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## **Extubation of Patients with Neuromuscular Weakness: A New Management Paradigm**

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## Abbreviations

ALS - amyotrophic lateral sclerosis

ARF - acute respiratory failure

CCM – critical care myopathy

CPF – cough peak flows

DMD - Duchenne muscular dystrophy

ICU – intensive care unit

IPPV - intermittent positive pressure ventilation

MAC - mechanically assisted coughing (mechanical insufflation-exsufflation with exsufflation-timed abdominal thrust)

NIV - noninvasive mechanical ventilation including noninvasive IPPV and high span bi-level PAP

NMD - neuromuscular disease

PAP – positive airway pressure

SBT – spontaneous breathing trial

SCI - spinal cord injury

SMA 1 – spinal muscular atrophy type 1

SpO2 – Pulse oxyhemoglobin saturation

VC - vital capacity

## ABSTRACT

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

**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 SBTs before or after extubation. Once SpO<sub>2</sub> was maintained  $\geq 95\%$  in ambient air patients were extubated to full noninvasive ventilation (NIV) support and aggressive mechanically assisted coughing (MAC). Rather than oxygen, NIV and MAC were used to maintain or return SpO<sub>2</sub> to  $\geq 95\%$ . Extubation success was defined as not requiring re-intubation during the hospitalization and was considered as a function of diagnosis, pre-intubation NIV experience, and vital capacity (VC) and assisted cough peak flows (CPF) at extubation.

**Results:** Before hospitalization 96 (61%) patients had no experience with NIV, 41 (26%) used it less than 24 hrs. per day, and 20 (13%) were continuously NIV dependent. First attempt protocol extubation success rate was 95% (149 patients). All 98 extubation attempts on patients with assisted CPF  $\geq 160$  L/m were successful. Dependence on continuous NIV and duration of dependence prior to intubation correlated with extubation success ( $p < 0.005$ ). Six of 8 patients who initially failed extubation succeeded on subsequent attempts, so only two with no measurable assisted CPF underwent tracheotomy.

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

## INTRODUCTION

Conventional extubation attempts follow successful “spontaneous breathing trials (SBTs)” and the passing of ventilator weaning parameters,<sup>1</sup> otherwise patients undergo tracheotomy. Patients are often extubated to supplemental oxygen and bi-level positive airway pressure (PAP) but settings are infrequently reported;<sup>2</sup> and extubation studies report very few if any NMD patients i.e. 18 of 162,<sup>3</sup> 17 of 900<sup>4</sup> and completely exclude unweanable patients.<sup>5-7</sup>

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

There are no guidelines for extubating unweanable NMD/CCM patients. Many are NIV dependent with no autonomous breathing ability for years before being intubated and they refuse tracheotomy.<sup>17-20</sup> Further, these patients can have ineffective CPF which can result in extubation failure due to airway secretion accumulation<sup>21-24</sup> but very few studies reported CPF<sup>2,21</sup> and none systematically used mechanically assisted coughing (MAC).<sup>4,24</sup> There are no “ventilator weaning parameters” that address the ability to expel secretions.

With success in decannulating unweanable traumatic tetraplegics and others to continuous NIV and MAC,<sup>20,25-27</sup> we used similar criteria to extubate unweanable NMD/CCM patients and report success rates.

## METHODS

The data were gathered in two centers, 113 patients in New Jersey and 44 in Portugal, using Table 1 inclusion criteria. The study was approved by the institutions’ review boards. All intubated patients were managed conventionally except for the use of

mechanically assisted coughing (MAC) via the tube. Although virtually unknown in critical care, MAC has been instrumental in avoiding pneumonia, respiratory failure, and hospitalizations for NIV dependent NMD patients.<sup>28-30</sup> Vital capacities (VCs) (Wright Mark 3 spirometer, Ferraris Ltd., London) and unassisted and assisted CPF (Access Peak Flow Meter, Health Scan Products Inc., Cedar Grove, New Jersey) were measured within 12 months before intubation for the local (Group 1) patients. 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.<sup>31-33</sup> Patients had to experience immediate distress, precipitous oxyhemoglobin desaturation and hypercapnia without stabilization before return to full ventilatory support both before and immediately post-extubation. Local patients were considered 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 risk for extubation failure is high when assisted CPF can not attain 160 L/m.<sup>22</sup> Considering that advanced a verbal bulbar ALS patients can rarely attain CPF of 160 L/m<sup>34,35</sup> we generally did not accept such patients for transfer (exclusion criteria). Local patients with 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.<sup>20,35</sup>

## Protocol

While intubated, sufficient ventilatory support was used to maintain normocapnia and normal respiratory rates. MAC (CoughAssist<sup>TM</sup>, Respironics Inc., Murrysville, Pa) was used at 40 to -40 cm H<sub>2</sub>O 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 minutes to maintain or return SpO<sub>2</sub> to  $\geq 95\%$  in ambient air. Tracheotomy would have been recommended if Table 1 criteria could not be met within 2 weeks of transfer.

