**Special Article**

**Negative Pressure Ventilation in Pediatric Critical Care Setting**

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**ABSTRACT**

Invasive ventilation is associated with both pulmonary and non-pulmonary complications. There has been a renewed interest in the use of negative pressure ventilation (NPV) for various medical conditions to minimise the complications associated with positive pressure ventilation. The routine use of NPV in an ICU setting still requires further studies and research. In this article, the authors review the clinical applications of NPV together with associated risks and limitations. Case reports of patients with cardiac, neuromuscular, and respiratory diseases managed with NPV on our unit are described. NPV improved the clinical condition in each of these patients and decreased the requirement for invasive therapy. [Indian J Pediatr 2007; 74 (5) : 483-488] E-mail: aakashdeeparora@hotmail.com

**Key words :** Non-invasive ventilation; Negative pressure ventilation; Chest cuirass ventilation; Continuous negative pressure ventilation; Continuous negative extra-thoracic pressure (CNEP) ventilation

Acute hypoxemic respiratory failure is an important cause of mortality and morbidity in children admitted to pediatric intensive care units (PICU). The standard strategy for treating children in respiratory failure is by invasive mechanical ventilation which can be associated with a number of pulmonary and extra-pulmonary complications. Non-invasive ventilation (NIV) which includes negative pressure ventilation (NPV), and positive pressure ventilation (PPV) via nasal continuous positive airway pressure (nCPAP) can be used to reduce the need for endotracheal intubation and associated complications. Experience in NIV in pediatric population has been limited to small case series and anecdotal reports. NPV was the primary mode of ventilation during polio epidemic. However, limitations of negative pressure ventilation lead to the widespread use of positive pressure ventilation including non-invasive PPV. But as more invasive ventilation was being used, the existence of complications became clear. These complications justified a renewed interest in NPV and with advancing technology ventilators that could provide negative pressure ventilation around a negative pressure baseline with the ability to actively control expiration and respiratory frequency produced a versatile alternative strategy for the intensive care physician. This article aims to summarise some practical aspects of initiating and maintaining NPV in the PICU for acute respiratory failure by describing its use on some of the patients hospitalised at St Mary’s Hospital PICU. It also provides a review of the published evidence related to NPV in children.

**Negative pressure ventilators :** NPV was initially started in 1950s using iron lung. However problems with the iron lung including its size, weight, accessibility of the patient and pooling of venous blood within the abdomen lead to invention of various improvised versions. Hayek oscillator was used later in which high frequency oscillation is applied by using an airtight cuirass placed around the chest wall. The cuirass is flexible with foam seals to fit around chest and abdomen. Different sizes of cuirass are available ranging from preterm newborn babies to adult patients. A jacket must fit well around the chest, but not too tight. There should be no leaks. It is designed to provide continuous negative pressure, or controlled ventilation including high frequency chest wall oscillation.

The RTX Respirator is an improved version of the original Hayek oscillator. There are seven modes of ventilation including cardiac synchronised mode and controlled biphasic mode. The three commonly used at St Mary’s PICU are continuous negative pressure, respiratory synchronised mode and secretion mode.

**Negative pressure ventilation (NPV):** NPV works by exposing the surface of the chest wall to sub-atmospheric...
pressures resulting in expansion of the lungs and drawing in of air. Both the inspiratory and expiratory phases can be fully controlled by modifying the negativity of the air pressure. The pressure can be changed at varying rates allowing for a total or partial control over the patient’s spontaneous ventilation. Alternatively, the negative pressure can be maintained at a constant level throughout the spontaneous respiratory cycle resulting in continuous extra thoracic pressure (CNEP) support.

Respiratory Synchronized Mode: This mode allows the patient to breathe spontaneously. The patient’s inspiratory and expiratory efforts are triggers for the respirator and thus full synchronisation is achieved. The trigger sensitivity can be set and is patient dependant.

Secretion Clearance (‘physio mode’): This is a physiotherapy tool used to enhance clearance of secretions from the lungs using two alternating modes. This is achieved by high frequency oscillations (up to 1200 oscillations per minute) around a negative baseline followed by an artificial cough that has a prolonged inspiratory phase followed by a forced short expiratory phase.

Clinical applications of negative pressure ventilation

NPV may be used in children who are not suitable for non-invasive PPV: children who do not tolerate a facemask, have facial deformities, who are claustrophobic or have excessive airway secretions. Table 1 summarises the use of NPV in paediatric practice.

