Noninvasive Ventilation to Facilitate Extubation in a Pediatric Intensive Care Unit

John F. Pope, MD*
and David J. Birnkrant, MD†

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Noninvasive ventilation has been used extensively to treat chronic respiratory failure associated with neuromuscular and other restrictive thoracic diseases, and is also effective in the treatment of acute respiratory failure, allowing some patients to avoid intubation. Noninvasive positive pressure ventilation is a potentially effective way to transition selected patients off endotracheal mechanical ventilation. The authors present a retrospective chart review of pediatric patients extubated with the use of noninvasive ventilation. Extubation with noninvasive positive pressure ventilation was attempted in 25 patients. The patients had a variety of diagnoses, including neuromuscular diseases, cerebral palsy with chronic respiratory insufficiency, asthma, and acute respiratory distress syndrome (ARDS), reflecting the diversity of patients with respiratory failure seen in our pediatric intensive care unit (ICU). Indications for noninvasive ventilation-assisted extubation were chronic respiratory insufficiency, clinical evidence the patient was failing extubation, or failure of a previous attempt to extubate. Extubation was successfully facilitated in 20 of 25 patients. Of the five patients failing an initial attempt at noninvasive ventilation-assisted extubation, two required tracheostomy, two were subsequently extubated with the aid of noninvasive ventilation, and one was subsequently extubated without the use of noninvasive ventilation. Risk factors for failure to successfully extubate with the assistance of noninvasive positive pressure ventilation included the patient's inability to manage respiratory tract secretions, severe upper airway obstruction, impaired mental status, and ineffective cough with mucus plugging of the large airways. All patients had mild to moderate skin irritation due to the mask interface. No patient had any serious or long-term adverse effect of noninvasive positive pressure ventilation. All patients left the hospital alive. Noninvasive positive pressure ventilation can facilitate endotracheal extubation in pediatric patients with diverse diagnoses who have failed or who are at risk of failing extubation, including those with neuromuscular weakness.

From the *Department of Pediatrics, Children's Hospital Medical Center of Akron, Northeastern Ohio Universities College of Medicine, Akron, OH, and †Department of Pediatrics, MetroHealth Medical Center, Case Western Reserve University School of Medicine, Cleveland, OH.

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Address correspondence to Dr Pope, Department of Pediatrics, Division of Critical Care, Children's Hospital Medical Center of Akron, One Perkins Square, Akron, OH 44308–1062, or e-mail: jpope@chmca.org

Weaning from mechanical ventilation and endotracheal extubation is generally accomplished uneventfully in children. Extubation failure, defined as the need for reintubation, was reported to occur in 16% of patients in a recent study of 208 children. Sixty-four percent of patients who failed extubation exhibited signs and symptoms of excessive respiratory muscle load or inadequate respiratory effort [1]. Failure to extubate is most commonly the result of inadequate respiratory pump function resulting from neuromuscular weakness, increased respiratory muscle load, or a combination of both [2]. Pediatric patients with various chronic and acute illnesses may have particular difficulty being liberated from mechanical ventilation. For example, when children with neuromuscular disease or mental retardation/cerebral palsy complicated by chronic respiratory insufficiency require mechanical ventilation, weaning is problematic and tracheostomy may be required to accomplish endotracheal tube removal. Weaning may also be difficult in pediatric patients with acquired neuromuscular weakness due to prolonged mechanical ventilation, pharmacologic neuromuscular blockade, or high spinal cord injuries.

Noninvasive positive pressure ventilation is a practical and effective method of chronic respiratory support for patients with various causes of respiratory insufficiency, including neuromuscular and other restrictive thoracic diseases [3]. Noninvasive ventilation can be used to treat acute respiratory failure, avoiding the need for intubation [4–6]. In contrast to these well-described clinical applications of noninvasive ventilation, there are few published reports of the use of noninvasive ventilation to facilitate endotracheal extubation in patients who require mechanical ventilation [7–9]. However, noninvasive positive pressure ventilation is a potentially effective way to transition selected patients off endotracheal mechanical ventilation by providing respiratory muscle support and decreasing the work of breathing. In this article we review our experience using noninvasive ventilation to facilitate endotracheal extubation in our pediatric intensive care unit (PICU).

