

Noninvasive Ventilation As an Alternative to Endotracheal Intubation During Tracheotomy in Advanced Neuromuscular Disease

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OBJECTIVE: To compare conventional tracheotomy with endotracheal intubation to tracheotomy with noninvasive positive-pressure ventilation (NPPV) in advanced neuromuscular disease. **METHODS:** This was a retrospective study of a historical cohort of patients tracheotomized while sedated and intubated versus patients tracheotomized under NPPV and local anesthesia. We recorded previous intubation difficulties, complications (eg, aspiration pneumonia), and hospital stay. **RESULTS:** Conventional tracheotomy was performed in 7 patients. We performed tracheotomy during NPPV with local anesthesia in 13 patients. All but 3 patients had risk factors for difficult intubation. Hospital stay was 23.3 ± 10.3 d in the conventional group and 25.3 ± 12.9 d in the NPPV group ($p = 0.87$). The number of pneumonias was higher in the conventional-tracheotomy group (4 vs 1, $p = 0.03$). **CONCLUSION:** In neuromuscular patients, performing tracheotomy with NPPV and local anesthesia may help avoid endotracheal intubation and reduce morbidity. *Key words:* tracheotomy, neuromuscular disease, noninvasive ventilation, difficult intubation. [Respir Care 2007; 52(12):1728–1733.   2007 Daedalus Enterprises]

Introduction

Tracheotomy is a major step for patients with neuromuscular disease, because it requires a higher level of care, which affects quality of life and increases dependence. Intolerance or nonfeasibility of noninvasive positive-pressure ventilation (NPPV) is the most common reason for tracheotomy of patients with Duchenne muscular dystrophy.¹ Other reasons include worsening of respiratory failure, with a low vital capacity (VC), persistent hypercarbia, and the need to increase ventilation time.²

One major condition to realize tracheotomy is to be able to maintain efficient ventilation throughout the procedure while providing an efficient analgesia. Because of their severely impaired pulmonary function, respiratory management of neuromuscular patients is very challenging. Neuromuscular patients are at risk of severe complications when they require deep sedation or anesthesia with endotracheal intubation. These patients present many factors that can lead to difficult intubation: skeletal deformities, tracheal deviation related to kyphoscoliosis, reduced neck mobility due to cervical fusion or myopathy, tongue hypertrophy, and reduced mouth opening.³

The main risk in case of difficult intubation is a delay in efficient airway protection; adverse events in this setting include desaturation, hypoxemia, aspiration pneumonia, and prolonged stay in the intensive care unit and/or hospital.⁴ Muscle relaxants and sedative anesthetic agents may induce severe respiratory depression and inefficient cough; moreover, muscle relaxants may cause rhabdomyolysis with fatal hyperkalemia.^{5,6} A heightened maximum effect of vecuronium has been reported in patients with Duchenne muscular dystrophy, which results in a large increase in time to recovery.⁷ Sedative agents can induce severe hypotension in patients with cardiac involvement, as seen in Duchenne muscular dystrophy; the potential depressive

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effect of sedatives on cardiac contractility must be considered when used in patients with cardiac dysfunction and at risk of life-threatening cardiac dysrhythmia.⁸

The difficulties and risks associated with endotracheal intubation in patients with advanced neuromuscular disease prompted us to compare our experience with 2 tracheotomy procedures: (1) conventional tracheotomy with sedation and intubation and (2) tracheotomy with local anesthesia and NPPV, as it was recently proposed for gastrostomy procedure in Duchenne muscular dystrophy or in amyotrophic lateral sclerosis.^{9–11}

