Optimum Insufflation Capacity and Peak Cough Flow in Neuromuscular Disorders

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Abstract

Rationale: For patients with neuromuscular disorders, lung insufflation with positive pressure is an accepted technique to increase inspiratory volume over VC to improve peak cough flow (PCF).

Objectives: The aim of the study was to determine the pressure or volume required to achieve the highest individual PCF.

Methods: In 40 patients dependent on noninvasive ventilation (VC, 16 ± 11% predicted; age, 20 ± 4 yr) and in 20 healthy control subjects, insufflation capacity (IC) was measured during titration from 10 to maximum 40 mbar using intermittent positive pressure breathing (IPPB) or the lung insufflation assist maneuver (LIAM) of the VENTI*logic* LS ventilator.

Measurements and Main Results: IPPB or LIAM titration resulted in a pressure–volume curve with an estimated total compliance of 0.23 ± 0.11 L/kPa in the patients and 1.0 ± 0.3 L/kPa in the controls and a plateau for IC at pressures between 30 and 40 mbar. IPPB or LIAM improved VC from 451 ± 229 ml to a maximum IC (IC_{max}) of 1,027 ± 329 ml, and PCF improved from 109 ± 45 to 202 ± 62 L/min ($P < 0.01$ for all). The highest individual PCF was achieved with 27 ± 6 mbar and an IC of 924 ± 379 ml, which was significantly below IC_{max} ($P < 0.01$).

Conclusions: A submaximal insufflation is ideal for generating the best individual PCF even in patients with severely reduced compliance of the respiratory system. Optimum insufflation capacity can be achieved using IPPB or LIAM with moderate pressures. Both techniques are equally effective and considered safe.

Keywords: lung insufflation maneuver; LIAM; intermittent positive pressure breathing; optimum insufflation capacity; peak cough flow

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In many inherited and acquired neuromuscular disorders (NMDs), respiratory complications pose a progressively increasing risk over the typical course of the disease. Inspiratory and expiratory muscle weakness, hypoventilation, and increasingly impaired cough competence cause frequent atelectasis and chest infection, which constitute the primary cause of morbidity and mortality in many NMDs (1, 2). For Duchenne muscular dystrophy and other NMDs, an incident rate of pneumonia has been reported that increases with disease progression (3).

Effective peak cough flow in healthy subjects exceeds values of 360 to 400 L/min (4, 5). A typical inspiration before coughing reaches 80 to 90% of VC. The minimum volume for generation of effective subsequent cough flows should be 50% of VC (6, 7). Glottic competence is the second essential prerequisite for sufficient cough because isometric compression of the inspired air with the glottis closed provides appropriate pressures and subsequent generation of high expiratory flow (8, 9). Peak cough flow (PCF) for mucus expectoration must exceed 160 to 200 L/min (3, 10). A PCF above 250 to 270 L/min has been shown to be sufficient to prevent pneumonia in patients with NMDs (3, 11).

Mechanical insufflation has been repeatedly proposed as a standard therapeutic measure to raise inspiratory VT over VC and thus to increase expiratory peak flows (10–15). Different active or passive insufflation techniques are available. Glossopharyngeal breathing can be performed by the patient without mechanical assistance but may not be possible with impaired function of bulbar muscles (16). Breath stacking can be performed with volume-cycled ventilators, but its success depends on the degree of glottic control (17). Insufflation with resuscitation bags and integrated one-way valves to compensate for bulbar weakness is feasible but requires several breaths and intermediate holds during filling of the bag (18). There are two techniques that use only positive pressure insufflation with continuous flow and avoid expiratory holds and stacking: intermittent positive pressure breathing (IPPB) (19–21) and the lung insufflation assist maneuver (LIAM). The latter recently became available as an integrated option in the VENTI*logic* LS ventilator (22). Whereas IPPB is a flow-controlled maneuver that is terminated at a preset maximum pressure, LIAM is pressure controlled with a preset insufflation time including a pressure plateau phase after the pressurization phase (Figure 1).

To date no study has investigated the individual insufflation volume and the corresponding pressure at which the best subsequent peak cough flows could be achieved. Typically, when administering deep lung insufflation, one aims at the deepest possible individual insufflation where the maximum volume that can be kept in the lungs by glottic closure has been defined as maximum insufflation capacity (IC_{max}) (11). The goal of this prospective study was to establish the relationship between the insufflation maneuver pressure, the resulting insufflation capacity, and the subsequent PCF. We hypothesized (*1*) that a certain insufflation maneuver pressure may be optimal to achieve the highest individual peak cough flow and (*2*) that this pressure is below the pressure needed to achieve the IC_{max}. The study was performed using two different techniques to allow conclusions to be drawn as to whether findings depend on certain techniques or on the fundamental effects of insufflation pressure.

Methods

The Ethics Committee of the University of Essen approved the study protocol, and all patients and healthy volunteers submitted informed written consent. During 2013 and 2014, 40 patients with various NMDs were included in the trial when they visited the hospital for routine examination. The protocol was published on ClinicalTrials.gov (Identifier: NCT01981915).

Study Population

The study population included 36 patients, 18 of whom had Duchenne muscular dystrophy(4 with spinal muscular atrophy and 14 with various congenital muscular dystrophies or myopathies). There were 25 male adolescents and young men and 11 young women (mean age, 20.1 ± 4.2 yr; range, 11–27 yr). Two young men with Duchenne muscular dystrophy, one young man with spinal muscular atrophy, and one young woman with congenital muscular dystrophy were enrolled in the IPPB and LIAM group; each group consisted of 20 participants. All patients had advanced respiratory compromise and received noninvasive ventilation; 10 patients used additional mouthpiece ventilation during the day. All but three patients were familiar with the IPPB device or a mechanical insufflator-exsufflator at the time they joined the study. The control group included 20 young and healthy volunteers (8 men and 12 women; mean age, 22 ± 4 yr).

