Mechanical In-exsufflation-Expiratory Flows as Indication for Tracheostomy Tube Decannulation

Case Studies

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Abstract: Mechanical insufflation exsufflation-expiratory flows (MIE-EFs) correlate with upper airway patency. Patients dependent on continuous noninvasive ventilatory support with severe spinal muscular atrophy type 1, now over 20 yrs old, have used MIE sufficiently effectively along with continuous noninvasive ventilatory support to avoid tracheotomy indefinitely. Although MIE-EFs can apparently decrease in amyotrophic lateral sclerosis to necessitate tracheotomy, they can increase over time and remain effective in all spinal muscular atrophy types. Two cases demonstrate an association between increasing MIE-EF and, ultimately, successful decannulation of a patient with spinal muscular atrophy type 2 who was continuous tracheostomy mechanical ventilation dependent and a patient with obesity hypoventilation syndrome. Only when MIE-EF increased to exceed 200 l/min did the decannulations succeed. Definitive noninvasive management (continuous noninvasive ventilatory support) of these patients may be possible only when MIE is effective, and the greater the MIE-EF, the greater its effectiveness. Thus, increasing MIE-EF can signal resolution of upper airway obstruction sufficiently to permit decannulation whether a patient is ventilator dependent or not.

Key Words: Mechanical Insufflation-Exsufflation, Successful Decannulation, Spinal Muscular Atrophy

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Mechanical insufflation-exsufflation (MIE) is ideally used via invasive or noninvasive interfaces at insufflation and exsufflation pressures of 40 to 70 cm H_2O.[1–3] The models T70 and E70 CoughAssists (Philips-Respironics Inc, Murrysville, PA) and the VOCSN ventilator (Ventec Life Systems, Seattle, WA) permit measurements of MIE exsufflation ("cough") flows (EFs). The greater the MIE-EF, the greater the upper airway patency and the more efficacious the expulsion of airway debris by MIE.[4]

In a recent amyotrophic lateral sclerosis (ALS) case study, over a 14-mo period, as vital capacity (VC) decreased from 1670 ml to 10 ml and continuous noninvasive ventilatory support (CNVS) became necessary for survival,[4] the MIE-EF also decreased from 320 l/min to 90 l/min, at which point it became ineffective at expulsing airway secretions; baseline pulse oxyhemoglobin saturation (SpO_2) decreased and remained below 95%; and tracheotomy became necessary to further prolong survival.[4–6] The decreasing MIE-EF in bulbar ALS patients is a result of stridor and the decreasing upper airway patency in the presence of upper motor neuron bulbar innervated muscle (BIM) dysfunction. Although decreasing over time in bulbar ALS patients, the MIE-EF does not generally decrease with advancing myopathic or lower motor neuron diseases.[4] Indeed, it has been noted that although MIE-EF may not exceed 100 l/min for infants with spinal muscular atrophy (SMA) type 1 who have small airways as well as severe BIM impairment, MIE-EF exceeds 200 l/min for consecutive CNVS-dependent SMA type 1 patients older than 20 yrs, none of whom have tracheostomy tubes. Therefore, there is a fundamental difference in the BIM impairment in lower *vs.* upper motor neuron involvement that permits MIE to be effective for the former but not the latter. The following case studies demonstrate an association between increasing MIE-EF and successful decannulation, and when MIE-EF are inadequate, possibly reversible obstructive lesions need to be sought. This study conforms to all Case Report guidelines and reports the required information accordingly (see Checklist, Supplemental Digital Content 1, http://links.lww.com/PHM/A640).

CASE STUDY 1

A 29-yr-old woman with SMA type 2 was intubated for pneumonia and acute on chronic respiratory failure. After failing all ventilator weaning parameters and spontaneous breathing trials, she underwent tracheotomy for continuous tracheostomy mechanical ventilation. She presented to us 2 mos later to be decannulated. Upon presentation, she was continuous tracheostomy mechanical ventilation dependent on "lung protection settings" of 300 ml, backup rate 20/m, but $PaCO_2$ was over 50 mm Hg and her VC was only 300 ml via the tube. After increasing her delivered volumes to normalize her CO_2 and decreasing the backup rate to 12/min, supplemental O_2 was

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permanently discontinued, her cuff was deflated and leak compensated by increasing the ventilator-delivered volumes to maintain normal CO_2 and to permit and encourage speech to recondition the glottis. After using MIE (T70 CoughAssist Respironics Inc, Murrysville, PA) via the tube hourly for 24 hrs with the cuff inflated, her VC increased to 320 ml, and the SpO_2 baseline remained normal, so she was decannulated and placed immediately on 700 ml volume preset nasal noninvasive ventilatory support (NVS). Her maximum MIE-EF was 114 l/min as read off the T70 CoughAssist screen; she had severe stridor and distress and required almost immediate replacement of the tracheostomy tube. Upper airway edema was suspected and she was placed on Decadron and decannulated 24 hrs later. This time, the stridor was almost equally severe and distress and re-cannulation again immediate, but MIE-EF had increased to 132 l/min. It was decided to continue the Decadron for three additional days then make a final attempt at decannulation. On the third attempt, the stridor was almost absent and MIE-EF was 225 l/min, but she still required continuous ventilatory support, that is, nasal CNVS on volume preset assist/control ventilation of 650 ml, rate l/min. She remained CNVS dependent via nasal interface with normal SpO_2 and no distress only while using it for 3 days, at which time she weaned to sleep-only NVS. Her lips were too weak for mouthpiece NVS. Post extubation, she used MIE with her family's assistance for decreases in SpO_2 below 95%. Her VC was still 320 ml. She was discharged to a local hotel and returned to our outpatient department 4 days later for follow-up, then returned home via commercial airline flight the following week.