Methods

We conducted a retrospective study to compare an initial period when patients were deeply sedated and intubated for tracheotomy to a period when patients underwent tracheotomy with local anesthesia and NPPV but without sedation. Patients with neuromuscular disease followed up at the 8-bed home ventilation unit of the Raymond Poincaré Teaching Hospital intensive care unit and who underwent elective tracheotomy were eligible. Patients were ventilated, using volume or pressure home ventilators, via nasal mask and/or mouthpiece during nighttime and daytime, for at least 8 hours a day. Tracheotomy was proposed to any patient who presented with one of the following: VC < 20% of predicted with persistent hypercarbia, recurrence of respiratory failure symptoms despite good adherence to home ventilation, or NPPV intolerance. In the conventional-tracheotomy group, patients intubated in the days before tracheotomy were excluded, as were patients with acute respiratory failure in the NPPV-tracheotomy group. The following data were obtained from the patient's medical record: neuromuscular diagnosis, VC, blood gas values before tracheotomy during spontaneous breathing (at least 2 h before, when possible) and during ventilation, and ventilation variables (mode, interface type, hours of ventilation per 24 h). We also recorded the Mallampati score before tracheotomy¹² and risk factors for intubation difficulties (tongue hypertrophy, difficult mouth opening, mouth aperture < 3 cm, inability to hyperextend the neck, and kyphoscoliosis with tracheal deviation demonstrated either clinically or on a chest radiograph).^{13,14} Physiological data, including pulse oximetry, continuous electrocardiogram, and blood pressure, were monitored throughout the intervention. We recorded the following complications: intubation difficulty or impossibility (in the conventional-tracheotomy group), ventilation impossibility or need for emergency intubation (NPPV-tracheotomy group), bleeding that required vascular ligation, decannulation, pneumonia,¹⁵ and death. We also recorded hospital stay. Consent was obtained from each patient or the patient's family. The study was approved by our institutional review board.

Tracheotomy Procedures

Coagulation tests, electrocardiography, and chest radiography were performed prior to tracheotomy. Each procedure was performed in the intensive care unit at bedside by 2 senior intensivists. Five of the co-authors (PA, BC, JG, DO, and HP) performed the procedures. A trained otolaryngologist (PA) was available if needed. Other attending personnel included a respiratory therapist and intensive care nurses. Surgical aseptic technique with sterile drapes was used. A 1.5–2-cm horizontal incision was made over the second tracheal ring. The soft tissues were dissected down to the trachea, a tracheal incision was made between the second and third ring, and a tube (Tracheoflex, Rusch, Duluth, Georgia) was inserted.

Conventional Tracheotomy. During the first period of the study, 1998–2001, we used a classical approach to tracheotomy. All the patients were screened for difficult intubation via clinical examination and measurement of Mallampati score. The suction unit, oxygen source, laryngoscopic blade, light source, endotracheal tube, and fiberoptic were prepared and verified prior to each intubation. Proper patient positioning was obtained with alignment of the oral, pharyngeal, and laryngeal axis to provide the best visualization of the vocal cords. All the patients were pre-oxygenated by breathing pure oxygen through a face mask, and were sedated with midazolam (5–10 mg) and fentanyl (100–300 µg), to perform direct laryngoscopy and insert the endotracheal tube. Difficult intubation was defined by 2 unsuccessful trials with laryngoscope and the requirement to use the fiberoptic endoscope. Impossible intubation was defined by emergency tracheotomy because of lack of effective tracheal access despite fiberoptic endoscopy.

NPPV Tracheotomy. During the second period of the study, 2001–2005, a procedure designed to avoid endotracheal intubation during tracheotomy was systematically proposed to patients who required tracheotomy. The patients were ventilated with their usual ventilation parameters and interface during the tracheotomy, and no endotracheal intubation was attempted. If necessary, oxygen could be added at the ventilator or mask to maintain oxygen saturation above 95%. Local anesthesia was achieved by injecting 2% lidocaine into the skin and soft tissues of the neck; small fentanyl boluses (30–50 µg/kg/min) were added in case of pain. Consciousness was maintained throughout the entire procedure, and patients were regularly asked about comfort, need for oropharyngeal suctioning, and pain.