Study Design

IPPB was used in one group of 20 patients and in the control group, and LIAM was administered in the other group of 20 patients. Baseline measurements of VC and peak cough flow were made with patients breathing spontaneously in a seated position. Insufflation was thereafter applied with increasing pressures, and insufflation capacity was measured during each insufflation. Subjects were instructed to close the glottis after each insufflation and to cough. PCF was measured with patients wearing a nose clip and performing a maximum cough into a pocket peak flow meter (PocketpeakTM; Ferraris Medical Ltd., Enfield, UK). Insufflation was delivered using a full face mask or a mouthpiece. The IPPB device (Alpha 200c; Salvia Medical GmbH and Co. KG, Kronberg, Germany) was set to a flow of 30 to 60 L/min to achieve an inspiration time of about 3 seconds. Inspiratory pressures started at a pressure of 10 mbar and were increased in increments of 5 to 20 mbar and in further increments of 2 mbar to a maximum of 40 mbar.

LIAM was administered as a single insufflation during noninvasive ventilation with the VENTI*logic* LS (Weinmann Medical Technology, Hamburg, Germany). The ventilator was set to the patient's personal settings, and the LIAM pressure was increased incrementally to 20 or 25 mbar (depending on the ventilation pressure) and then further as described for the IPPB group. Inspiration time was set to

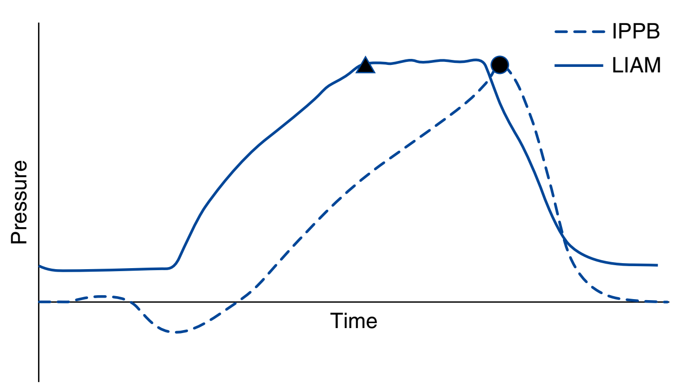

Figure 1. Qualitative representation of pressure profiles for the lung insufflation assist maneuver (LIAM) (*solid line*) and intermittent positive pressure breathing (IPPB) (*dotted line*) for one patient. For LIAM, peak maneuver pressure (delta pressure above normal inspiratory pressure), insufflation time, and expiratory time after insufflation are adjustable maneuver parameters.

3 seconds, which included a brief plateau phase. Characteristic pressure profiles are qualitatively shown in Figure 1.

Measurements were performed two or three times at each pressure level, and a data pair of volume and PCF for the highest insufflation capacity was documented. The individual pressure titration was stopped when further increase did not lead to additional improvement or if patients did not tolerate higher pressures. The highest insufflation volume was defined as IC_{max}. Insufflation volumes were plotted against insufflation pressures, allowing an estimation of the individual total compliance of chest wall and lungs (CL_{tot}). Then CL_{tot} was calculated from the linear section of this compliance curve (Δinsufflation volume/Δpressure).

Measurements

V_T and insufflation capacity (IC) were determined by integration of inspiratory flow. Flow measurement was performed using a pneumotachograph (PNT) (4700 series, 0–160 L/min; Hans Rudolph Inc., Shawnee, KS) and a differential pressure transducer (DIGIMA ± 2 cm H_2O; Special-Instruments GmbH, Noerdlingen, Germany). Airway pressure was derived at a T-piece using a pressure transducer (0–80 cm H_2O) (DIGIMA; Special-Instruments GmbH). The PNT and T-piece were positioned between the hose system and the patient interface. Analog signals were digitized at 100 Hz using a data acquisition device (USB 6009; National Instruments, Austin, TX). Data were visualized and stored using customized Labview Software (National Instruments). The pressure signal was calibrated at 0 and 40 mbar. The flow signal was calibrated at 0 L/min and by forcing an air bolus through the PNT using a 1,000-ml syringe (Care Fusion, Germany GmbH, Höchberg, Germany) until the integrated volume was in a range of 1,000 ml ± 0.4%.

Statistical Analysis

Analysis was performed with Statistica 6.0 (StatSoft, Inc., Tulsa, OK). The impact of insufflation on VC and PCF was tested using the paired Wilcoxon test. Group comparison between IC_{max} and optimum insufflation capacity (OIC) was tested using ANOVA. Interrelationships between VC and PCF were analyzed using the Spearman's rank correlation. Results are presented as mean ± SD. A P value <0.05 was considered significant.