Materials and Methods

The data were collected by retrospective chart review on all patients extubated with noninvasive positive pressure ventilation assistance from December 1993 to July 1997. Selection of candidates for use of noninvasive positive pressure ventilation was at the discretion of the attending pediatric critical care specialist. No patient extubated with the assistance of positive pressure noninvasive ventilation during this time period was excluded. Prior to attempted extubation, all patients were weaned to minimal mechanical ventilatory support. All but two patients had noninvasive ventilation with the BiPAP® ST/D unit (Respironics, Murrysville, PA); in one patient a PB335® ventilator (Nellcor Puritan Bennett, Carlsbad, CA) was used, and the remaining patient was ventilated with a Quantum® PSV (Healthdyne Technologies, Marietta, GA). The BiPAP, PB335, and Quantum devices are pressure-limited, flow-sensitive ventilators with individually set inspiratory and expiratory pressure supports. The inspiratory pressure, expiratory pressure, and supplemental oxygen flow were adjusted to maintain adequate chest rise and tidal volume, decrease accessory muscle use and respiratory rate, and provide adequate oxygenation and ventilation as measured by arterial blood gases and/or oxyhemoglobin saturations. Multiple ventilator–patient interfaces were used. Except for the patients less than 3 years of age, all patients were initially extubated with full-face masks. Patients 3 years of age and younger were extubated with nasal mask interfaces. One of the infants with spinal muscular atrophy was ventilated with a custom adaptation of an INCA infant nasal cannula assembly (Ackrad Laboratories, Cranford, NJ) [10]. However, once it became commercially available, infants and toddlers were ventilated with a "small child" size pediatric mask (Respironics, Pittsburgh, PA). Nasal masks or pillows were used when patients were transitioned to chronic noninvasive ventilation.

Results

During the study time period 653 patients received mechanical ventilation in our PICU. Extubation with the use of noninvasive ventilation was attempted in 25 patients. The median age of the patients was 15 years (range 0.2–19 years); 5 of 25 patients were less than 3 years old. Patient ages, diagnoses, and indications for noninvasive ventilation-assisted extubation are shown in Table 1. Fourteen of the 25 patients had chronic respiratory insufficiency (10 patients with severe neuromuscular disease; 4 patients with mental retardation/cerebral palsy and chronic lung disease). The remaining 11 patients had various acute causes of respiratory failure. The median duration of endotracheal mechanical ventilation was 9 days (range 1–23 days).

Indications for use of noninvasive ventilation were chronic respiratory insufficiency (10 patients); clinical evidence the patient was failing within 2 hours of extubation (11 patients); or prior failure to extubate without noninvasive ventilation, necessitating one or more reintubations (4 patients) (Table 1). The "chronic respiratory insufficiency group" consisted of 10 patients with neuromuscular disease. Four of the patients were infants (ages 2, 4, 7, and 9 months) with spinal muscular atrophy type I. Three of the patients were intubated for placement of gastrostomy tubes. The fourth infant was intubated after a respiratory arrest at home. All four infants were supported with noninvasive ventilation at the time of extubation because of their profound weakness. Extubation using noninvasive ventilation was initially unsuccessful in one baby due to severe postextubation stridor. He was treated with dexamethasone and successfully extubated with noninvasive ventilation 48 hours later [11]. The remaining patients in the chronic respiratory insufficiency group had Duchenne muscular dystrophy (four patients) (ages 15–17 years) or spinal muscular atrophy type II (two patients) (ages 9 and 17 years). Baseline pulmonary function tests in these six patients showed severe respiratory muscle weakness, with five of six patients having forced vital capacities (FVCs) $\leq$ 16% of predicted (mean FVC = 15.7% predicted). Two of these patients failed noninvasive ventilation-assisted extubation due to an inability to manage airway secretions. One patient with spinal muscular atrophy type II and severe hypoxic-ischemic encephalopathy eventually required a tracheostomy. The second, a patient with Duchenne muscular dystrophy and pneumonia, was reintubated after he failed an initial attempt at extubation with noninvasive ventilation due to mucus plugging; he was extubated using noninvasive ventilation 3 days later.

The "clinically failing after extubation" group consisted of 11 patients who had severe respiratory distress within 2 hours of extubation. The managing physician felt each was going to require mechanical ventilation and noninvasive positive pressure ventilation was used to avoid reintubation. Two patients in this group failed noninvasive ventilation. One patient was recovering from acute respiratory distress syndrome (ARDS) secondary to traumatic rupture of his thoracic aorta. He did not tolerate noninvasive ventilation due to agitation and diffi-

Table 1. Indications, Characteristics, Diagnoses, and Outcomes of Patients Transitioned to Noninvasive Ventilation

