

# Role of non-invasive ventilation in difficult-to-wean children with acute neuromuscular disease

V G Reddy, M P Nair, F Bataclan

## ABSTRACT

Weaning from mechanical ventilation in children could be time-consuming and on many occasions, leads to reintubation with its associate complications. We report two children with acute neuromuscular disease, in whom bi-level positive airway pressure (BiPAP) as a mode of non-invasive ventilation was successfully used to wean the child from ventilators and prevented the need for tracheostomy. Despite the limited number of studies published in the literature suggesting BiPAP as a mode of weaning from mechanical ventilation, the technique when applied correctly seems to be safe and effective in weaning and avoiding tracheostomy.

**Keywords:** acute disseminated encephalomyelitis, bi-level positive airway pressure, Guillain-Barré syndrome, neuromuscular disorder, non-invasive ventilation

*Singapore Med J 2004 Vol 45(5):232-234*

## INTRODUCTION

Bi-level positive airway pressure (BiPAP), a form of non-invasive ventilation (NIV), involves the delivery of mechanically-assisted breaths without the use of an endotracheal or tracheostomy tube. NIV preserves normal swallowing, speech and cough mechanisms. Currently, ventilation is delivered via either a nasal mask or facemask. NIV as a mode of ventilation has been used in children with acute respiratory failure<sup>(1,2)</sup>. Children with neuromuscular disorders presenting with acute respiratory failure usually require long-term ventilation. Extubation is sometimes difficult and prolonged, requiring tracheostomy. Tracheostomy in children has been associated with an overall complication rate as high as 40%<sup>(3)</sup>. We describe our experience with the use of BiPAP as a form of NIV in two cases, where BiPAP was successfully applied to wean the children from ventilators and avoidance of tracheostomy.

## CASE REPORTS

### Case 1

An 11-year-old girl was admitted to the paediatric intensive care unit with a diagnosis of acute Guillain-Barré syndrome. On the third day of admission, she became tachypnoeic (48/min) and tachycardic (140/min). Arterial blood gas showed pH of 7.2 (range 7.35-7.45), PaCO<sub>2</sub> of 8 kPa (range 4.5-6 kPa), PaO<sub>2</sub> of 7.45 kPa (range 11-12.5 kPa), and oxygen saturation of 80% (range 90-99%) on breathing 60% oxygen via a Venturi mask. At the same time, there was absence of her gag reflex. She was intubated under anaesthesia, and mechanically-ventilated in volume control mode using a Siemens Servo 900C ventilator with a FiO<sub>2</sub> of 0.45. She received intravenous immunoglobulin (IVIg) 400mg/kg for five days. Her parents refused to give consent for an elective tracheostomy. After two weeks of uncomplicated ventilation, the ventilatory mode was changed to synchronised intermittent mandatory ventilation with pressure support (SIMV rate of 12/min, PS of 8cm H<sub>2</sub>O, PEEP 3cm H<sub>2</sub>O). After 16 days of SIMV mode, the ventilatory support was gradually decreased to a SIMV rate of 5/min, PS of 7cm H<sub>2</sub>O, PEEP 7cm H<sub>2</sub>O alternating with spontaneous ventilation via a T-piece, as long as she tolerated the T-piece without respiratory distress and maintained oxygen saturation above 92%.

On the 34th day, after two hours of T-piece trial, she was extubated. She received 40% oxygen by mask while breathing spontaneously. Within two hours of extubation, she complained of difficulty in breathing and became tachycardic (148/min), tachypnoeic (rate 46/min), and hypoxic (7.1 kPa) and hypercarbic (PaCO<sub>2</sub> 9.8 kPa) while breathing 100% oxygen. Direct laryngoscopy under general anaesthesia revealed minimal laryngeal oedema, which could not explain the severity respiratory insufficiency. She was reintubated with a smaller endotracheal tube. After 24 hours, extubation was attempted again but she became tachypnoeic (rate 52/min), hypercarbic (PaCO<sub>2</sub> 8.8 kPa) and hypoxic (PaO<sub>2</sub> 7 kPa) while breathing 100% oxygen.