Statistics

Qualitative values were compared with the Fisher's exact test, and quantitative values were compared with the

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Table 1. Study Cohort Characteristics

NPPV Patient No.	Neuromuscular disease	Age (y)	Sex	VC (mean mL)	VC (mean % of predicted)	P _{aCO₂} (mean mm Hg)*	Ventilation Time (mean h/d)
1	Duchenne muscular dystrophy	32	M	ND	ND	ND	24
2	Duchenne muscular dystrophy	28	M	700	15	42.8	20
3	Duchenne muscular dystrophy	17	M	360	9	37.3	8
4	Duchenne muscular dystrophy	20	M	300	10	60.0	20
5	Duchenne muscular dystrophy	35	M	ND	ND	ND	24
6	Acid maltase deficiency	22	F	590	16	ND	24
7	Duchenne muscular dystrophy	20	M	610	13	51.7	8
8	Duchenne muscular dystrophy	19	M	440	12	45	10
9	Duchenne muscular dystrophy	21	M	400	10	43.9	12
10	Duchenne muscular dystrophy	25	M	410	8	46.7	8
11	Duchenne muscular dystrophy	28	M	520	11	55.9	15
12	Becker muscular dystrophy	29	M	770	16	46.0	11
13	Duchenne muscular dystrophy	19	M	400	10	53.6	12
Mean ± SD		24.2 ± 5.7	NA	500 ± 143	11.8 ± 2.7	48.3 ± 6.9	15.1 ± 6.4

Intubation Patient No.	Neuromuscular disease	Age (y)	Sex	VC (mean mL)	VC (mean % of predicted)†	P _{aCO₂} (mean mm Hg)*†	Ventilation Time (mean h/d)†
1	Duchenne muscular dystrophy	26	M	375	8	46.4	12
2	Duchenne muscular dystrophy	20	M	560	13	51.1	12
3	Limb girdle muscular dystrophy 2C	30	M	650	14	53.0	11
4	Duchenne muscular dystrophy	28	M	ND	ND	ND	24
5	Congenital muscular dystrophy 1C (Fukutin-related protein deficit)	37	F	440	14	46.0	24
6	Duchenne muscular dystrophy	22	M	300	8	58.2	8
7	Limb girdle muscular dystrophy 2C	36	M	590	12	53.1	8
Mean ± SD		27.2 ± 6	NA	485 ± 136	11.5 ± 2.6	51.3 ± 4.2	14.1 ± 6.4

*During spontaneous breathing.
†The noninvasive positive-pressure ventilation (NPPV) and conventional-intubation groups had similar vital capacity (VC) (p = 0.90), P_{aCO₂} (p = 0.356), and ventilation h/d (p = 0.81).
ND = no data available
NA = not applicable

nonparametric Mann-Whitney test. p values < 0.05 were considered statistically significant.

Results

Of the 20 patients who met inclusion criteria during the study period (1998–2005), 16 had Duchenne muscular dystrophy or Becker muscular dystrophy, and 4 had other muscle diseases (limb-girdle muscular dystrophy, limb girdle muscular dystrophy 2C, n = 2), acid maltase deficiency (n = 1), or congenital muscular dystrophy with fukutin-related gene deficit (fukutin-related protein, congenital muscular dystrophy 1C, n = 1). There were 18 men and 2 women, and the subjects' mean ± SD age was 25.7 ± 6.1 y (Table 1). The average hours of ventilation per day was 14.7 ± 6.5 h. Seven patients (2 in the conventional group and 5 in the NPPV group) were ventilated

for > 20 h/d, and 5 were totally dependent on the ventilator (2 in the conventional group and 3 in the NPPV group). Mean ± SD VC was 495 ± 141 mL (11.7 ± 2.7 percent of predicted). P_{aCO₂} during at least 2 hours free of mechanical ventilation was obtained from only 16 patients; the mean value was 48.7 ± 5.3 mm Hg. No patient in either group had bulbar dysfunction with patent clinical dysphagia.

All patients except 3 (1 in the conventional group and 2 in the NPPV group, Table 2) had risk factors for difficult intubation, with a Mallampati score higher than II (p = 0.92). Difficult intubation occurred in 4 patients in the conventional group; intubation with fiberoptic endoscopy was successful in 3 of these patients but failed in one patient, who therefore underwent emergency tracheotomy while being ventilated with a manual insufflator. All these patients experienced oxygen desaturation. There were no major cardiac arrhythmias.