Indication	Patients	Age range (years)	Diagnoses (number of patients)	Outcomes (number of patients)
Chronic respiratory insufficiency	10	0.2–17	Type I SMA s/p gastrostomy (3) s/p respiratory arrest (1) Type II SMA pneumonia (1) s/p respiratory arrest (1) DMD s/p respiratory arrest (2) s/p gastrostomy (1) pneumonia (1)	Reintubation (2)* Tracheostomy (1) Extubated (7)
Clinically failing after extubation	11	1.8–17	ARDS (4) Status asthmaticus (3) Cardiomyopathy (2) Pneumonia (1) Spinal cord injury (1)	Reintubation (1)† Tracheostomy (1) Extubated (9)
Failed previous extubation attempt	4	2–19	MRCP with pneumonia (4)	Extubated (4)
Total	25	0.2–19		Failed (5) Extubated (20)

SMA = spinal muscular atrophy; DMD = Duchenne muscular dystrophy; ARDS = acute respiratory distress syndrome; MRCP = mental retardation and cerebral palsy.

*Patients failed initial attempt at extubation with noninvasive ventilation and were reintubated. Subsequent extubations with noninvasive ventilation were successful.

†Patient maintained extubation with continuous noninvasive ventilation for 3 days before being reintubated for increasing distress. Subsequently extubated without noninvasive ventilation 5 days later.

culty handling his respiratory secretions and required tracheostomy. The second patient was a 22-month-old girl with progressive encephalopathy who was intubated for status epilepticus. She tolerated noninvasive ventilation for 3 days after extubation but required reintubation due to respiratory syncytial virus pneumonia. She was eventually extubated without noninvasive ventilation.

The "failed extubation" group consisted of four patients with mental retardation/cerebral palsy who had failed at least one attempt at extubation prior to using noninvasive ventilation. The patients in this group were successfully extubated with noninvasive ventilation assistance a mean of 6 days after reintubation (range 4–8 days). The primary reason for extubation failure was upper airway obstruction (three patients) or whole lung atelectasis (one patient) (Table 1). In addition, all four patients had chronic lung disease due to recurrent aspirations, three patients had chronic supplemental oxygen dependency, and two patients had chronic carbon dioxide retention.

Of the 20 patients successfully managed with noninvasive ventilation, 11 were weaned off noninvasive ventilation (7 of 11 were weaned within 72 hours); 9 were transitioned to chronic nocturnal noninvasive ventilation (all had neuromuscular disease). All patients left the hospital alive.

All patients had mild to moderate skin irritation due to application of the mask interface. No patient had skin ulceration or scarring. One patient had gastric distention due to insufflation, requiring venting of her gastrostomy tube. No patient had hemodynamic instability or suffered any long-term adverse effects of noninvasive ventilation. Noninvasive ventilation was never discontinued due to an adverse effect.

Discussion

Noninvasive positive pressure ventilation has been used extensively to treat chronic respiratory failure associated with neuromuscular and other restrictive thoracic diseases. For these patients, nocturnal noninvasive positive pressure ventilation can reverse symptoms of chronic alveolar hypoventilation, normalize oxyhemoglobin saturation, decrease frequency of hospitalization and prolong survival [2,12].

Noninvasive positive pressure ventilation is also effective in the treatment of acute respiratory failure, allowing some patients to avoid intubation [4]. Recent reports have focused on the use of noninvasive positive pressure ventilation in pediatric critical care units and have demonstrated that children with respiratory failure associated with various diagnoses can be successfully managed with noninvasive ventilatory support. These reports have shown noninvasive positive pressure ventilation can decrease the work of breathing, improve oxygenation and ventilation, and prevent intubation in children with respiratory failure [5,6]. While some studies of noninvasive ventilation for acute respiratory failure include adult [13] or pediatric [5,6,9] patients clinically failing extubation, few studies have specifically reported the use of noninvasive ventilation to facilitate extubation. In one study of acute respiratory failure, 18 of 28 pediatric patients had noninvasive ventilation to avoid reintubation, with a success rate of 89%. Patients with neuromuscular disease were excluded [5]. Meduri et al. [13] reported using noninvasive ventilation in 158 adults with acute respiratory failure. For 37 patients, acute respiratory failure occurred postextubation and 24 of these 37 (65%) were managed successfully with noninvasive positive pressure ventilation. Tobias and Wilson [9] reported the use of noninvasive ventilation to avoid reintubation in three children who developed respiratory failure following cardiac surgery. Udwadia et al. [7] reported the use of noninvasive positive pressure ventilation to extubate 18 of 22 adult patients with neuromuscular disease, chest wall restriction, primary lung disease, or cardiac disorders. Nine of 22 patients were postoperative. Restrick et al. [8] reported extubation using noninvasive positive pressure ventilation in 13 of 14 adult patients with chronic air flow limitation, chest wall restriction, or neuromuscular disease, two of whom were postoperative.