In an attempt to avoid further airway trauma from tracheal intubation or the need for tracheostomy,

Department of  
Anaesthesia  
and Intensive  
Care Unit  
College of Medicine  
Sultan Qaboos  
University Hospital  
PO Box 35,  
PC 123-SQU  
Muscat, Sultanate  
of Oman

V G Reddy, MD,  
EDIC, FCARCSI  
Associate Professor  
and Consultant

Department of  
Child Health

M P Nair, MD,  
DNB, DM  
Consultant  
in Neonatology

F Bataclan, MD  
Registrar

Correspondence to:  
Dr Venu Gopal Reddy  
Tel: (96) 8 513850  
Fax: (96) 8 513850  
Email: venu@  
squ.edu.om

the decision was made to use NIV, applied through a nasal mask using a pressure-cycled ventilator in spontaneous mode (BiPAP, Respironics Inc, Murrysville, Pennsylvania, USA). After carefully explaining the procedure, the child was asked to hold the facemask and to breathe. Initially the inspiratory positive airway pressure (IPAP) was set at 5cm H<sub>2</sub>O, expiratory positive airway pressure (EPAP) at 3cm H<sub>2</sub>O, with 2L/min of oxygen to maintain oxygen saturation above 90%. The mask was fixed with the help of headgear, and the IPAP and EPAP were gradually increased to 10cm and 4cm H<sub>2</sub>O, respectively, until the child felt comfortable, generated a tidal volume of 270-300 mL per breath, and with adequate chest expansion and satisfactory blood gases.

Subsequently, the mode was changed to spontaneous/timed mode (14-BPM). Arterial blood gas taken at 30 minutes showed normal PaCO<sub>2</sub> (5 kPa) and PaO<sub>2</sub> (13 kPa). She remained stable, and was able to eat and communicate verbally. During the daytime, she was gradually allowed to breathe on her own for an average of one hour, after which she would receive BiPAP. This mode was continued over the next six days and during this period, IPAP was gradually decreased during the daytime until she was able to breathe on her own without inspiratory pressure support. On the 6<sup>th</sup> day, she could breathe spontaneously for the whole day without the help of BiPAP but required the assistance of the ventilator during the night. On the 8<sup>th</sup> day, she was completely liberated from the ventilator. She was transferred to the ward on the 9<sup>th</sup> day, and discharged satisfactorily on the 12<sup>th</sup> day.

## Case 2

A 7-year-old girl was referred to the paediatric intensive care unit from a peripheral hospital with a diagnosis of acute disseminated encephalomyelitis and bronchopneumonia. Magnetic resonance (MR) imaging of the spine revealed oedema and swelling of the spinal cord (C1-T2 levels) with no extrinsic compression. On admission, she was in respiratory failure (tachypnoea 56/min, PaCO<sub>2</sub> 8.8 kPa, PaO<sub>2</sub> 7.5 kPa, oxygen saturation 77% with 100% oxygen). She received intravenous immunoglobulin (400mg/kg/day for five consecutive days). She was electively intubated and ventilated in pressure control mode using a Siemens Servo 900C ventilator. Repeat MR imaging after one week revealed extension of the lesion from the medulla to T8 level. Mechanical ventilation was continued for 28 days and when she showed signs of respiratory improvement, the ventilatory mode was changed to synchronised intermittent mechanical ventilation with pressure support (SIMV of 10 and with PS of 8cm H<sub>2</sub>O, PEEP 3cm H<sub>2</sub>O). Any attempt at

decreasing the SIMV rate or PS resulted in respiratory distress and hypoxia. A tracheostomy was contemplated at this time but her parents refused to give their consent.