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Table 2. Differences Between the NPPV and Conventional Ventilation Subjects

NPPV Patient No.	Risk Factors for Difficult Intubation	Mallampati Score	Hospital Stay (d)*	Complications
1	Tongue hypertrophy, reduced mouth opening, neck immobility	IV	10	None
2	Neck immobility	III	20	None
3	Neck immobility	III	30	None
4	Neck immobility	III	27	None
5	Tongue hypertrophy, reduced mouth opening, neck immobility	IV	21	None
6	None	II	63	Early decannulation
7	Reduced mouth opening, neck immobility	IV	36	None
8	Neck immobility	III	11	None
9	Tongue hypertrophy, reduced mouth opening, neck immobility	IV	30	None
10	Tongue hypertrophy, reduced mouth opening, reduced neck mobility	IV	20	Seizure, pneumonia
11	Tongue hypertrophy, reduced mouth opening, reduced neck mobility	IV	20	None
12	Reduced neck mobility	II	21	None
13	Tongue hypertrophy, reduced mouth opening, neck immobility	IV	20	None

Intubation Subject No.	Risk Factors for Difficult Intubation	Mallampati Score	Hospital Stay (d)	Complications
1	Kyphoscoliosis with tracheal deviation, reduced mouth opening, neck immobility	IV	41	Failed intubation, pneumonia
2	Tongue hypertrophy, reduced mouth opening, neck immobility	IV	11	Failed intubation
3	Tongue hypertrophy, kyphoscoliosis with tracheal deviation	III	15	Pneumonia
4	Tongue hypertrophy, difficulty opening the mouth, neck immobility	IV	21	Difficult intubation, pneumonia
5	Tongue hypertrophy, reduced mouth opening, neck immobility	IV	35	None
6	Tongue hypertrophy, reduced mouth opening, neck immobility	IV	15	Difficult intubation
7	Reduced neck mobility	II	25	Pneumonia

*The noninvasive positive-pressure ventilation (NPPV) and conventional-intubation groups had similar duration of hospitalization ($p = 0.87$) and number of patients with a Mallampatti score > 2 ($p = 0.92$). There were significantly fewer pneumonias in the NPPV group than in the conventional-ventilation group ($p = 0.03$).
NPPV = noninvasive positive-pressure ventilation

There were 5 pneumonias, all of which occurred within the first 5 days after tracheotomy, which suggests aspiration as the cause of the pneumonia. Pneumonia was more common in the conventional-tracheotomy group ($p = 0.03$) (see Table 2); the only pneumonia in the NPPV group was diagnosed after a seizure due to lidocaine injection (error of dosage). Inadvertent decannulation occurred in one patient in the NPPV group, and was corrected immediately at bedside, without any consequence. There were no desaturations or arrhythmias in the NPPV group. There were no cases of bleeding or death. Mean \pm SD total hospital stay was 24.6 ± 12.4 d for the whole study group, 25.3 ± 12.9 d for the NPPV

group, and 23.3 ± 10.3 d for the conventional group ($p = 0.87$).

Discussion

In these neuromuscular patients with severe restrictive respiratory failure, and who required tracheotomy (either because of disease progression or acute respiratory failure), endotracheal intubation and deep sedation for tracheotomy were associated with a significantly higher incidence of pneumonia than was tracheotomy under local anesthesia and NPPV. There was no significant difference in hospital stay between the NPPV and conventional groups.

Our purpose was to evaluate if tracheotomy under local anesthesia and NPPV reduced complications associated with intubation during tracheotomy in patients with severe pulmonary restrictive failure. Our method can be compared to gastrostomy insertion procedures in patients with amyotrophic lateral sclerosis or Duchenne muscular dystrophy.^{9,10} Our objective is to reduce hospital morbidity due to pulmonary complications related to intubation difficulties.¹⁶

Progress in medical management of neuromuscular disorders has resulted in longer survival. People with advanced neuromuscular disease, such as muscular dystrophy, now more frequently require invasive and painful procedures, and, therefore, deep sedation or anesthesia. In this series, patients who underwent tracheotomy had advanced disease with severe cardiac and respiratory involvement. They were particularly at risk of heightened sensitivity to sedatives and cardiorespiratory complications.

Some neuromuscular diseases are associated with major skeletal deformities that limit neck extension and/or cause macroglossia and reduced mouth opening.¹⁷ We know of no recommendations available for intubation of patients with neuromuscular disease. The American Society of Anesthesiologists Task Force on Management of the Difficult Airway recently published an update of practice guidelines.^{5,18,19} Factors that predict difficult intubation include an inter-incisor distance < 3 cm, a Mallampati score > II, decreased mandibular space compliance, and a short thick neck with impossible extension. Most of our patients had such risk factors.