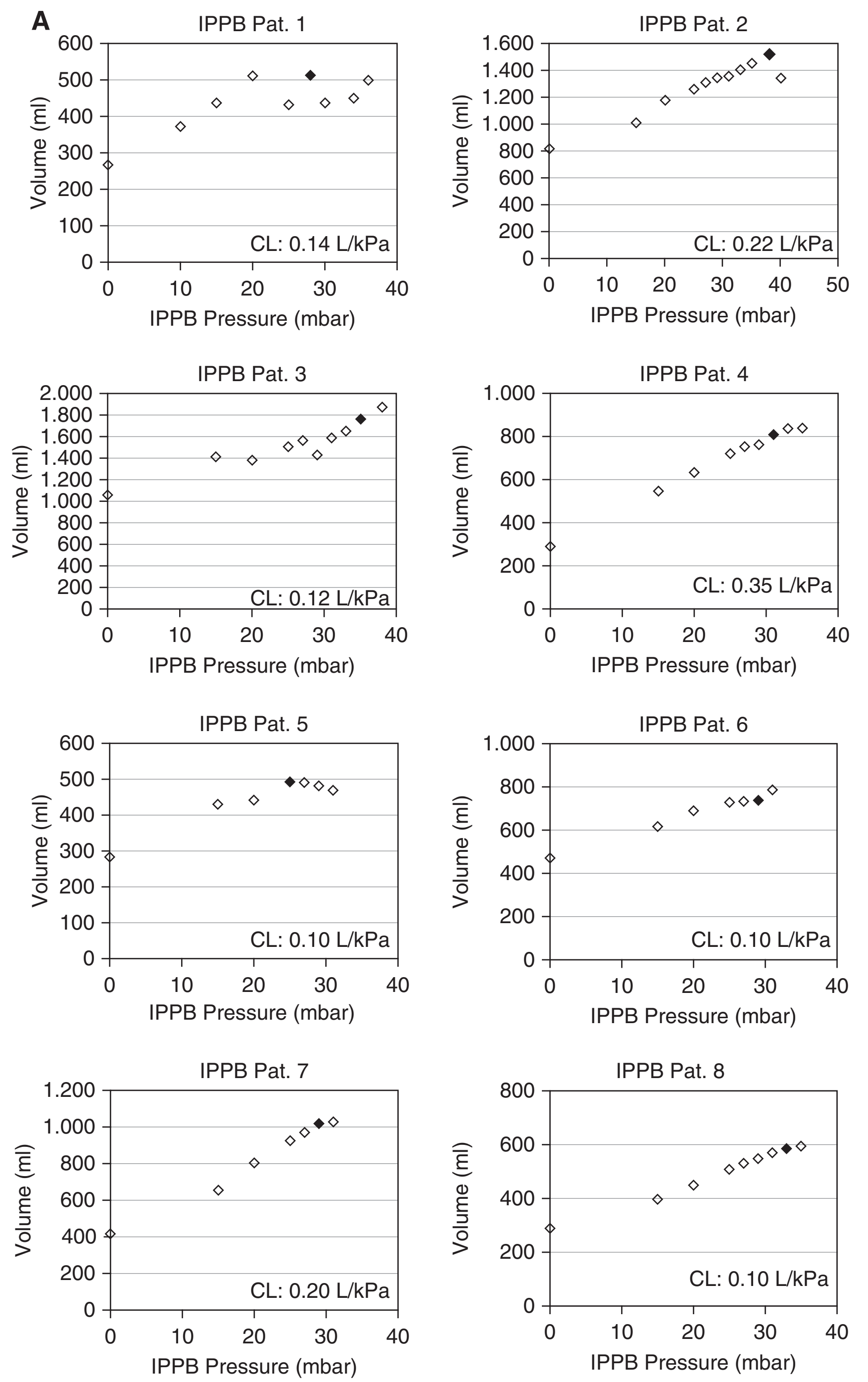

Figure 2. Impact of intermittent positive pressure breathing (IPPB) titration on insufflation capacity (ml) in patients 1 through 8 (*A*) and in patients 9 through 14 (*B*). Impact of lung insufflation assist maneuver (LIAM) titration on insufflation capacity (ml) in patients 1 through 8 (*C*) and in patients 9 through 15 (*D*). *Open diamond*: preset pressure (*x* axis) and highest corresponding insufflation capacity (*y* axis). *Closed diamond*: pressure/insufflation capacity resulting in the highest peak cough flow. CL = total compliance.

Results

Six patients in the IPPB group and five patients in the LIAM group were unable to perform glottis closure after insufflation and therefore could not adequately hold the insufflated volume and increase their PCF.

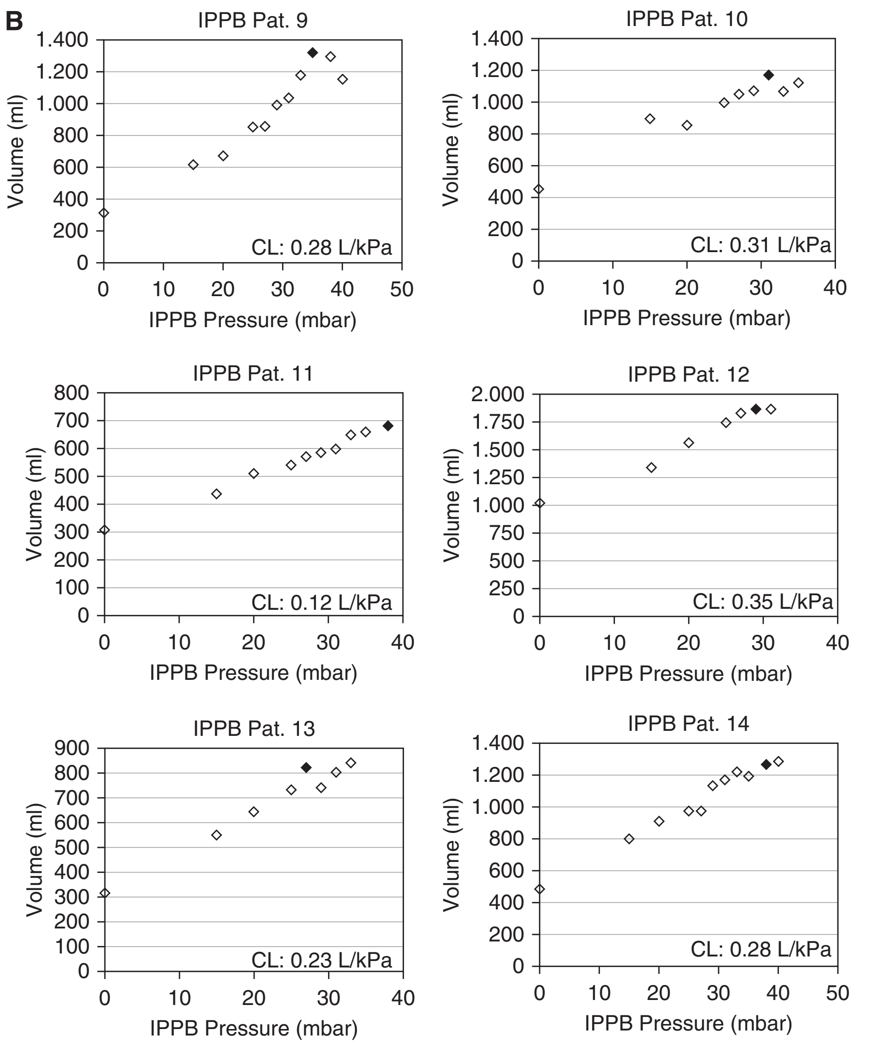

Figure 2. (Continued).

