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Original Research

CRITICAL CARE MEDICINE

Extubation of Patients With Neuromuscular Weakness

A New Management Paradigm

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Background: **Successful extubation conventionally necessitates the passing of spontaneous breathing trials (SBTs) and ventilator weaning parameters. We report successful extubation of patients with neuromuscular disease (NMD) and weakness who could not pass them.**

Methods: **NMD-specific extubation criteria and a new extubation protocol were developed. Data were collected on 157 consecutive "unweanable" patients, including 83 transferred from other hospitals who refused tracheostomies. They could not pass the SBTs before or after extubation. Once the pulse oxyhemoglobin saturation (Spo_2) was maintained at ≥ 95% in ambient air, patients were extubated to full noninvasive mechanical ventilation (NIV) support and aggressive mechanically assisted coughing (MAC). Rather than oxygen, NIV and MAC were used to maintain or return the Spo_2 to ≥ 95%. Extubation success was defined as not requiring reintubation during the hospitalization and was considered as a function of diagnosis, preintubation NIV experience, and vital capacity and assisted cough peak flows (CPF) at extubation.**

Results: **Before hospitalization 96 (61%) patients had no experience with NIV, 41 (26%) used it < 24 h per day, and 20 (13%) were continuously NIV dependent. The first-attempt protocol extubation success rate was 95% (149 patients). All 98 extubation attempts on patients with assisted CPF ≥ 160 L/m were successful. The dependence on continuous NIV and the duration of dependence prior to intubation correlated with extubation success ($P < .005$). Six of eight patients who initially failed extubation succeeded on subsequent attempts, so only two with no measurable assisted CPF underwent tracheotomy.**

Conclusions: **Continuous volume-cycled NIV via oral interfaces and masks and MAC with oximetry feedback in ambient air can permit safe extubation of unweanable patients with NMD.**

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Abbreviations: ALS = amyotrophic lateral sclerosis; CCM = critical care myopathy; CPF = cough peak flows; IPPV = intermittent positive pressure ventilation; MAC = mechanically assisted coughing; NIV = noninvasive mechanical ventilation; NMD = neuromuscular disease; PAP = positive airway pressure; SBT = spontaneous breathing trial; SMA 1 = spinal muscular atrophy type 1; Spo_2 = pulse oxyhemoglobin saturation; VC = vital capacity

Conventional extubation attempts follow successful "spontaneous breathing trials" (SBTs) and the passing of ventilator weaning parameters,[1] otherwise patients undergo tracheotomy. Patients are often extubated to supplemental oxygen and bilevel positive airway pressure (PAP), but settings are infrequently reported,[2] and extubation studies report very few if any patients with neuromuscular disease (NMD) (eg, 18 of 162,[3] 17 of 900[4]) and completely exclude unweanable patients.[5-7]

Patients with preexisting NMD make up only 4% to 12.5% of cases in critical care,[8-10] but about 25% in weaning centers.[11] While acquired critical care myopathy (CCM) is common, it is an often unrecognized[12,13] cause of extubation failure by conventional approaches.[14-16]

There are no guidelines for extubating unweanable patients with NMD and CCM. Many are dependent on noninvasive mechanical ventilation (NIV) with no autonomous breathing ability for years before being intubated, and they refuse tracheotomy.[17-20] Further, these patients can have ineffective cough peak flows (CPFs), which can result in extubation failure due to airway secretion accumulation,[21-24] but very few studies

have reported CPF[2,21] and none systematically used mechanically assisted coughing (mechanical insufflation-exsufflation with exsufflation-timed abdominal thrust) (MAC).[4,24] There are no "ventilator weaning parameters" that address the ability to expel secretions. With success in decannulating unweanable patients with traumatic tetraplegia and others to continuous MAC and NIV, which includes noninvasive intermittent positive pressure ventilation (IPPV) and high-span bilevel PAP,[20,25-27] we used similar criteria to extubate unweanable patients with NMD and CCM and report the success rates.