Once Table 1 criteria were met the oro or nasogastric tube was removed to facilitate post-extubation nasal NIV. The patient was then extubated directly to NIV on assist/control 800 to 1500 ml, rate 10-14/min in ambient air. Pressure control of at least 18 cm H<sub>2</sub>O was used if abdominal distension developed. The NIV was provided via a combination of nasal, oro-nasal, and mouth piece interfaces.<sup>36</sup> Assisted CPF, CPF obtained by abdominal thrust following air stacking, were measured within 3 hours as the patient received full volume-cycled NIV support. Patients kept 15 mm angled mouth pieces accessible (Figure 1) and weaned themselves, when possible, by taking fewer and fewer intermittent positive pressure ventilations (IPPVs) as tolerated. Diurnal nasal IPPV was used for those who could not secure the mouth piece. Patients took as much of the delivered volumes as desired. They used nasal or oronasal interfaces (Figures 2, 3) for night time ventilation. For episodes of SpO<sub>2</sub><95%: ventilator positive inspiratory pressure (PIP), interface or tubing air leakage, CO<sub>2</sub> 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.<sup>37</sup> Once “air stacked”, an abdominal thrust was provided to manually assist the cough,<sup>29,37</sup> and



these assisted CPF 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 oro-nasal interfaces up to every 20 min until SpO<sub>2</sub> no longer dipped below 95% and the patients felt clear of secretions. In 7 cases, post-extubation oral intake was considered unsafe so open modified Stamm gastrostomies were performed under local anesthesia using NIV as in Figure 2 without complication.<sup>38</sup>

Extubation was considered successful if the patient was discharged without re-intubation. When re-intubation was necessary, the patient was again extubated after achieving Table 1 criteria. Multiple failures necessitated tracheotomy. Extubation success was considered as a function of diagnosis, patient group, VC, CPF, and pre-intubation NIV experience. VC was measured 3-6 months after extubation. Total days intubated were compared pre- and post-transfer.

### **Statistical Analysis**

Results are expressed as mean±standard deviation. Statistical analyses were carried out using SPSS 12.0. Differences in means were compared using the Wilcoxon rank test. A p value ≤0.05 was considered significant. Univariate comparisons of potential predictive factors for failure (F) or success (S) were run with the Fisher Exact Test for categorical variables and Wilcoxon rank-sums test for continuous variables.

## **RESULTS**

The 157 patients, mean age 37±21 years, included 139 (89%) with NMD who were intubated for acute respiratory failure/compromise (ARF) due to pneumonia and/or surgery and 18 CCM patients (11%). The 74 local patients (Group 1) were intubated at our

institutions and 83 others were intubated elsewhere. Demographic data are in Table 2. Group VC and CPF data are in Table 3. Twenty (13%) of the 157 patients had been continuously NIV dependent for 12.2 (range=1 to 47) years before being intubated. Forty-one (26%) used NIV part-time (<24 hrs/day) and 96 (61%) used no NIV before intubation. All patients satisfied Table 1 criteria in less than 2 weeks.

Univariate comparisons of potential predictive factors for extubation success yielded significant differences for continuous NIV use ( $p<0.0001$ ) and for duration of continuous NIV use and MAC prior to intubation ( $p=0.0038$ ) indicating that experience with NIV and MAC was significant in predicting success. Given only 15 failures in 8 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  $\text{CPF} \geq 160$  L/m were successful. Fifty-nine of 74 attempts (80%) on patients with  $\text{CPF} < 160$  L/m were successful including 52 of 60 (87%) at first extubation. Six patients who initially failed, succeeded on 2<sup>nd</sup> (4 patients) and 3<sup>rd</sup> (3 patients) attempts. One with advanced bulbar ALS and one with facioscapulohumeral muscular dystrophy, both with no measurable assisted CPF, failed a total of 5 extubations and underwent tracheotomy. Only one of the eight who failed any extubation attempt had pre-intubation experience with NIV and MAC but she and several others had suboptimal post-extubation care provider support for aggressive MAC. She and eight bulbar ALS patients with little residual bulbar-innervated muscle function required oro-nasal interfaces for a closed system of post-extubation NIV (Hybrid NE, Teleflex Medical, Research Triangle Park, NC) (Figure 2). All nine had gastrostomy tubes for total enteral nutrition. One Lipseal nocturnal NIV user for 28 years prior to intubation was extubated back to Lipseal NIV (Figure 3).<sup>36</sup>