The remaining 29 patients presented with advanced respiratory impairment. Characteristics were similar in both groups, with no significant differences between the IPPB and the LIAM groups in age (22 ± 4 vs. 20 ± 4 yr), baseline VC (483 ± 276 ml [15 ± 8% predicted] vs. 425 ± 181 ml [17 ± 12% predicted]), and baseline PCF (109 ± 45 vs. 110 ± 45 L/min).

There was a close correlation between VC (ml) and PCF ($r = 0.80$; $P < 0.01$) and between VC % predicted and PCF ($r = 0.67$; $P < 0.01$).

Impact of IPPB on IC and PCF

In 11 of 14 patients, IC_{max} was identified by titrating the pressure until no further volume increments could be achieved. In 3 of 14 patients, titration was stopped before reaching the highest possible volume because further increase could not be tolerated.

The pressure–volume curves of the IPPB titration are displayed in Figures 2A and 2B. The mean estimated CL_{tot} of all 14 patients was 0.21 ± 0.1 L/kPa.

Insufflation with IPPB resulted in an IC_{max} of 134 ± 68% above baseline VC (Table 1; Figure 3A). The OIC to achieve the best PCF was below IC_{max} in 13 patients and equal to IC_{max} in one patient ($P < 0.01$). OIC was 89% of IC_{max} ($P < 0.01$). The pressure to reach OIC was significantly below the IC_{max} pressure ($P < 0.01$) (Table 1). IPPB augmentation improved PCF from 109 ± 43.3 to 201.4 ± 71.9 L/min ($P < 0.001$) (Figure 3B).

Impact of LIAM on IC and PCF

In 11 of 15 patients, IC_{max} was identified by titrating the pressure until no further volume increments could be achieved. In 4 of 15 patients, titration was stopped before reaching the highest possible volume because further increase could not be tolerated.

The pressure–volume curves of the LIAM titration are displayed in Figures 2C and 2D. The mean estimated CL_{tot} of all 15 patients was 0.25 ± 0.12 L/kPa.

Insufflation with LIAM resulted in an IC_{max} 150 ± 79% above baseline VC (Table 2; Figure 4A). The OIC to achieve the best PCF was below IC_{max} in 14 patients and was equal to IC_{max} in one patient ($P < 0.01$). OIC was 91% of IC_{max}. The pressure to reach OIC was significantly below the IC_{max} pressure ($P < 0.05$) (Table 2). LIAM augmentation improved PCF from 110 ± 49.1 to 204.7 ± 52.2 L/min ($P < 0.001$) (Figure 4B).

Impact of Passive Lung Insufflation in Healthy Control Subjects

In four subjects, no reliable pressure–volume curves could be obtained due to leakage or poor adherence to the protocol. In the remaining 16 subjects, passive lung insufflation with IPPB resulted in a pressure–volume curve with a slope of the straight line of 1.0 ± 0.2 L/kPa (CL_{tot}); an upper inflection point at 30.8 + 2.9 mbar and 3,480 + 821 ml, respectively; and a plateau. Cumulative pressure–volume curves of the patients and the control subjects are displayed in Figure 5.

Limitations of the Study

Flow and IC were measured during inspiration. IC assessment based on expiratory flow after insufflation may be a superior method because potential overestimation due to leakage could be overlooked. However, our method allowed us to switch to PCF measurement directly after insufflation within each breath and had the advantage of generating a data pair of IC and PCF. Leakage was closely monitored, and data were considered valid only when no sign of leakage could be detected. For measurement of PCF, we used a simple pocket peak flow meter in favor of the PNT. PNT may ascertain the most accurate flow measurements, as has been argued by Chatwin and colleagues (10),