Materials and Methods

The data were gathered in two centers, with 113 patients in New Jersey and 44 in Portugal, using the inclusion criteria described in Table 1. The study was approved by the institutions' review boards. All intubated patients were treated conventionally except for the use of MAC via the tube. Although virtually unknown in critical care, MAC has been instrumental in avoiding pneumonia, respiratory failure, and hospitalizations for NIV-dependent patients with NMD.[28-30] Vital capacities (VCs) (Wright Mark 3 spirometer; Ferraris Ltd; London, England) and unassisted and assisted CPFs (Access Peak Flowmeter; Health Scan Products Inc.; Cedar Grove, NJ) were measured within 12 months before intubation for the local patients (group 1). The other 83 intubated patients were transferred intubated from other hospitals after one to four failed extubation attempts (group 2) or after failing multiple SBTs (group 3).

VC was measured via the tube with the cuff inflated following clearing of the airways by MAC just prior to extubation. Patients were ready for extubation and inclusion in this study only when all Table 1 criteria were satisfied and SBTs failed, as described.[31-33] Patients had to experience immediate distress, precipitous oxyhemoglobin desaturation, and hypercapnia without stabilization before return to full ventilatory support both before and immediately postextubation. Local patients were considered to be group 1 because their greater experience with NIV and MAC could have affected outcomes. All transferred patients had been told that extubation and survival were not possible without tracheotomy.

We reported that the risk for extubation failure is high when assisted CPF cannot attain 160 L/m.[22] Considering that patients with advanced averbal bulbar amyotrophic lateral sclerosis (ALS) can rarely attain a CPF of 160 L/m[34,35] we generally did not accept such patients for transfer (exclusion criteria). Local patients with a CPF < 160 L/m were offered extubation if acknowledging that three extubation failures would necessitate tracheotomy. Other, at least temporary, exclusion criteria were medical instability, inadequate cooperation, and imminent surgery.[20,35]

Table 1—*Extubation Criteria for Unweanable Ventilator-Dependent Patients*

Afebrile and normal WBC count
Aged 4 y and older
No ventilator-free breathing tolerance with 7-cm pressure support in ambient air on the basis of NMD or CCM
VC < 20% of normal
$Paco_2 \leq 40$ mm Hg at peak inspiratory pressures < 35 cm H_2O on full-setting assist/control mode at a rate of 10-13/min
$Spo_2 \geq 95\%$ for 12 h or more in ambient air
All oxyhemoglobin desaturations < 95% reversed by MAC and suctioning via translaryngeal tube
Fully alert and cooperative, receiving no sedative medications
Chest radiograph abnormalities cleared or clearing
Air leakage via upper airway sufficient for vocalization upon cuff deflation

CCM = critical care myopathy; MAC = mechanically assisted coughing; NMD = neuromuscular disease; Spo_2 = pulse oxyhemoglobin saturation; VC = vital capacity.

Protocol

While intubated, sufficient ventilatory support was used to maintain normocapnia and normal respiratory rates. MAC (CoughAssist; Respironics, Inc.; Murrysville, PA) was used at 40 to −40 cm H_2O or greater to rapidly achieve clinically full chest expansion to clinically complete emptying of the lungs, with exsufflation-timed abdominal thrusts. The MAC sessions were up to every 20 min to maintain or return the pulse oxyhemoglobin saturation (Spo_2) to ≥ 95% in ambient air. Tracheotomy would have been recommended if the Table 1 criteria could not be met within 2 weeks of transfer.

Once the Table 1 criteria were met, the orogastric or nasogastric tube was removed to facilitate postextubation nasal NIV. The patient was then extubated directly to NIV on assist/control of 800 to 1,500 mL, at a rate of 10 to 14 min in ambient air. Pressure control of at least 18 cm H_2O was used if abdominal distension developed. The NIV was provided via a combination of nasal, oronasal, and mouthpiece interfaces.[36] Assisted CPF and CPF obtained by abdominal thrust following "air stacking" were measured within 3 h as the patient received full volume-cycled NIV support. Patients kept 15-mm angled mouthpieces accessible (Fig 1), and weaned themselves, when possible, by taking fewer and fewer IPPVs as tolerated. Diurnal nasal IPPV was used for those who could not secure the mouthpiece. Patients took as much of the delivered volumes as desired. They used nasal or oronasal interfaces (Figs 2, 3) for nighttime ventilation. For episodes of Spo_2 < 95%, ventilator positive inspiratory pressure, interface or tubing air leakage, CO_2 retention, ventilator settings, and MAC were considered.