The VC, CPF, and duration of NIV use as a function of post-extubation weaning capacity are in Table 4. Weaning from full to part-time NIV took 3-21 days and was usually accomplished at home. As supine VCs increased to approach 1000 ml we encouraged patients to sleep without NIV with SpO<sub>2</sub> and EtCO<sub>2</sub> monitoring and when these remained stable for 2 weeks to discontinue NIV. The mean extubation VC and assisted CPF for the 29 patients  $\geq 18$  years of age who were successfully extubated at first attempt despite assisted CPF < 160 L/m were  $245 \pm 114$  (range 120-620) ml and  $97 \pm 39$  (range 0-150) L/m, respectively. No clinically or radiographically apparent barotrauma was noted for any patients.

The 83 Group 2 and 3 patients had been intubated for  $11 \pm 9.1$  (range=1-80) days before transfer and  $2 \pm 1$  (range=1-11) days on our units before extubation ( $p < 0.005$ ). Upon admission, on 21% FiO<sub>2</sub>, 71 (85%) of the patients' SpO<sub>2</sub>s settled below 95%. Increased NIV support to normalize CO<sub>2</sub> and especially MAC normalized SpO<sub>2</sub> generally within 24 to 48 hours to satisfy a criterion for extubation.

The intensivists and respiratory therapists estimated that noninvasive necessitated more time than invasive respiratory management. The extubation itself required about 1.5 hours 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 tracheostomy patient were discharged home. One hundred thirty-four patients are alive using NIV (Table 4). Nine patients (6%) died of cardiac failure, 6 (4%) from lung disease/respiratory failure, 2 (2%) with bulbar-ALS died after tracheotomy from sepsis and decubiti, and 9 (6%) died of unknown causes. Although offered, no patients accepted tracheotomy following successful extubation.

## DISCUSSION

There are no extubation studies on continuously NIV dependent NMD patients.<sup>6</sup> A recent controlled post-extubation 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<sub>2</sub> alone or in conjunction with bi-level PAP at spans up to 14 cm H<sub>2</sub>O, pressures inadequate for normal alveolar ventilation for our patients.<sup>39</sup> A meta-analysis of 12 extubation studies to bi-level PAP demonstrated decreased mortality, ventilator associated pneumonia, length of stay, and resort to tracheotomy but unweanable NMD patients were uniformly excluded.<sup>6</sup> Eligibility for extubation was based on “readiness for weaning” and failure of SBTs after 30 minutes or more.<sup>6</sup> While this justifies extubation to NIV for primarily lung/airways disease patients with some autonomous breathing ability and for whom SpO<sub>2</sub>>90% may be acceptable with or without supplemental O<sub>2</sub>, 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<sub>2</sub> 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<sub>2</sub> below 95% despite continuous NIV and MAC in ambient air is the indication for tracheotomy.<sup>35</sup>

Besides hypoventilation, ineffective CPF have been associated with extubation failure.<sup>21,22</sup> 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<sub>2</sub>.<sup>30,40,41</sup> Our success stemmed not only from providing

continuous full-setting NIV via a variety of interfaces, but from frequent and aggressive MAC to expel secretions and maintain or return SpO<sub>2</sub> over 95%.

In our earlier study of extubation/decannulation to NIV considering extent of need for NIV, age, VC, and maximum assisted CPF, only assisted CPF $\geq$ 160 L/m predicted success in 62 extubation/decannulation attempts on 49 consecutive patients including 34 with no ventilator-free breathing ability.<sup>22</sup> None of the 15 attempts on those with maximum CPF<160 L/m succeeded as opposed to an 87% first extubation success rate in this study. The most likely reasons for the difference between then and now are: baseline SpO<sub>2</sub> criterion for extubation 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 were decannulated, not extubated; the patients were in various hospital locations; and MAC was used less often and without family involvement.<sup>22</sup>