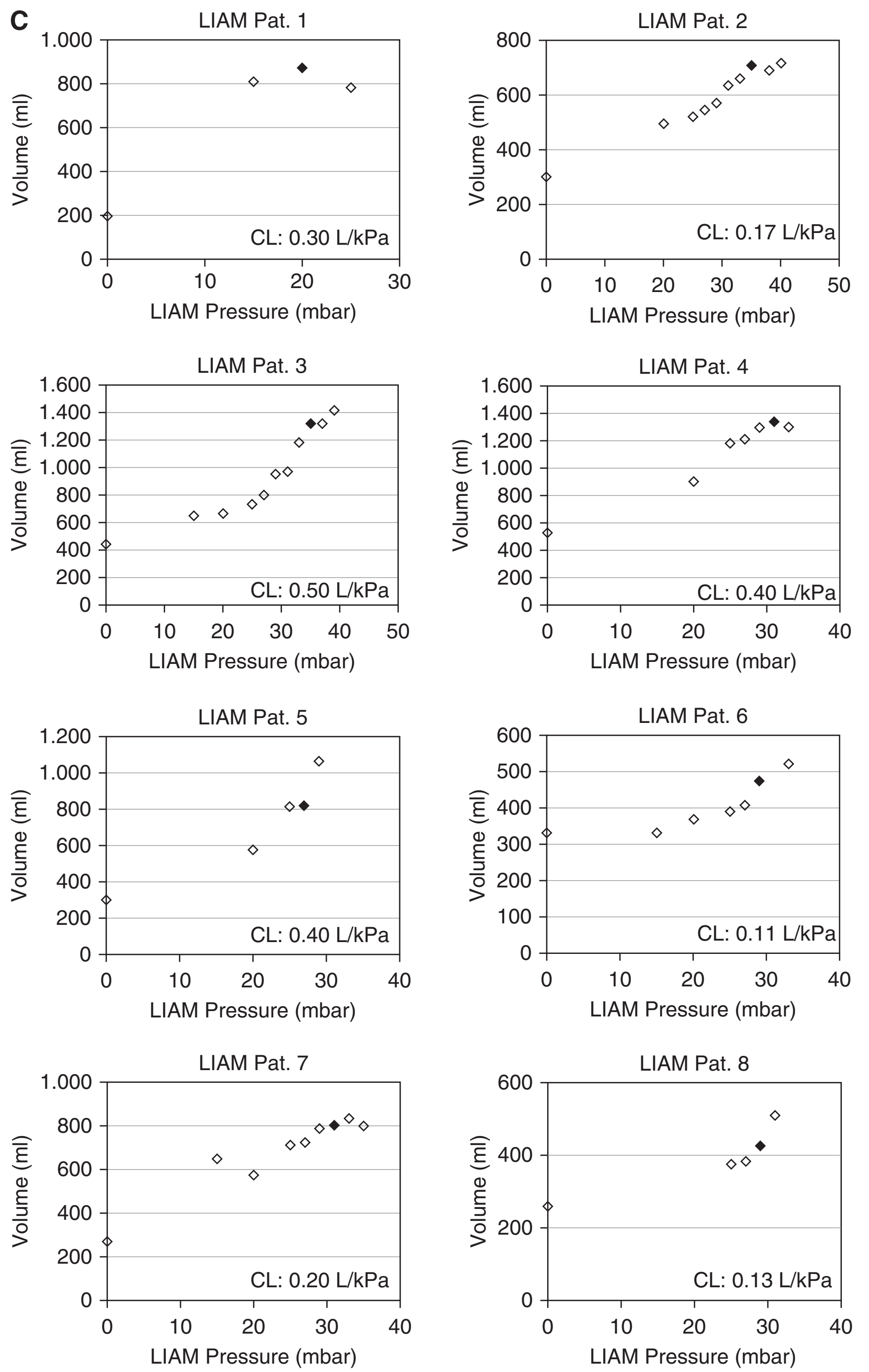

Figure 2. (Continued).

but we noted that a PNT adds unnatural flow resistance to coughing and thus leads to underestimation of the patient's cough competence. In fact, according to manufacturer specifications, the back pressure of the PNT we used is about 7 mbar at a flow of 160 L/min. A corresponding specification of the peak flow meter is not known but can be reasonably presumed to be minimal. Furthermore, Sancho and colleagues have shown that PCF can be measured reliably with a portable peak flow meter (23).

Discussion

Mechanical ventilation, mainly use as noninvasive ventilation, has become a standard therapy to efficiently address chronic hypoventilation in NMDs. However, even under effectively administered noninvasive ventilation, progressive weakness of respiratory muscles with shallow breathing and low pulmonary and chest wall excursions contribute to continuous deterioration of lung function. Furthermore, insufficient cough and impaired secretion clearance result in atelectasis and recurrent pulmonary infections. Accepted interventions to improve inspiratory capacity, expiratory flow, and cough are manual chest physiotherapy and techniques such as breath stacking, mechanical insufflation, and mechanical insufflation-exsufflation (MI-E) (12, 13, 15, 21, 24–27).

According to our daily clinical observation in patients with advanced NMD, cough can be sufficiently augmented by deep lung insufflation. We aimed to study the correlation between the pressures applied and the resulting IC and PCF in more detail. Given our clinical experience and concerns about using insufflation pressures above 40 mbar, we hypothesized that the best PCF could be achieved below a maximum lung insufflation and that we could identify an OIC.

Our results strongly support this hypothesis. We were able to show a clear relationship between insufflation pressure, IC, and cough flow with the best PCF after insufflation significantly below IC_{max}.

Twenty-nine of the enrolled 40 patients complied with the protocol and easily learned the assigned maneuver. Eleven patients were not able to hold their breath after insufflation and lost part of the insufflated volume during the shift to the peak flow meter. It is likely that some patients with bulbar weakness and glottic closure incompetence may not sufficiently increase PCF after deep lung insufflation and therefore may have to use other techniques.

Based on the titration curves plotted, we propose the following implications for respiratory system mechanics. When we assume (*1*) that flow has ceased at the end of each inspiration and (*2*)