Over the next five days, mild cough reflex during suctioning of the endotracheal tube was noticed. On the 33<sup>rd</sup> day, a decision was made to use non-invasive ventilation using BiPAP. After carefully explaining and demonstrating the procedure, the child was asked to hold the facemask and to breathe. The IPAP was set at 5cm H<sub>2</sub>O, EPAP at 3cm H<sub>2</sub>O with 1L/min of oxygen to maintain her oxygen saturation above 90%. Initially, she refused to accept the facemask, but subsequently relented with constant encouragement. The mask was fixed with the help of headgear and the IPAP and EPAP was gradually increased to 11cm and 5cm H<sub>2</sub>O, respectively, until the child felt comfortable, generated a tidal volume of 175mL per breath, and chest expansion was adequate with satisfactory blood gases. No sedation was prescribed. Three hours later, the facemask was replaced by a nasal mask.

At night, the mode was changed to spontaneous/timed mode (16BPM) as a back-up. She was continuously monitored for heart rate, respiratory rate, blood pressure, oxygen saturation and arterial blood gases. From the 6<sup>th</sup> day, the levels of support were gradually decreased and the child was allowed to breathe spontaneously without the help of the ventilator until she would ask for the ventilator. She was completely removed from BiPAP on the 8<sup>th</sup> day, successfully transferred to the ward on the 9<sup>th</sup> day, and discharged from the hospital on the 15<sup>th</sup> day.

## DISCUSSION

Respiratory failure occurs when pulmonary gas exchange is sufficiently impaired to cause hypoxemia, with or without hypercarbia. Depending upon the severity of respiratory failure, some children would require mechanical ventilation. BiPAP refers to a technique of augmenting alveolar ventilation without the help of an endotracheal tube. In BiPAP, a facemask or nasal mask is used to deliver the ventilation. Use of a nasal mask rather than a facemask allows easier access to clear secretion, improves patient comfort, and permits the child to cough, speak, eat, and play. Nasal masks are not very efficient in mouth-breathers or in some cases, mouth leakage may occur during sleep. BiPAP ventilation reduces the work of breathing by delivering an inspiratory pressure and by decreasing the expiratory work of breathing by providing end-expiratory pressure<sup>(4)</sup>. BiPAP has the unique advantage of independent control of inspiratory and expiratory pressures.

IPAP supports inspiration by augmenting the tidal volume. The EPAP prevents airway/alveolar collapse

during exhalation, improves oxygenation, and prevents rebreathing of CO<sub>2</sub>. Tracheostomy in children has been associated with an overall complication rate as high as 40%<sup>(3)</sup>. In one report, 13 patients with the Guillain-Barré syndrome had tracheostomy and four patients had complications directly related to tracheostomy<sup>(5)</sup>. Paediatric applications of BiPAP ventilation have focused primarily on patients with chronic respiratory failure. In critically-ill paediatric patients, NIV has been applied with success in acute respiratory failure in order to avoid intubation, improve gas exchange, and prevent atelectasis<sup>(4)</sup>.

There continues to be a paucity of literature on the use of BiPAP as a mode of weaning in children<sup>(4)</sup>. Niranjana and Bach<sup>(6)</sup> reported the successful use of NIV in children affected with Duchenne muscular dystrophy who required ventilatory support. Another study from the same group of authors reported the successful use of NIV after extubation in children with spinal muscular atrophy<sup>(7)</sup>. Both studies demonstrated the safety of NIV as an alternative to tracheostomy. In both reports, the children were affected with chronic neuromuscular disease rather than acute neuromuscular disease, as in our case. The extubation was protocol-driven, and carried out only when the child required no supplemental oxygen to maintain a saturation of more than 94% and improvement of the chest radiographical abnormality. Even though BiPAP has been successfully used for weaning from prolonged mechanical ventilation, our cases are unique in that we describe for the first time in children with acute neuromuscular disorder, the efficacy of BiPAP as a mode to wean from ventilator and to avoid tracheostomy.