Patients were taught to maximally expand their lungs by air stacking (retaining consecutive) ventilator delivered volumes to the largest volume the glottis could hold.[37] Once the lunges were air stacked, an abdominal thrust was provided to manually assist the cough,[29,37] and these assisted CPFs were measured. For patients using pressure-cycling, air stacking volumes were provided by manual resuscitator. The therapists, nurses, and in particular, the family and personal care attendants provided MAC via oronasal interfaces up to every 20 min until the Spo_2 no longer dipped below 95% and the patients felt clear of secretions. In seven cases, the postextubation oral intake was considered unsafe, so open

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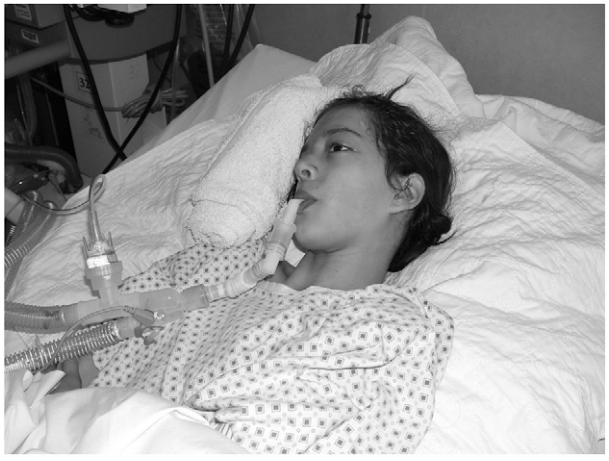

FIGURE 1. A 10-year-old girl with neurofibromatosis status post-spinal cord tumor resection, extubated with a vital capacity (VC) of 180 mL and no ventilator-free breathing ability, using a 15-mm angled mouthpiece (Malincrodt-Puritan-Bennett; Pleasanton, CA) for ventilatory support. Current VC 380 mL and minimal ventilator-free breathing ability. The patient provided written consent for the use of this photograph.

modified Stamm gastrostomies were performed under local anesthesia using NIV, as in Figure 2, without complication.[38]

Extubation was considered successful if the patient was discharged without reintubation. When reintubation was necessary, the patient was again extubated after achieving the Table 1 criteria. Multiple failures necessitated tracheotomy. Extubation success was considered as a function of diagnosis, patient group, VC, CPF, and preintubation NIV experience. VC was measured 3 to 6 months after extubation. The total days intubated were compared pretransfer and posttransfer.

Statistical Analysis

Results are expressed as mean ± SD. Statistical analyses were carried out using SPSS 12.0 (SPSS, Inc.; Chicago, IL). Differences in means were compared using the Wilcoxon rank test. A P value $\leq .05$ was considered significant. Univariate comparisons of potential predictive factors for failure or success were run with the Fisher exact test for categorical variables and the Wilcoxon rank-sums test for continuous variables.