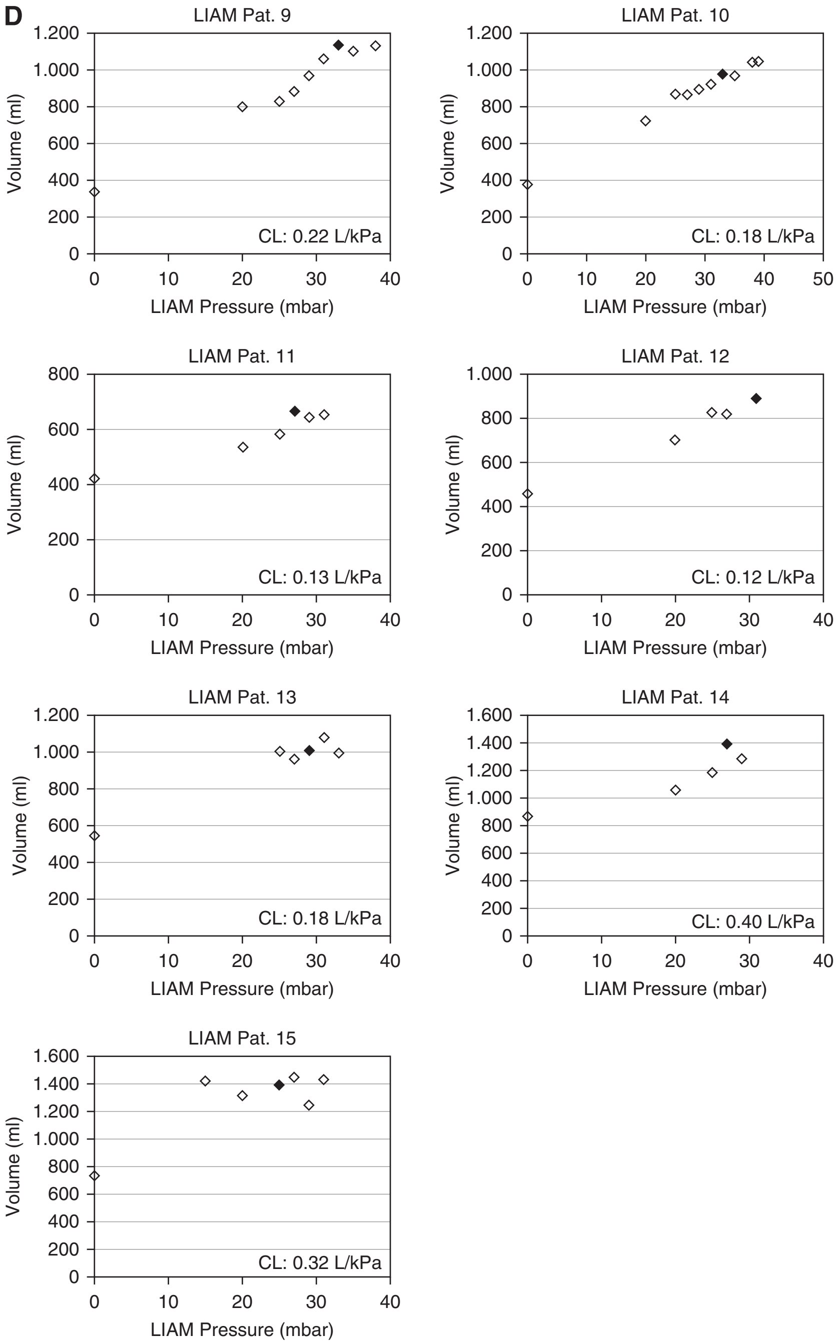

Figure 2. (Continued).

Table 1. Insufflation with intermittent positive pressure breathing to maximum insufflation capacity and optimum insufflation capacity with corresponding pressures (n = 14)

VC baseline (ml)	MIC (*ml*)	OIC (*ml*)	*P* Value
483 ± 276	1,060 ± 464	939 + 431	<0.01
	Pressure (*mbar*) 30.8 ± 5.2	Pressure (*mbar*) 26.8 + 5.3	<0.01

Definition of abbreviations: MIC = maximum insufflation capacity; OIC = optimum insufflation capacity.

that patients were passive during insufflations, we see static equilibrium between maneuver pressure and elastic recoil of the respiratory system. Zero flow at the maneuver end was a prerequisite for data validity because flow persistence could also indicate leakage. The second assumption is likely to be fulfilled at the end of a maneuver and, particularly, for volumes substantially above VC (Figures 2A–2C). The usage of esophageal pressure probes is regarded as standard for the measurement of muscle activity during mechanical ventilation. In our setting, we believe that any contribution of muscle work at maneuver end is very unlikely for the given reasons. Deriving compliance information from these titration curves may be regarded as a methodical weakness. Nevertheless, with this minor uncertainty we regard these data as an approximation to the compliance curve of the respiratory system. This assessment is strongly supported by the fact that the compliance we found in healthy control subjects with the same method is very similar to an accepted reference value of 1 L/kPa (28). In the majority of our patients and control subjects, we found a flattening in this pressure–volume relation in the region of 30 mbar, indicating that the lungs had been filled to the upper inflection point of the compliance curve.

The primary goal of deep lung insufflation is the generation of increased lung volume to improve subsequent cough. This approach has been repeatedly reported as successful, and Bach and colleagues (17, 25) and Kang and Bach (11) in particular have shown that air stacking with bag resuscitators or insufflation with the MI-E is effective for recruitment of lung volume and enhancement of PCF. Two publications on mechanical lung insufflation have influenced many clinicians. One work reported on nearly doubling lung volume by passive maximum insufflation in a group of 282 patients with VC <70% predicted and various NMDs where pressures between 40 and 70 mbar have been used (25). In addition, an inverse correlation between residual VC and assisted PCF has been demonstrated (10). We studied a small but homogenous group of adolescents