The 25 patients in our study represent 4% of the patients requiring mechanical ventilation in our PICU during the time period studied. The 80% extubation success rate in our patients using noninvasive positive pressure ventilation is consistent with the 65–89% success rates reported by others [5,13]. The primary reason for reintubation in three of the five patients who failed was excessive airway secretions. Two of these three patients had impaired mental status and one had an ineffective cough. The two patients with mental status changes required tracheostomies and the third patient was extubated to noninvasive ventilation after antibiotic treatment. These patients illustrate relative contraindications to noninvasive ventilation: inability to handle respiratory secretions/risk of aspiration, and mental status change/lack of patient cooperation. Patients with neuromuscular weakness and ineffective cough can be managed with noninvasive ventilation, provided they are able to cooperate and do not have significantly increased respiratory tract secretions [14]. The two remaining patients required reintubation for postextubation stridor and viral pneumonia. Both were subsequently extubated, one with and one without the aid of noninvasive ventilation. In total, 22 of 25 patients (88%) were successfully extubated with the aid of noninvasive positive pressure ventilation, 23 patients on the first attempt and 2 patients on the second attempt.

The retrospective nature of this study and the lack of control subjects makes it impossible to prove that our patients could not have extubated without noninvasive ventilation. However, these patients were very poor candidates for successful extubation. The 10 patients in the "chronic respiratory insufficiency" group all had neuromuscular disease with severe weakness. The four infants with spinal muscular atrophy type I had a rapidly degenerative neuromuscular disease characterized by progressive respiratory insufficiency leading to death before the age of 2 years [15]. Although these four infants could not undergo formal pulmonary function testing, they all exhibited clinical signs of respiratory insufficiency. The remaining six patients in this group had a significant risk of extubation failure, with pulmonary function tests reflecting severe respiratory muscle weakness [16]. The "failed extubation" group consisted of four patients with mental retardation and cerebral palsy who were successfully extubated with noninvasive ventilation after one or more previous attempts at extubation failed. Although the primary reason for failing extubation was upper airway obstruction (in three of four patients), the patients also had chronic lung disease, oxygen dependency, and/or carbon dioxide retention, making the cause of extubation failure multifactorial. Potential reasons for difficulty extubating these patients included increased work of breathing due to partial upper airway obstruction, decreased respiratory muscle strength due to muscle fatigue and/or malnutrition, and increased respiratory muscle pump load due to chronic lung disease (e.g., due to recurrent aspiration). The patients in the "clinically failing after extubation" group had severe respiratory distress and would have been immediately reintubated if noninvasive ventilation had not been available. The purpose of this report is not to prove the necessity of noninvasive ventilation in accomplishing extubation. Rather we have demonstrated the efficacy and practicality of noninvasive positive pressure ventilation in transitioning selected pediatric patients off mechanical ventilation through an endotracheal tube.

We can speculate on the mechanism by which noninvasive ventilation facilitates endotracheal extubation. Noninvasive positive pressure ventilation has been shown to provide similar levels of physiologic support when compared to endotracheal mechanical ventilation [17]. Noninvasive positive pressure ventilation has been shown to decrease the work of breathing in patients with neuromuscular and chronic obstructive pulmonary disease, as evidenced by decreased electromyographic activity in the diaphragm and accessory muscles of respiration, a decrease in the transdiaphragmatic pressure-time index, and a less negative esophageal pressure [18]. In addition, noninvasive positive pressure ventilation likely improves the asynchrony of rib cage and abdominal wall motion which characterizes the breathing pattern of patients with neuromuscular disorders [19,20]. Extubation failure is most often the result of inadequate respiratory pump function [2] and work of breathing has been shown to increase immediately after endotracheal extubation [21]. We hypothesize that noninvasive ventilation assisted our patients postextubation, when there was increased demand on their weak respiratory muscles. Noninvasive positive pressure ventilation also relieved partial upper airway obstruction in our patients with mental retardation/cerebral palsy, most likely by stenting open their collapsible upper airways with positive pressure.

Side effects of noninvasive ventilation were minimal in our patients, consisting primarily of skin irritation, usually over the bridge of the nose, due to pressure from the mask interface. Careful attention to mask fit, application of thin skin pads, alternating interfaces, providing time off ventilation if possible, and frequent examination of pressure points are all very important in preventing skin breakdown.

Conclusion

Noninvasive positive pressure ventilation is an effective method of assisting endotracheal extubation in selected pediatric patients, especially those with primary or secondary neuromuscular weakness or mental retardation/cerebral palsy with upper airway obstruction and chronic lung disease. Further studies will be necessary to establish the optimal uses of noninvasive positive pressure ventilation in pediatric respiratory medicine.

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