygraphy with nasal CPAP pressure recording showed paradoxical but regular respiratory movement, $SaO_2 > 80\%$ (Fig. 2B). Repeated ultrasound scanning confirmed the absence of diaphragmatic movement. After 4 weeks of successful nasal mask CPAP therapy our patient thrived and could be discharged home with a weight of 7.9 kg (>3rd percentile).

Discussion

We reported on an 8-month-old boy with congenital heart disease, repeated heart operations, bilateral phrenic nerve paralysis, long-term mechanical ventilation, unsuccessful plication of the right diaphragm, and nasal mask CPAP as ventilatory assistance in bilateral diaphragmatic paralysis.

Diaphragmatic paralysis in children who require multiple cardiac surgery is more common than in those who require a single operation [9]. Ultrasonography and cardiorespiratory polygraphy are useful diagnostic means of establishing the presence of paradoxical respiratory movement and for deciding management [4]. The efficacy of treatment with endotracheal intubation and CPAP has been confirmed [4], but mechanical ventilation in patients less than 2 years of age with phrenic nerve paralysis after cardiac surgery may be necessary for several months [9]. Long-term mechanical ventilation causes pulmonary problems such as nosocomial lung infections, pleural atelectases, infiltrations, and pneumonia.

In previously described patients with bilateral phrenic nerve paralysis, mechanical ventilation was required in all 11 patients, 5 of which underwent tracheotomy [2]. Tracheotomy was preferred to bilateral diaphragmatic plication because it reduced the risk of an obstructed endotracheal tube, allowed immediate oral intake, and simplified the weaning process from the ventilator [7].

Diaphragmatic plication in patients with unilateral or bilateral diaphragmatic dysfunction is safe, reliable, and of most value in this age group [9]. In our case diaphragm plication of the right side was unsuccessful. The paralysis of the left side initially recovered but subsequently recurred. We believe that the secondary dysfunction of diaphragm may have been caused by scarring. In order to avoid renewed mechanical ventilation and tracheotomy (which the parents declined), we started an alternative treatment of ventilatory assistance with nasal mask CPAP. This therapy is an effective treatment for infants with upper airway respiratory problems during sleep [3]. The compliance of the boy and of the parents was not problematic, the pulmonary symptoms disappeared, breathing movements became regular, the boy's weight increased, and he became more interested in his environment.

Conclusion

We demonstrate nasal mask CPAP as an effective alternative treatment to mechanical respiratory support in a boy with bilateral phrenic nerve paralysis, thereby avoiding tracheotomy.

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