Table 1. Clinical conditions in which NIV has been used

<table>
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<tr>
<th>Condition</th>
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<tr>
<td>Respiratory distress syndrome of the Newborn</td>
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<tr>
<td>Bronchiolitis</td>
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<tr>
<td>Neuromuscular disorders</td>
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<tr>
<td>Central hypoventilation syndrome</td>
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<tr>
<td>Cystic fibrosis</td>
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<tr>
<td>Traumatic head injury</td>
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<tr>
<td>Lower respiratory tract infection</td>
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<tr>
<td>Chronic restrictive lung disease</td>
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<tr>
<td>Post-cardiac surgery (e.g. Fontan circulation, Tetrology of Fallot repair, thoracoplasty)</td>
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<tr>
<td>Pneumatocele</td>
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<tr>
<td>Failed extubation to avoid re-intubation</td>
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<tr>
<td>Difficult weaning due to chest wall weakness</td>
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A Cochrane review was published in 2003 that evaluated the effectiveness of CNEP or CPAP in paediatric patients with acute hypoxaemic respiratory failure (AHRF) due to non-cardiogenic causes. It concluded that description of clinical improvements with the use of non-invasive support required further studies. An update of this review was published in 2005 by Shah et al. Only 1 study was found eligible for inclusion in which patients with bronchiolitis on CNEP were compared to control patients. Those on CNEP had a reduced FiO₂ to less than 30% within 1 hour of initiation of therapy compared to control patients. The authors concluded that CNEP may be beneficial but more trials were needed. The authors now describe the role of NPV in various medical conditions through some cases managed in the present unit.

1. CNEP in bronchiolitis: It has been postulated that air trapping in infants with bronchiolitis resulting from small airway closure due to their easily collapsible chest wall may be reduced by the application of negative pressure. In our present Pediatric Intensive Care Unit (PICU), nCPAP and CNEP have been used to support infants with respiratory failure due to bronchiolitis. In unpublished data from their present unit, the authors tried to determine the efficacy and predictors of success/failure of non-invasive techniques of respiratory support for infants with respiratory failure due to bronchiolitis. 80 patients with bronchiolitis in respiratory failure were managed on invasive mechanical ventilation while 60 patients were either given CPAP (n=49) or CNEP (n=11). Of these 60 patients, 39 infants (65%) were successfully managed while twenty-one infants (35%) subsequently required mechanical ventilation. Underlying risk factors such as prematurity, chronic lung disease, congenital heart disease and neuromuscular disorders were more frequent in this unsuccessful group. The authors concluded that non-invasive respiratory support is an effective alternative to conventional ventilation for infants with bronchiolitis without underlying risk factors. Samuels et al report an intubation rate of 0.4% in their unit with the early use of negative pressure ventilation in patients with bronchiolitis. Al-Bakhi retrospectively compared the use of negative pressure ventilation to invasive positive pressure ventilation in infants with recurrent apnoea secondary to acute bronchiolitis. Negative pressure was associated with a reduced rate of intubation, shorter PICU stay and lesser use of sedation. In a commentary on the work done by Al-Bakhi, Henderson from Bristol feels that despite the lack of evidence from randomised controlled trials, non-invasive respiratory support offers a potential advantage of avoiding endotracheal intubation, at least for some infants.

2. CNEP in cardiac patients and effects on hemodynamic status: Negative pressure ventilation improves cardiac output by lowering mean thoracic pressure which maximises the venous return to the heart. In patients with cardiogenic shock or right heart failure, continuous negative pressure supports FRC while biphasic external cuirass ventilation decreases the afterload of the right ventricle thereby increasing the pulmonary blood flow. This optimises oxygenation and alveolar ventilation.

The authors successfully managed a case of severe heart failure on NPV and avoided the risks of intubation. A 14-yr-old boy presented with a history of upper respiratory tract infection followed by increasing fatigue.
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and breathlessness. He was febrile, tachycardic, tachypnoeic with crepitations at the left lung base and a gallop rhythm. Oxygen saturations were 84% in air and he was exhausted. Capillary refill time was 3 seconds, blood pressure was 90/40 mm Hg. He had a 3 cm palpable liver. Arterial blood gas revealed Ph 7.30, BE –8, CO2 3.5kPa. These findings were consistent with cardiac failure. ECG showed low amplitude complexes suggestive of myocarditis. A heart echo revealed bi-ventricular dilatation. He initially required 15 liters of oxygen by rebreathing mask to attain oxygen saturations of 90%. He was then started on CNEP using the Hayek oscillator via a chest cuirass with initial settings of -10 cm H2O in addition to facemask oxygen. His clinical status improved within a few hours with a reduction in tachycardia and tachypnoea and improved blood pressure. The oxygen requirements came down gradually. Endotracheal intubation was avoided. This could have been difficult considering the risks of induction agents in myocarditis. This boy did not require any inotropic support during his stay in PICU.