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 SMA type 1 according to an almost identical protocol.<sup>42</sup> The difference may be due to 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 for a comparable infant population, the success rate was significantly less (87% vs. 100%) ( $p<0.05$ ) than for patients with assisted CPF $\geq$ 160 L/m. Unmeasurable assisted CPF indicate 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 (NIV) dependent NMD patients.<sup>20,35</sup> Here we considered

unweanable NMD patients for whom intubation could not be avoided. Upon extubation most patients with VC of 200 ml or more eventually weaned from continuous to part-time NIV by taking fewer and fewer mouth piece IPPVs. Thus, the paradigm of weaning then extubation can be changed to extubation to permit self-weaning for NMD patients. The notion that early tracheotomy after intubation somehow facilitates ventilator weaning<sup>43</sup> should be reassessed for patients with NMD. NIV is also associated with over 75% fewer ventilator associated pneumonias.<sup>6,44</sup> Use of mouth pieces rather than “mask” interfaces in acute care facilitated speech, oral intake, comfort, glossopharyngeal breathing,<sup>45</sup> eliminated risk of skin pressure sores, and permitted air stacking to maintain pulmonary compliance,<sup>37,45</sup> diminish atelectasis, and facilitate manually assisted coughing,.

The purpose here was not to facilitate ventilator weaning nor 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.<sup>46</sup> Avoidance of tracheostomy for continuous ventilator (NIV) users can also better maintain quality of life,<sup>47-49</sup> significantly diminish long-term pneumonia and respiratory hospitalization rates,<sup>50</sup> maximize ventilator-free breathing,<sup>46</sup> and facilitate return home.<sup>49</sup>

In conclusion, unweanable intubated NMD patients 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 NMD patients who satisfy Table 1 criteria and now offer extubation to most with CPF less than 160 L/m.

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**Author activities:**

Dr. John Bach: wrote all drafts of the paper and, all with Dr. Hamdani, extubated all the patients at University Hospital, in Newark, New Jersey.

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. Miguel Gonçalves: performed the extubations on all of the patients in Porto, Portugal, gathered data on the Portuguese patients, and added material to the text of the manuscript.

Dr. Joao-Carlos Winck: over saw the extubations on all of the Porto patients and added material to the Introduction and Discussion of the manuscript.

Table 1- Extubation Criteria for Unweanable Ventilator Dependent Patients
<ul style="list-style-type: none"><li>• Afebrile and normal white blood cell count</li><li>• Age 4 years and older</li><li>• No ventilator-free breathing tolerance with 7 cm pressure support in ambient air on the basis of neuromuscular disease or critical care myopathy</li><li>• Vital capacity less than 20% of normal</li><li>• PaCO<sub>2</sub> 40 mm Hg or less at peak inspiratory pressures less than 35 cm H<sub>2</sub>O on full setting assist/control mode at a rate of 10-13/minute</li><li>• Oxyhemoglobin saturation (SpO<sub>2</sub>) ≥ 95% for 12 hours or more in ambient air</li><li>• All oxyhemoglobin desaturations below 95% reversed by mechanically assisted coughing and suctioning via translaryngeal tube</li><li>• Fully alert and cooperative, receiving no sedative medications</li><li>• Chest radiograph abnormalities cleared or clearing</li><li>• Air leakage via upper airway sufficient for vocalization upon cuff deflation</li></ul>



**Table 2 – Demographic Data**

	<b>Group 1</b>	<b>Group 2</b>	<b>Group 3</b>	<b>Total</b>
<b>N (%)</b>	74 (47%)	45 (29%)	38 (24%)	157 (100%)
<b>Sex (%)</b>	52 male(70%) 22 female(30%)	28 male(62%) 17 female(38%)	17 male(45%) 21 female(55%)	97 male(62%) 60 female(38%)
<b>Diagnoses (%)</b>	ICUMy–15(20%) SCI – 13(18%) ALS – 11(15%) MG – 9(12%) MD – 8(11%) oNMD-8(11%) SMA- 5(7%) DMD – 3(4%) PPS – 2(3%)	SMA- 10(22%) MD – 9(20%) DMD – 8(18%) MG – 6(13%) PPS – 4(9%) oNMD- 4(9%) ALS – 3(7%) SCI – 1(2%)	SMA- 10 (26%) DMD – 9(24%) MD – 5(13%) PPS – 5(13%) oNMD- 4(11%) SCI – 3(8%) ALS – 2(5%)	SMA- 25 (16%) MD – 22 (14%) DMD – 20(13%) SCI – 17 (11%) ALS – 16(10%) oNMD- 16 (10%) ICUMy–15(10%) MG – 15 (10%) PPS – 11 (7%)
<b>Use of NIV pre-intubation</b>	NoNIV–51 (69%) Cont – 10(14%) Noct – 13(17%)	noNIV-24(53%) Cont – 7(16%) Noct – 14(31%)	noNIV– 21 (55%) Cont – 3 (8%) Noct – 14(37%)	noNIV– 96(61%) Cont – 20 (13%) Noct – 41(26%)