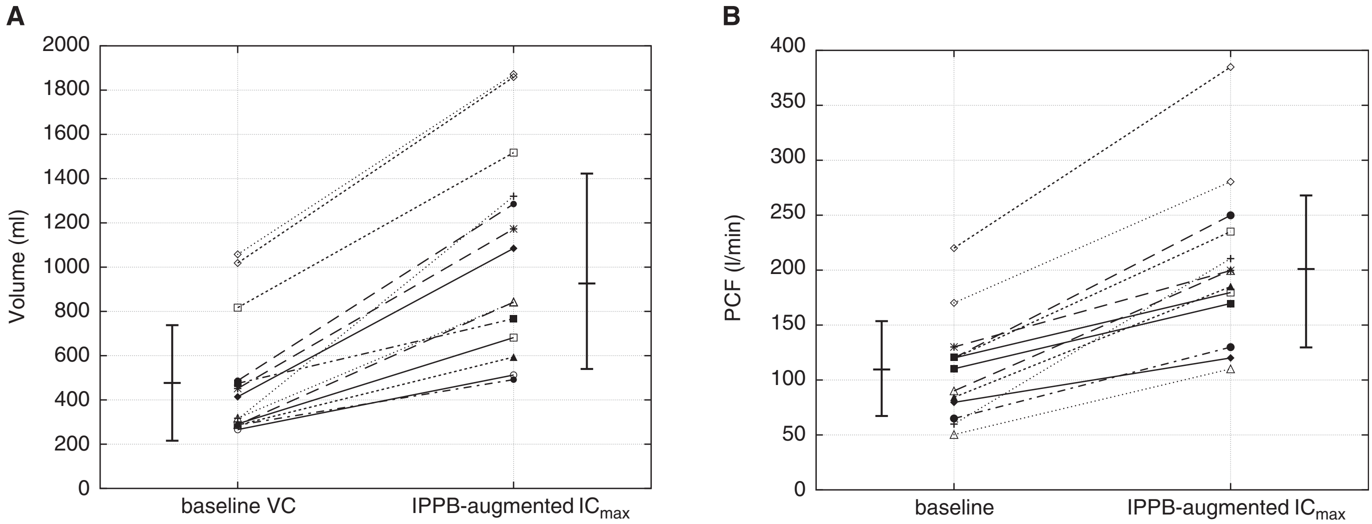

Figure 3. (*A*) Baseline vital capacity (VC) and IPPB-augmented maximum insufflation capacity (IC_{max}). (*B*) Baseline peak cough flow (PCF) and IPPB-augmented PCF.

and young adults with advanced respiratory failure and a mean VC below 500 ml. Slow insufflation using moderate pressures resulted in a mean IC_{max} over 1,000 ml. In contrast to other studies, where fixed and high pressures have been applied, IC_{max} was identified in our cohort by incrementally titrating the pressure until no further volume improvement was achieved or the patient could not tolerate a higher pressure. With this approach, IC_{max} could be achieved below 40 mbar at a mean pressure of about 30 mbar in many subjects even though those patients had severely reduced respiratory system compliance.

Furthermore, we found the highest individual PCFs after prior insufflation volumes significantly below IC_{max} (Figures 2A–2D) at significantly lower pressures (Tables 1 and 2). In Figures 2A through 2D we indicated the point with the highest PCF in the patients' individual volume-pressure curve, and, for the majority of our patients, this volume seemed to represent the upper inflection point of an estimated compliance curve. We describe the IC corresponding to the highest PCF as OIC. We hypothesize that an OIC achieved by slow insufflation with a pressure of approximately 30 mbar is an effective method to generate the highest PCF in patients with advanced respiratory restriction and cough insufficiency due to NMDs. In particular cases where patients may demand higher pressures, our strategy would be to increase the insufflation time to fully allow air distribution in the lung. One patient who participated in this study described his perception as "air getting deeper into the lung" when using LIAM with an insufflation time of 3 seconds instead of 1.5 seconds at the same pressure. Chatwin and colleagues have pointed out that the highest PCF is not necessarily associated with the highest maneuver pressures and have proposed mechanisms like vocal cord dysfunction or upper airway collapse during additional exsufflation in their study (29). With our findings, we conclude that severe lung or chest wall restriction is characterized by a mere downward displacement of the compliance curve without shift to the right, compared with healthy subjects. Consequently, we regard higher pressures beyond the upper inflection point as counterproductive or even hazardous. Insufflation for optimal PCF should instead target the upper inflection point. In our cohort, the patients' cough was severely impaired, with a mean baseline PCF of 110 L/min. In accordance with other studies, we also found a strong correlation (r = 0.8) between residual VC and unassisted PCF (3, 11). After titration of maneuver pressure to OIC, the PCF could be significantly and relevantly enhanced by 85% to a mean of 200 L/min. In the majority of patients, PCF could thus also be increased above 160 L/min, which marks a therapeutically relevant improvement (3). In a clinical setting, if PCF is deemed insufficient based on insufflation alone, a combination of manual abdominal compression or insufflation/exsufflation techniques may be alternative treatment options.

Table 2. Insufflation with lung insufflation assist maneuver to maximum insufflation capacity and optimum insufflation capacity with corresponding pressures (n = 15)

VC Baseline (*ml*)	MIC (*ml*)	OIC (*ml*)	*P* Value
425 ± 181	993 ± 313	906 ± 331	<0.01
	Pressure (*mbar*)	Pressure (*mbar*)	
	29.1 ± 5.7	26.3 + 6.9	<0.05

Definition of abbreviations: MIC = maximum insufflation capacity; OIC = optimum insufflation capacity.

We studied the impact of increasing inspiratory pressures on IC_{max}, OIC, and PCF using two continuous flow maneuvers, IPPB and LIAM. IPPB generally is used for insufflation with or without mechanical ventilation and has been subjected to repeated scientific investigation (12, 19, 21). LIAM has become available only in recent years as an integrated ventilator