Myocarditis significantly increases the risk of cardiac arrest and mortality related to anaesthetic agents because of the dysrhythmic potential. Low cardiac output can complicate the post operative course of patients undergoing cardiac surgeries like Fontan repair and Tetrology of Fallot repair. Shekerdemian et al investigated the effect of NPV on cardiac output in the early postoperative period after right heart surgery and found that NPV improved cardiac output and stroke volume while systemic and pulmonary vascular resistances were reduced significantly. The same group also studied acute post-operative cardiopulmonary hemodynamics in 9 children who underwent Fontan procedure and found an increase in pulmonary blood flow during CNEP with values falling to base line with PPV. They concluded that increased stroke volume resulted in augmentation of pulmonary blood flow during CNEP ultimately resulting in increased cardiac output. Sideno et al showed that biphasic cuirass ventilation when applied with external high frequency oscillations can be used to improve cardiac function after coronary artery bypass grafting.

3. CNEP in Staphylococcal pneumatocele: Tension pneumothorax could lead to cardiopulmonary instability. In some instances, it could subsequently rupture forming a pneumothorax or a broncho-pleural fistula (BPF), especially during PPV. The proposed check-valve mechanism for the development of pneumatoceles predicts that PPV could lead to distension of these airspaces and formation of tension pneumothoraces.

The authors successfully managed on NPV a 10-month-old girl who presented with a 10-day history of cough and fever and respiratory distress. On examination, she had absent breath sounds on the right infra-mammary region. She had severe respiratory distress with intercostal and sub-costal recessions and her blood gas showed a rising PCO2 (10.2 kPa). Chest X-ray was consistent with a right-sided pneumatocele. Possibilities of using elective intubation or facial CPAP were discussed and a decision was made to initiate CNEP at –10 cm H2O. Within a few hours her clinical parameters improved. Oxygen requirements reduced and she did not require endotracheal intubation and PPV that could have predisposed this patient to the enlargement of the pneumatocele with subsequent rupture and prolonged air leak.

4. CNEP in neuromuscular disorders: CNEP has been used in obstructive sleep apnoeas, tracheobronchomalacia, central hypoventilation syndrome and neuromuscular diseases. Studies have shown an improved quality of life and physical activity and improvements in blood gases when these patients were managed on non-invasive techniques. Jackson et al investigated the long-term (5 yr) effectiveness of cuirass-assisted ventilation in patients with chest wall disease and neuromuscular disorders. There was no significant change in arterial oxygen tension (PaO2), lung volumes, respiratory muscle strength, hemoglobin, right heart failure, exercise tolerance, mental function and symptom scores after 5 yr compared to after 1 yr. They concluded that nocturnal cuirass-assisted ventilation has a role in long-term management of patients with neuromuscular and skeletal chest wall disorders. Fig. 1 shows a patient of neuro-muscular disease being ventilated on RTx ventilator.

![Fig. 1. Patient with Neuromuscular disorder being ventilated on RTx Ventilator](image-url)
subsequently discharged home with his own CNEP ventilator. He was readmitted only once in the subsequent 6 mth.

5. CNEP in neonatal respiratory failure: Negative pressure ventilation is described as effective in neonatal respiratory distress syndrome. 26,27,29 Negative pressure has an important role, particularly in the resolution phase of respiratory distress syndrome in preterm infants, and may reduce the risk of developing chronic lung disease. 30,10 Negative pressure ventilation provides a guaranteed trans-pulmonary pressure gradient in preterm infants with hyaline membrane disease. 31 Samuels et al have demonstrated the reduced need for endotracheal intubation along with decreased duration of oxygen requirement in patients treated with CNEP as compared to the ones treated with standard therapy. 30 British investigators report that when comparing CNEP with standard respiratory support for neonatal respiratory distress syndrome in preterm infants, outcomes 10 to 15 yr later appear to be similar between groups with some cognitive advantages noted for patients treated with CNEP. 32

6. CNEP in Central Hypoventilation Syndrome (CHS): In this disease entity, there is no respiratory drive during sleep and these children are ventilated at night usually through tracheostomy by positive pressure. Linton DM successfully converted 2 patients with CHS from PPV via a tracheostomy to NPV via biphasic cuirass ventilation. This allowed closure of the tracheostomy. 33 Hartman et al studied 9 patients with central hypoventilation syndrome (CHS) who were treated with CNEP. They concluded that CNEP is an effective means of respiratory support in CHS patients. 34 It improves the patient’s quality of life during daytime. The child can breathe on his own and eat or drink as advised. It will also help the child to communicate more efficiently with parents and care team.