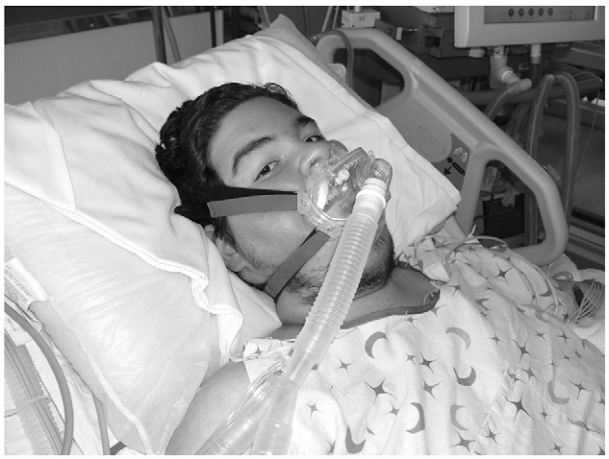

FIGURE 2. A 20-year-old man with Duchenne muscular dystrophy transferred for extubation after failing three extubations over a 26-day period. He used a 15-mm angled mouthpiece, as in Figure 1, for daytime ventilatory support and a lip-seal phalange with nasal prongs for nocturnal ventilatory support. His VC was 260 mL at extubation in October 2007 and 720 mL in July 2009. See Figure 1 legend for expansion of the abbreviation. The patient provided written consent for the use of this photograph.

RESULTS

The 157 patients, mean age 37 ± 21 years, included 139 (89%) with NMD who were intubated for acute respiratory failure and compromise due to pneumonia and/or surgery and 18 patients with CCM (11%). The 74 local patients (group 1) were intubated at our institutions, and 83 others were intubated elsewhere. Demographic data are in Table 2. VC and CPF data are in Table 3. Twenty (13%) of the 157 patients had been continuously NIV dependent for 12.2 years (range = 1-47) before being intubated. Forty-one (26%) used NIV part-time (< 24 h/d), and 96 (61%) used no NIV before intubation. All patients satisfied the Table 1 criteria in < 2 weeks.

Univariate comparisons of potential predictive factors for extubation success yielded significant differences for continuous NIV use ($P < .0001$) and for the duration of continuous NIV use and MAC use prior to intubation ($P = .0038$), indicating that experience with NIV and MAC was significant in predicting success. Given only 15 failures in eight patients, it was not possible to run multivariable logistic regression models considering diagnosis, patient group, VC, and CPF.

Of 172 extubations on 157 patients, all 98 on patients with assisted CPF ≥ 160 L/m were successful. Fifty-nine of 74 attempts (80%) on patients with CPF < 160 L/m were successful, including 52 of 60 (87%) at first extubation. Six patients who initially failed, succeeded on second (four patients) and third (three patients) attempts. One with advanced bulbar ALS and one with facioscapulohumeral muscular dystrophy, both with no measurable assisted CPF, failed a total of five extubations and underwent tracheotomy. Only one of the eight who failed any extubation attempt had preintubation experience with NIV and MAC, but she and several others had suboptimal postextubation care provider support for aggressive MAC. She and eight patients with bulbar ALS with little residual bulbar-innervated muscle function required oro-nasal interfaces for a closed system of postextubation NIV (Hybrid NE; Teleflex Medical; Research Triangle Park, NC) (Fig 2). All nine had gastrostomy tubes for total enteral nutrition. One lip-seal nocturnal NIV user for 28 years prior to intubation was extubated back to lip-seal NIV (Fig 3).[36]

Data on VC, CPF, and duration of NIV use as a function of postextubation weaning capacity are included in Table 4. Weaning from full-time to part-time

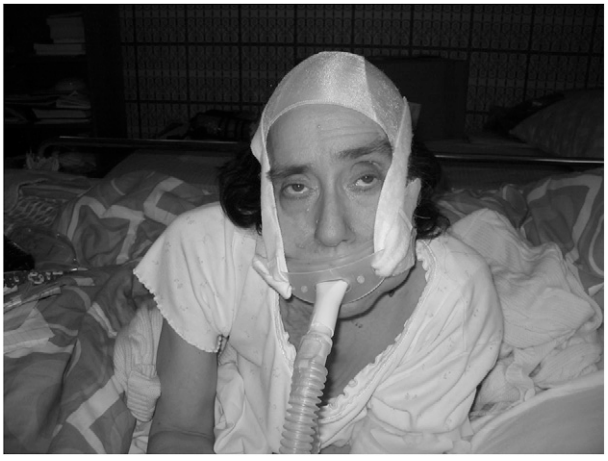

FIGURE 3. A 59-year-old woman with spinal cord injury at birth and 31 years of dependence on a daytime mouthpiece and nocturnal lip-seal (seen here) ventilation. She was extubated back to noninvasive mechanical ventilation despite a VC of 130 mL and no autonomous breathing ability and continued to use simple 15-mm angled-mouthpiece ventilation for daytime ventilatory support and lip-seal ventilation overnight with the nose clipped to prevent air leakage. Current VC is 340 mL. She is employed full-time as a psychologist. See Figure 1 legend for expansion of the abbreviation. The patient provided written consent for the use of this photograph.