Evidence from recent literature suggests that protocol-directed extubation is an useful approach to liberate patients from mechanical ventilation. Protocol-driven weaning reduces the number of days on mechanical ventilation, better patient outcomes, increased safety, and cost savings<sup>(8,9)</sup>. On the other hand, we had no protocol to wean. We believe a T-piece trial of two hours of spontaneous breathing is sufficient to decide extubation and discontinuation of mechanical ventilation<sup>(10)</sup>. The success of NIV depends upon acceptance by the child, age of the child, and familiarity and experience of the clinician in using the ventilator. We preferred the spontaneous mode since the respiratory rate was adequate and the mode synchronised with patient's respiratory effort.

We initially encouraged both children to hold the facemask themselves to get a "feel" of it before applying the head strap. Children may refuse to accept the facemask or nasal mask. The child's co-operation is extremely important in order to obtain a high success rate with NIV. Repeated assurance and demonstration of the comfort of facemask or nasal masks compared to endotracheal tubes usually helps overcome the fear. We believe that close contact between the parent and the attending doctor is more important than prescribing sedation. The specific side-effects related to the use of NIV include skin breakdown commonly over the bridge of the nose, nasal dryness, conjunctivitis, air leak, gastric distension, and aspiration of the gastric contents. It has been noted that gastric distension is uncommon in the paediatric age group<sup>(1,2,4)</sup>.

#### ACKNOWLEDGEMENT

We would like to gratefully acknowledge Dr Bhaskar Ramachandran, Consultant Anaesthetist, Bahrain, who critically reviewed the manuscript and offered invaluable suggestions.

#### REFERENCES

1. Marino P, Rosa G. Treatment of acute respiratory failure by prolonged noninvasive ventilation in a child. *Can J Anaesth* 1997; 44:727-31.
2. Padman R, Lawless ST, Ketrwick RG. Noninvasive ventilation via bilevel positive airway pressure support in paediatric practice. *Crit Care Med* 1998; 26:169-73.
3. Donnelly MJ, Lacey PD, Maguire AJ. A twenty-year (1971-1990) review of tracheostomies in a major pediatric hospital. *Pediatr Otol Rhinol Laryngol* 1996; 35:1-9.
4. Fortenberry D, Del Toro J, Jefferson LS, Evey L, Haase D. Management of paediatric acute hypoxemic respiratory insufficiency with bilevel positive pressure (BiPAP) nasal mask ventilation. *Chest* 1995; 108:1059-64.
5. Gracey DR, McMichan JC, Divertie MB, Howard FM Jr. Respiratory failure in Guillain-Barre syndrome: a 6-year experience. *Mayo Clin Proc* 1982; 57:742-6.
6. Niranjana V, Bach JR. Noninvasive management of pediatric neuromuscular ventilatory failure. *Crit Care Med* 1998; 26:1952-3.
7. Bach JR, Niranjana V, Weaver B. Spinal muscular atrophy type 1: a non-invasive respiratory management. *Chest* 2000; 117:1100-5.
8. Grap MJ, Strickland D, Torney L, Keane K, Lubin S, Emerson J, et al. Collaborative practice: development, implementation, and evaluation of a weaning protocol for patients receiving mechanical ventilation. *Am J Crit Care* 2003; 12:454-60.
9. Chan PKO, Fischer S, Stewart TE, Hallett DC, Patricia HG, Lapinsky SE, et al. Practicing evidence-based medicine: the design and implementation of a multidisciplinary team-driven extubation protocol. *Crit Care* 2001; 5:349-54.
10. Esteban A, Alia I, Gordo F, Fernandez R, Solsona JF, Vallverdu I, et al. Extubation outcome after spontaneous breathing trials with T-tube or pressure support ventilation. The Spanish Lung Failure Collaborative Group. *Am J Respir Crit Care Med* 1997; 156:459-65.