7. CNEP in respiratory distress following extubation: Some children who have been extubated require some form of respiratory support. CPAP is often an initial choice though it may not be tolerated or may not be suitable because of excessive airway secretions, facial deformities, claustrophobia, etc. In the present unit, though CPAP still remains a first line of management in these selected groups of patients, NPV is used frequently, either alone or with concurrent CPAP as required.

8. CNEP in trauma: CNEP can be used alone or in combination with invasive PPV to treat respiratory failure in children with traumatic injuries. CNEP can also help improve cardiac output and subsequently oxygen delivery to tissues in patients with head trauma. Torelli et al compared the haemodynamic effects of application of CNEP (-10 cmH\(_2\)O) and PEEP (10 cm H\(_2\)O) in consecutive 15 mechanically ventilated patients with head trauma. They observed a significant increase in cardiac index (CI) and oxygen delivery index during CNEP compared with PEEP. They concluded that CNEP improves CI in mechanically ventilated patients with and without associated lung damage. 35

9. CNEP in weaning from PPV: CNEP can be used as an adjunct whilst the patient is weaned from PPV. As negative extra-thoracic pressures are applied to a patient on PPV, the requirements for pressure support are reduced and physiological improvements are observed in oxygenation, ventilation and cardiac output. 36 Takeda et al have shown that application of external biphasic (cyclical negative and positive pressure) cuirass ventilation in patients already receiving PPV facilitates secretion clearance and supports ventilation during temporary disconnections from PPV for endotracheal tube suctioning. 37 Patients who retain upper airway protective reflexes and who have good pulmonary compliance but are otherwise too weak to generate tidal volumes can be easily weaned from PPV to NPV. They can be extubated onto mandatory negative pressure breaths that can later be weaned to continuous negative pressure mode as tolerated.

10. NPV in chest physiotherapy and secretion clearance: Clearing of airway secretions can be enhanced by cuirass ventilation when used in high frequency oscillations followed by supra-normal inspiratory to expiratory excursions. This mode can be used at regular intervals in a patient with copious secretions difficult to eliminate for example in cystic fibrosis, chronic lung disease and acute chest syndrome of sickle cell disease. Mechanical high frequency secretion mode clearance and conventional chest physiotherapy were compared and found to be equally effective in 20 patients with bronchiectasis. 38

Monitoring the use of NPV

Pulse oximetry, end-tidal CO\(_2\), arterial or capillary blood gas monitoring are useful tools to evaluate patient’s clinical progress on NPV. If NPV is added to PPV, further reduction in positive pressure requirements, improvements in oxygenation and ventilation should suggest that its application is beneficial. Children on long term NPV may be monitored by using pulse oximetry alone. But, parents have to be taught how to detect clinical signs of acute respiratory distress.

Potential complications and limitations of NPV

The main disadvantages of NPV relate to the use of cuirass: physical access to the patient for nursing care may be restricted and, despite being transparent, accurate assessment of chest wall movement and signs of respiratory distress may be difficult. Though it is desirable to obtain a good seal around the patient, one has to be careful to protect the skin at contact points and minimise skin injury and avoid venous obstruction. The most serious problem with NPV is extra-thoracic upper airway obstruction when the abductors fail to contract to
oppose the negative pressure within the trachea generated during the inspiratory cycle of NPV. This problem can be overcome by the addition of nasal CPAP. There is a possibility of reflux and aspiration associated with dynamic effects on the lower oesophageal sphincter.  

CONCLUSION

In summary, NPV appears to be a promising modality to deliver NIV for a selected group of pediatric patients. The potential benefits of NPV include reduced airway complications, improved pulmonary parenchymal inflation at reduced airway pressures, improved hemodynamics thereby reduced cardiovascular compromise, decreased sedation requirements and improved enteral nutrition. Due to paucity of data, there is a need for further clinical evaluation of the use of NPV before this mode of ventilation becomes a recommended part of ICU management. Training of staff for this mode of respiratory support is possible and infants can be monitored without invasive techniques used in intensive care units. This approach could drastically reduce the cost incurred in treating patients on more invasive modes which in turn require invasive monitoring in an ICU set up which is an important aspect of medical management in developing countries.

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