NIV took 3 to 21 days and was usually accomplished at home. As supine VC increased to approach 1,000 mL, we encouraged patients to sleep without NIV but with Spo_2 and end-tidal CO_2 monitoring, and when these remained stable for 2 weeks to discontinue NIV. The mean extubation VCs and assisted CPFs for the 29 patients ≥ 18 years of age who were successfully extubated at first attempt despite assisted CPF < 160 L/m were 245 ± 114 mL (range 120-620) and 97 ± 39 L/m (range 0-150), respectively. No clinically or radiographically apparent barotrauma was noted for any patients.

The 83 patients in groups 2 and 3 had been intubated for 11 ± 9.1 days (range = 1-80) before transfer and 2 ± 1 days (range = 1-11) on our units before extubation ($P < .005$). Upon admission, on 21% fraction of inspired O_2, 71 (85%) of the patients' Spo_2 levels settled below 95%. Increased NIV support to normalize CO_2 and especially MAC normalized Spo_2 generally within 24 to 48 h to satisfy a criterion for extubation.

The intensivists and respiratory therapists estimated that noninvasive treatment necessitated more time than invasive respiratory treatment. The extubation itself required about 1.5 h for a specifically trained respiratory therapist to train the patients and care providers in NIV and MAC. In part because only one local nursing/rehabilitation facility would accept NIV users, all except one patient who had a tracheostomy were discharged home. One hundred thirty-one patients are alive using NIV (Table 4). Nine patients (6%) died of cardiac failure, six (4%) from lung disease/respiratory failure, two (2%) with bulbar ALS died after tracheotomy from sepsis and decubiti, and nine (6%) died of unknown causes. Although offered, no patients accepted tracheotomy following successful extubation.

DISCUSSION

There are no extubation studies on continuously NIV-dependent patients with NMD.[6] A recent controlled

Table 2—*Demographic Data*

Characteristics	Group 1[a]	Group 2[b]	Group 3[c]	Total
Subjects, No. (%)	74 (47)	45 (29)	38 (24)	157 (100)
Sex, No. (%)	52 male (70)	28 male (62)	17 male (45)	97 male (62)
	22 female (30)	17 female (38)	21 female (55)	60 female (38)
Diagnoses, No. (%)	ICUMy, 15 (20)	SMA, 10 (22)	SMA, 10 (26)	SMA, 25 (16)
	SCI, 13 (18)	MD, 9 (20)	DMD, 9 (24)	MD, 22 (14)
	ALS, 11 (15)	DMD, 8 (18)	MD, 5 (13)	DMD, 20 (13)
	MG, 9 (12)	MG, 6 (13)	PPS, 5 (13)	SCI, 17 (11)
	MD, 8 (11)	PPS, 4 (9)	oNMD, 4 (11)	ALS, 16(10)
	oNMD, 8 (11)	oNMD, 4 (9)	SCI, 3 (8)	oNMD, 16 (10)
	SMA, 5 (7)	ALS, 3 (7)	ALS, 2 (5)	ICUMy, 15 (10)
	DMD, 3 (4)	SCI, 1 (2)	. . .	MG, 15 (10)
	PPS, 2 (3)	. . .	. . .	PPS. 11 (7)
Use of NIV preintubation, No. (%)	No NIV, 51 (69)	No NIV, 24 (53)	No NIV, 21 (55)	No NIV, 96 (61)
	Cont, 10 (14)	Cont, 7 (16)	Cont, 3 (8)	Cont, 20 (13)
	Noct, 13 (17)	Noct, 14 (31)	Noct, 14 (37)	Noct, 41 (26)

ALS = amyotrophic lateral sclerosis; Cont = continuous noninvasive ventilation; DMD = Duchenne muscular dystrophy; ICUMy = ICU-acquired neuromuscular disease; MD = muscular dystrophy; MG = myasthenia gravis; NIV = noninvasive ventilation; Noct = nocturnal noninvasive ventilation; oNMD = other neuromuscular disease; PPS = postpolio syndrome; SCI = spinal cord injury; SMA = spinal muscular atrophy, including types 1, 2, and 3, and other neuromuscular disease.