Diagnoses: amyotrophic lateral sclerosis (ALS), Duchenne muscular dystrophy (DMD), intensive care unit acquired NMD (ICUMy), muscular dystrophy (MD), myasthenia gravis (MG), post-polio syndrome (PPS), spinal cord injury (SCI), spinal muscular atrophy (SMA) including types 1, 2 and 3, and other NMD (oNMD); NIV-noninvasive ventilation; Cont – continuous NIV; Noct – nocturnal NIV; Group 1 – local patients; Group 2 – patients transferred after failing extubations in other institutions; Group 3 - patients transferred after failing multiple SBT's in other institutions.

**Table 3 - Pulmonary function**

	<b>Group 1</b>	<b>Group 2</b>	<b>Group 3</b>	<b>Total</b>
<b>N (%)</b>	74 (47%)	45 (29)	37 (24%)	157
<b>Assisted CPF at extubation (L/min)</b>	187 ± 85	162 ± 86	178 ± 62	177 ± 77
<b>VC (ml) at extubation</b>	355 ± 171	273 ± 189	295 ± 155	315 ± 173
<b>VC (ml) 3-6 months post- extubation</b>	1121±748	709±679	617±412	877±698

Intergroup differences were not statistically significantly different except for post-extubation VC with that of Group 1 being greater than for Groups 2 and 3 ( $p < 0.05$ )

VC – vital capacity; CPF – cough peak flows

Table 4

## Post-Extubation Long-Term Noninvasive Ventilation Use for 155 Patients

	Weaned in 1 week	Weaned to part-time NIV (mos)	Unweanable (mos)
N	23	62	72
VC at extubation	423±157	344±152	259±179
CPF at extubation	204±58	179±73	158±85
VC 6 mos. later	1797±683	896±649	502±353
Duration ventilator use		48±55 (1-204)	71±62(1-228)

VC=vital capacity (ml)

CPF=cough peak flows (L/min)

## Legends

### Figure 1

Ten 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 15 mm angled mouth piece (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.

### Figure 2

Twenty year old man with Duchenne muscular dystrophy transferred for extubation after failing 3 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 (Hybrid, Teleflex Medical, Research Triangle Park, NC) for nocturnal ventilatory support. His VC was 260 ml at extubation October 2007 and 720 ml in July 2009. The patient provided written consent for the use of this photograph.

### Figure 3

Fifty-nine year old woman with spinal cord injury at birth, 31 years of dependence on daytime mouth piece, and nocturnal lip seal (seen here) ventilation. She was extubated back to NIV despite a vital capacity of 130 ml and no autonomous breathing ability and continued to use simple 15 mm angled mouth piece ventilation for daytime ventilatory support and lip seal overnight with nose clipped to prevent air leakage. Current VC is 340 ml. She is employed full-time as a psychologist. The patient provided written consent for the use of this photograph.

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10 year old girl with neurofibromatosis status-post spinal tumor resection, extubated with a vital capacity of 180 ml and no ventilator-free breathing ability, using a 15 ml angled mouth piece for ventilatory support.

1004x753mm (72 x 72 DPI)

The patient provided written consent for the use of this photograph.



22 year old man with Duchenne muscular dystrophy transferred for extubation after failing 3 attempts over a 26 day period. He uses a 15 mm angled mouth piece as in Figure 1, for daytime ventilatory support and a lipseal phalange with nasal prongs for nocturnal ventilatory support. His VC was 260 ml at extubation.

1004x753mm (72 x 72 DPI)

The patient provided written consent for the use of this photograph.



59 year old woman with spinal cord injury at birth, 31 years of continuous dependence on noninvasive ventilation (NIV) using a mouth piece during the day and lipseal (see here) for sleep. She was extubated back to NIV despite of VC of 130 ml and no autonomous breathing ability.

288x216mm (180 x 180 DPI)

The patient provided written consent for the use of this photograph.



## Extubation of Patients with Neuromuscular Weakness: A New Management Paradigm

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