CASE STUDY 2

A 340-lb, 5′5″ 53-yr-old woman with obesity hypoventilation syndrome was hospitalized for ventilatory failure, was intubated, underwent tracheotomy, was weaned from continuous tracheostomy mechanical ventilation, and was referred to us for decannulation. Her VC measured via the mouth when the tube was out and the ostomy covered was 900 ml. Although she was comfortably breathing via a trach collar, with the tube out and the ostomy covered, she became dyspneic and stridorous and had MIE-EF 75 l/min. Her tube was changed to a cuffless fenestrated #6 Shiley and she was referred to the otolaryngology department to assess the upper airway. Granulation tissue was removed. Seven weeks later, she returned for decannulation and was decannulated. Her VC measured via the mouth with the ostomy covered was 1170 ml; she no longer had dyspneic breathing via the upper airway, stridor was absent, and MIE-EF was 230 l/min, so she was successfully decannulated and remains so 4 yrs later.

DISCUSSION

The patient in case 1 was continuous tracheostomy mechanical ventilation dependent with no ventilator free-breathing ability, either via a trach collar or via the upper airway with the tube capped. Her chest radiograph demonstrated only chronic opacities. She received all nutrition via an indwelling gastrostomy tube since age 3. She failed decannulation twice before it succeeded. Although the VC remained unchanged during all three attempts, the MIE-EF improved progressively during 4 days of Decadron administration. This decreased upper airway edema and permitted greater MIE-EF. The effectiveness of MIE to expulse her prodigious airway secretions is the main reason ventilator unweanable patients can be extubated to CNVS with ventilatory pump failure with resort to tracheotomy.[1,2] Whereas saliva aspiration can be avoided indefinitely by head positioning,[7] it is the tracheostomy tube that typically causes airway secretions that cause tracheostomy mechanical ventilation users to need to be suctioned an average of 7.7 times per day.[8]

Many patients with severe SMA type 1 have no BIM function at all, have no ability to close the glottis, and have been CNVS dependent for over 20 yrs and yet have no tracheostomy tubes.[7] Although our babies with SMA type 1 have less than 100 l/min of MIE-EF, all four of our local CNVS-dependent patients with SMA type 1 who are older than 20 yrs and have no BIM function have MIE-EFs over 250 l/min. This indicates that despite the absence of any BIM function, MIE can effectively expulse airway secretions to permit them to avoid invasive mechanical ventilatory support.

Life has now been extended past age 50 for some patients with Duchenne muscular dystrophy by over 25 yrs of CNVS,[9] to age 25 in SMA type 1 by CNVS from as young as 4 mos of age, by over 60 yrs for postpoliomyelitis patients, by up to 38 yrs for high-level spinal cord tetraplegia,[2] by over 10 yrs for two of our patients with obesity hypoventilation, but only by a mean of 12.8 ± 16.2 mos for 33% of 360 patients with ALS.[10] The ALS patients developed increasing stridor and decreasing MIE-EF such that tracheotomy became necessary for continued survival.[1] Thus, while patients with myopathic and lower motor neuron disorders can become CNVS dependent and remain so, indefinitely, this is not the case for ALS patients. Whereas 760 CNVS-dependent patients with Duchenne muscular dystrophy, ALS, and SMA type 1 were reported from 22 centers in 2010, and all with Duchenne muscular dystrophy and SMA type 1 succeeded in becoming CNVS dependent, this was true for only 33% of the ALS patients who required tracheotomies before they could become CNVS dependent.[4]

Definitive noninvasive respiratory management is possible only with access to effective MIE when there are secretions to expulse and cough flows are inadequate to expulse them. The higher the MIE-EF, the greater its effectiveness. Thus, whereas the decreasing MIE-EF associated with advancing bulbar ALS signaled need for tracheotomy,[4] these cases demonstrate how increasing MIE-EF can signal resolution of upper airway obstruction to permit safe decannulation whether a patient is ventilator dependent or not.

Total absence of BIM function does not prevent noninvasive respiratory management, but upper motor neuron loss of BIM function and diminishing MIE-EF does.[4] In 2016, the authors analyzed CNVS outcomes as a function of myopathic, lower motor neuron, and upper motor neuron/central nervous system BIM impairment. Duration of part-time NVS and CNVS use was recorded and unassisted cough peak flows and MIE-EF were measured at each outpatient visit and before tracheostomy. It was found that about 200 l/min or more of MIE-EF correlated with successful noninvasive management using CNVS to thereby avoid tracheostomy.[11] It was also reported that cough peak flows of 160 l/min correlated with successful extubation of ventilator unweanable patients.[3] Thus, although there can be no absolute expiratory flow value that guarantees successful

transition from invasive to noninvasive respiratory management, flows of 150 to 200 l/min may be the minimum needed for successful long-term transition. These flows have become principal criteria for decannulation.[3] Reversal of upper airway edema or resection of granulation tissue can augment MIE-EF and permit successful decannulation as for these cases.

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