[a]Local patients.

[b]Patients transferred after failing extubations in other institutions.

[c]Patients transferred after failing multiple spontaneous breathing trials in other institutions.

Table 3—*Pulmonary Function*

Characteristics	Group 1[a]	Group 2[b]	Group 3[c]	Total
Subjects, n (%)	74 (47%)	45 (29)	37 (24%)	157
Assisted CPF at extubation, L/min	187 ± 85	162 ± 86	178 ± 62	177 ± 77
VC at extubation, mL	355 ± 171	273 ± 189	295 ± 155	315 ± 173
VC 3-6 mo postextubation, mL	1,121 ± 748	709 ± 679	617 ± 412	877 ± 698

Intergroup differences were not statistically significantly different except for postextubation VC, with that of group 1 being greater than for groups 2 and 3 ($P < .05$). CPF = cough peak flows. See Table 1 for expansion of the other abbreviation.

[a]Local patients.

[b]Patients transferred after failing extubations in other institutions.

[c]Patients transferred after failing multiple spontaneous breathing trials in other institutions.

postextubation respiratory failure study of 106 patients included only two with restrictive syndromes, but none with NMD, and all had passed SBTs. They were extubated to supplemental O_2 alone or in conjunction with bilevel PAP at spans up to 14 cm H_2O, pressures inadequate for normal alveolar ventilation for our patients.[39] A metaanalysis of 12 extubation studies to bilevel PAP demonstrated decreased mortality, ventilator-associated pneumonia, length of stay, and resort to tracheotomy, but unweanable patients with NMD were uniformly excluded.[6] Eligibility for extubation was based on "readiness for weaning" and failure of SBTs after 30 min or more.[6] While this justifies extubation to NIV for patients who primarily have lung/airways disease with some autonomous breathing ability and for whom $Spo_2 > 90\%$ may be acceptable with or without supplemental O_2, our patients had no ability to sustain breathing before or after extubation with VCs as low as 0 mL. Thus, no control group extubation to O_2 or less than full NIV would be possible, ethical, or permissible by any review board. While there are always limitations of uncontrolled studies when comparing two approaches, this was a study of only one approach to extubate patients not previously considered extubatable. For our long-term NIV users, aspiration causing persistent $Spo_2 < 95\%$ despite continuous NIV and MAC in ambient air is the indication for tracheotomy.[35]

Besides hypoventilation, ineffective CPF have been associated with extubation failure.[21,22] MAC is essentially noninvasive suctioning via noninvasive or invasive interfaces. It can clear the left airways that are often not cleared by invasive suctioning and can acutely increase VC and Spo_2.[30,40,41] Our success stemmed not only from providing continuous full-setting NIV via a variety of interfaces but also from frequent and aggressive MAC to expel secretions and maintain or return $Spo_2 > 95\%$.

In our earlier study of extubation/decannulation to NIV, considering the extent of need for NIV, age, VC, and maximum assisted CPF, only assisted CPF ≥ 160 L/m predicted success in 62 extubation/decannulation attempts on 49 consecutive patients, including 34 with no ventilator-free breathing ability.[22] None of the 15 attempts on those with maximum CPF < 160 L/m succeeded, as opposed to an 87% first-attempt extubation success rate in this study. The most likely reasons for the difference between then and now are: baseline Spo_2 criterion for extubation of 92% vs 95%, and thus the earlier patients had more residual airway secretions or lung disease at extubation; 5% vs 39% of patients with pre-extubation experience with NIV and MAC; less hospital staff experience with NIV and MAC; 50 of 62 patients were decanulated, not extubated; the patients were in various hospital locations; and MAC was used less often and without family involvement.[22]