In an earlier study, the relative risk of difficult intubation increased with Mallampati score.³ Seventeen of our 20 patients were Mallampati class III or IV, and 1 patient was impossible to intubate.

NPPV (via nasal mask, face mask, mouthpiece, or laryngeal mask) has been proposed in children with facial deformities, cervical spine rigidity, and neuromuscular disease undergoing invasive medical or surgical procedures to provide safe respiratory support during deep sedation or anesthesia.^{9,11,13,14,20} These techniques require well-trained physicians who are able to adapt the ventilator settings in response to the loss of spontaneous breathing due to sedation, or to change the method of ventilation during the procedure. No change in ventilator settings was necessary in our patients. The risk that NPPV might expose the patients to a lack of efficient airway protection during the procedure must be counterbalanced against the risks linked to endotracheal intubation.

Regional anesthesia is used to eliminate hazards associated with general anesthesia and to facilitate chest physiotherapy in patients with Duchenne muscular dystrophy.⁵ The advantages of avoiding general anesthesia include maintenance of the ability to cough and to swallow, as well as to breathe spontaneously, even in the event of

decannulation in patients with low VC and low respiratory reserve. Neuromuscular patients often present piecemeal deglutition and impairment of breathing-swallowing interaction.²¹ Swallowing disorders and inability to clear the airway^{22,23} may be worsened by the use of sedative drugs, anesthetics, and muscle relaxants, which increase the risk of aspiration pneumonia.⁵ Considering that intubation is often difficult and hazardous in patients with advanced neuromuscular disease, this additional factor may explain the high rate of aspiration pneumonia in the intubation group. On the other hand, the NPPV procedure allowed patients to remain conscious and therefore preserved voluntary breathing, swallowing, and coughing, and facilitated suctioning of oropharyngeal secretions, if needed, which reduced the risk of aspiration.

One of our patients experienced a seizure, which might have been an adverse effect of lidocaine injection, resulting from unexpected penetration of the compound into the bloodstream. This complication was anecdotal and probably more related to the high concentration of the anesthetic used than to the procedure itself. This complication was not observed again after we switched to a less concentrated product.

Tracheotomy is a surgical procedure that should be performed only by experienced physicians. Recently introduced techniques include percutaneous tracheotomy via puncture and progressive dilation with a bougie²⁴ or a grip,²⁵ and tracheotomy via endoscopic access.²⁶ These methods do not require in-depth knowledge of neck anatomy, compared to surgical tracheotomy. During tracheotomy, ventilation can be provided via laryngeal mask or Combitube.^{20,27,28} These techniques are not recommended for patients with cervical deformity²⁹ and therefore do not seem suitable for patients with neuromuscular diseases; further evaluation of these techniques is necessary in this population. Cannula malposition has been reported in up to 25% of cases, and bleeding due to blood vessel injury can occur.^{30,31}

The present study has major limitations because of the small number of subjects, the retrospective design, and the long study period (over 8 years). Practices may have been influenced by evolution of procedures; however, there were no major changes in therapies or supportive approaches (ventilation techniques, interfaces, and cough assistance) in the neuromuscular population during the study period. Intubation and tracheotomy were performed by the same team of trained senior physicians, and the sedation protocol with midazolam and fentanyl was not modified during the study period.

We did not find a difference in hospital stay between the groups. However, regardless of complications during or after the procedure, the post-tracheotomy period is dedicated to the education of the patient and family in regard to the tracheostomy tube and suctioning techniques. Dis-

charge from the hospital depends directly on the acquisition of these techniques and on the organization of the patient's environment. Therefore, hospital stay is not solely influenced by the complications from the procedures, which might account for the lack of difference in stay.

Conclusions

Our results suggest that NPPV tracheotomy with local or locoregional anesthesia is an acceptable option for patients with neuromuscular disease who are at high risk of complications from endotracheal anesthesia. This new procedure could be favored over tracheotomy with intubation and general anesthesia in patients with advanced neuromuscular disease.

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