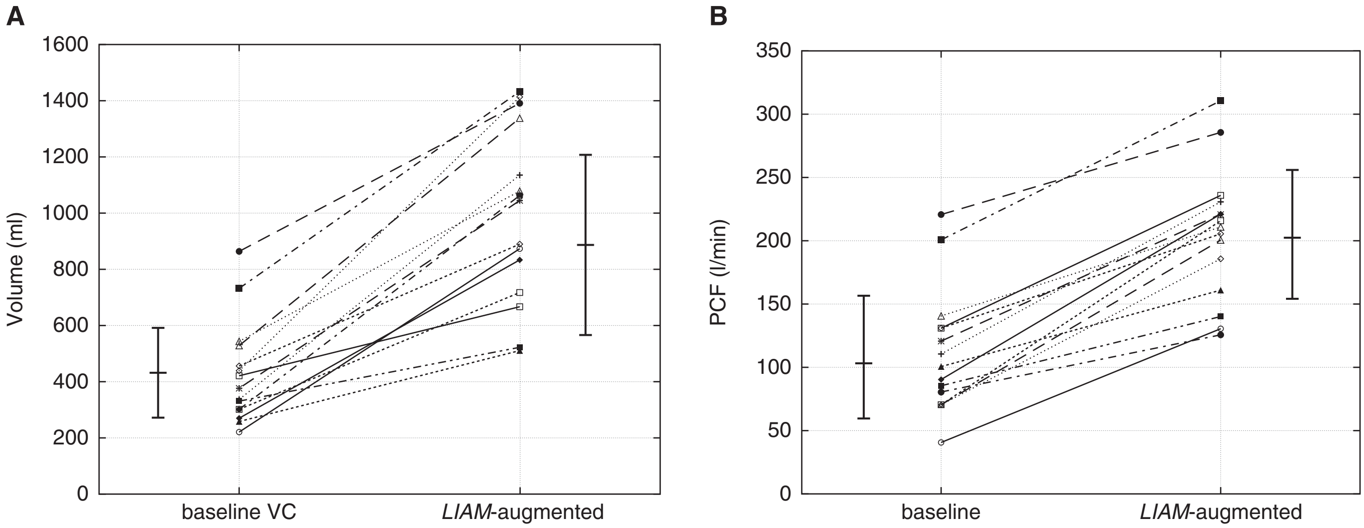

Figure 4. (*A*) Baseline vital capacity (VC) and LIAM-augmented maximum insufflation capacity (IC_{max}). (*B*) Baseline peak cough flow (PCF) and LIAM-augmented PCF.

function, and this is the first study that presents data on its effects on IC and PCF. Both techniques show some differences in their parameterization and pressurization rate, but both allow gentle flow or pressure profiles over several seconds, qualitatively shown in Figure 1. We found both methods similarly effective and comparable in patient comfort. For LIAM, slow inspiration is combined with an end-inspiratory pressure plateau, which facilitates synchronization of glottis closure with the maneuver.

Moderate insufflation pressures as used here may also limit discomfort and any potential remaining risk of pneumothorax. Pneumothorax has been documented in a few cases associated with mechanical ventilation (30), mechanical insufflation with resuscitation bag (31), and the use of MI-E (32). Through personal communication with our patients and colleagues, we also realized that concerns regarding pneumothorax as a potential complication of mechanical assisted coughing are quite common. Given these considerations, treatment may be introduced only hesitantly or might be administered at low and suboptimal pressures.

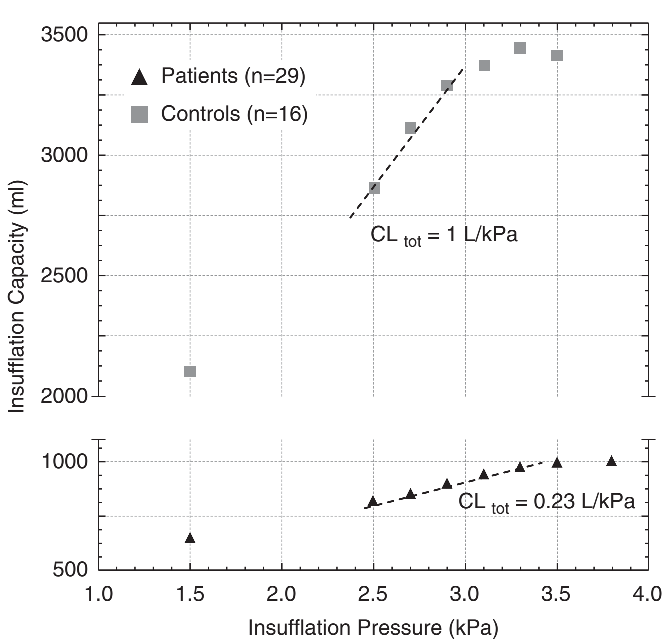

Figure 5. Estimated total compliance of chest wall and lungs (CL_{tot}) in patients and control subjects. Standard deviation (SD) in patients is 300–350 ml, SD in control subjects is 950–1,100 ml.

The methods used in this study are limited to insufflation without subsequent exsufflation. The combined MI-E, which has been studied especially in patients with NMDs (10, 27, 33, 34), is increasingly popular and is available in some new devices (35). However, the combination of insufflation and exsufflation does not inevitably result in higher PCF. Senent and colleagues reported on some patients with amyotrophic lateral sclerosis who did not achieve the best PCF based on insufflation/exsufflation but on insufflation alone (15). Chatwin and colleagues found an improvement in pediatric and adult patients with NMD from 169 ± 90 ml to 235 ± 111 ml with MI-E at pressures between +30 and −30 mbar (10), which is a lesser improvement than reported here. On the other hand, a PCF improvement of 250% at pressures between +40 and +60 mbar has been documented for postpoliomyelitis but for a different pathology (33). The authors of an ATS consensus statement acknowledged the clinical value of MI-E but also recommended further studies due to the limited data on this modality (24). A very recent Cochrane Review found that MI-E is effective in augmenting cough flow; however, MI-E was not found to be significantly better than other techniques (36).

Commentary

In patients with advanced NMDs, deep lung insufflation has become a mainstay in the management of insufficient cough. However, which pressures or volumes are safe and which parameters are required to achieve the highest individual PCF has not been evaluated. This study refines the current concept of lung insufflation to a maximum insufflation capacity for NMDs in children and young adults. Rather than targeting an IC_{max}, lung insufflation to the patient's individual OIC results in the best individual cough flow and allows limitation of pressures to approximately 30 mbar and therefore appears to be safe. ■

Author disclosures are available with the text of this article at www.atsjournals.org.

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