The 87% first-attempt extubation success rate on patients with maximum CPF < 160 L/m in this study is greater than the 82.4% (61 of 74) success rate reported for extubating NIV-dependent infants with spinal muscular atrophy type 1 (SMA 1), according to an almost identical protocol.[42] The difference may be the result of the ability of these patients, as opposed to babies, to cooperate with NIV and MAC. The SMA 1 infants may have also had more severe bulbar-innervated muscle dysfunction. Thus, while higher than those

Table 4—*Postextubation Long-Term Noninvasive Ventilation Use for 155 Patients*

Characteristics	Weaned in 1 wk	Weaned to Part-Time NIV	Unweanable
Subjects, n	23	62	72
VC at extubation, mL	423 ± 157	344 ± 152	259 ± 179
CPF at extubation, L/min	204 ± 58	179 ± 73	158 ± 85
VC 6 mo postextubation, mL	1797 ± 683	896 ± 649	502 ± 353
Duration ventilator use, mo (range)	. . .	48 ± 55 (1-204)	71 ± 62 (1-228)

See Tables 1 and 3 for expansion of abbreviations.

for a comparable infant population, the success rate was significantly less (87% vs 100%) ($P < .05$) than for patients with assisted CPF ≥ 160 L/m. Unmeasurable assisted CPFs indicate an inability to close the glottis and are associated with stridor, saliva aspiration, and less effective NIV and MAC.

An NIV/MAC protocol has been used to avoid over 100 hospitalizations for continuously ventilator-dependent (NIV) patients with NMD.[20,35] Here we considered unweanable patients with NMD for whom intubation could not be avoided. Upon extubation, most patients with a VC of 200 mL or more eventually were weaned from continuous to part-time NIV by taking fewer and fewer mouthpiece IPPVs. Thus, the paradigm of weaning then extubation can be changed to extubation to permit self-weaning for patients with NMD. The notion that early tracheotomy after intubation somehow facilitates ventilator weaning[43] should be reassessed for patients with NMD. NIV is also associated with over 75% fewer ventilator-associated pneumonias.[6,44] Use of mouthpieces rather than "masks" interfaced in acute-care facilitated speech, oral intake, comfort, and glossopharyngeal breathing[45]; eliminated the risk of skin pressure sores; and permitted air stacking to maintain pulmonary compliance,[37,45] diminish atelectasis, and facilitate manually assisted coughing.

The purpose here was not to facilitate ventilator weaning or to consider long-term outcomes, but to extubate unweanable patients. Benefits included no mortality, fewer days intubated, no tracheostomies, and return home. Decannulation, too, can facilitate ventilator weaning.[46] Avoidance of tracheostomy for continuous ventilator (NIV) users can also better maintain quality of life,[47-49] significantly diminish long-term pneumonia and respiratory hospitalization rates,[50] maximize ventilator-free breathing,[46] and facilitate return home.[49]

In conclusion, unweanable intubated patients with NMD who satisfy specific criteria can be successfully extubated to full NIV and MAC. Patients with measurable assisted cough flows should no longer be advised to refuse intubation for fear of extubation failure and tracheotomy. We no longer consider tracheotomy for any ventilator-dependent patients with NMD who satisfy Table 1 criteria, and now offer extubation to most with CPF < 160 L/m.

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Author contributions: *Dr Bach:* wrote all drafts of the paper and, with Dr Hamdani, extubated all the patients in New Jersey.
Mr Gonçalves: performed the extubations on all of the patients in Portugal, gathered data on the Portuguese patients, and added material to the text of the manuscript.
Dr Hamdani: performed the extubations on some of the patients in New Jersey, gathered data on the New Jersey patients, and added material to the text of the manuscript.
Dr Winck: oversaw the extubations on all of the Portuguese patients and added material to the introductory and "Discussion" sections of the manuscript.
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