

ΕΝΩΣΗ ΠΝΕΥΜΟΝΟΛΟΓΩΝ ΕΛΛΑΔΑΣ

ετησιό σύνεσριό



30 Maΐou - 2 Ιουνίου 2019

Αθήνα, Ξενοδοχείο Royal Olympic

Παρασκευή 31 Μαΐου 2019

09.00-10.30 ΣΤΡΟΓΓΥΛΗ ΤΡΑΠΕΖΑ

ΜΗ ΕΠΕΜΒΑΤΙΚΟΣ ΜΗΧΑΝΙΚΟΣ ΑΕΡΙΣΜΟΣ (ΜΕΜΑ)

Προεδρείο: Α. Μαραδιά, Ι. Σιγάλα, Α. Δέρβας Βασικές αρχές ΜΕΜΑ *Ε. Δήμα* Ενδείξεις εφαρμογής ΜΕΜΑ στα ΤΕΠ Α. Κυριακούδη ΜΕΜΑ στο σπίτι Α. Βλάμη Παρακολούδηση ασδενών με κατ΄ οίκον ΜΕΜΑ: Τι πληροφορίες μου δίνει η κάρτα μνήμης των συσκευών *Ε. Περράκη*

ΜΕΜΑ στο σπίτι Α. Βλάμη

Βλάμη Αικατερίνη

Πνευμονολόγος

Επιμελήτρια Α' Β' ΠΑΝΕΠΙΣΤΗΜΙΑΚΗ ΠΝΕΥΜΟΝΟΛΟΓΙΚΗ ΚΛΙΝΙΚΗ

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Εργαστήριο Μελέτης Ύπνου Πανεπιστημιακό Γενικό Νοσοκομείο ΑΤΤΙΚΟΝ



RESOURCE DIRECTORY FOR VENTILATOR-ASSISTED LIVING

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- VENTILATOR EQUIPMENT AND AIDS, MANUFACTURERS' WEBSITES
- ORGANIZATIONS, ASSOCIATIONS AND FOUNDATIONS
- VENTILATOR CARE FACILITIES
- CONGREGATE HOMES FOR LONG-TERM VENTILATOR USERS Fuminiko Tasuma, MD (Muscular dystrophy)

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Home mechanical ventilation registration in Greece. Preliminary results from the Hellenic HMV Network

Eva serasli, Irini Lekka, Vasilis Kilintzis, Theodoros Moysiadis, Michalis Agrafiotis, Katerina Fekete, Dimitris Lagonidis, Athanasia Pataka, Kostas Porpodis, Vasiliki Paschidou, Paschalis Steiropoulos, Georgia Trakada, Georgia Chasapidou, Despina Ioannidou, Vassilis Koulouras, Dimitris Matamis, Christina Matei, Maria Sdougka, Elena Volakli, Nikos Maglaveras, Venetia Tsara *European Respiratory Journal 2017*

Abstract

Home Mechanical Ventilation (HMV) started in Greece during the 1990's, however systematic registration of patients has not been attempted previously. Preliminary results derived from the implementation of a HMV registration system, applied within the Hellenic HMV Network, are presented

Methods:

The HMV registration system is a flexible, on-line platform that supports recording of multi-visit patient data from multiple centers. Collected data is stored in a single data repository and can then be reviewed and exported in user defined datasets for statistical analysis. The system is based on the web server Apache, an embedded scripting language (PHP) and the open source database MySQL.

Results: The Hellenic HMV Network was launched in October 2016 with the primary aim of registering current practice. Currently, **125 patients from 11 centers are registered** with the following breakdown to underlying disease: **22.5% obstructive**, **32.4% restrictive**, **9.8% mixed (combined) lung disease**, **29.4% SAS.** Twenty-one % of patients are still smoking, 94.8% have clinical symptoms, 92.3% hypercapnia, 92.2% hypoxemia and **92.2% experience nocturnal** hypoventilation. NIV was prescribed in **76.9% and IV via tracheostomy in 5.1%.** The prescribed equipment was **BPAP S (42.7%), BPAP S/T (11.1%),** CPAP (25.6%) and **Home Ventilators (2.6%).**

Conclusion: These preliminary results indicate that NIV is the preferred type of HMV. Considering the prevailing economic crisis, there is a need to expand and continue the registration recordings with the view to better understand and eventually overcome the existing problems towards implementing effective medical interventions and sustainable health policies.



Patterns of home mechanical ventilation use in Europe: results from the Eurovent survey

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ABSTRACT: The study was designed to assess the patterns of use of home mechanical ventilation (HMV) for patients with chronic respiratory failure across Europe.

A detailed questionnaire of centre details, HMV user characteristics and equipment choices was sent to carefully identified HMV centres in 16 European countries.

A total of 483 centres treating 27,118 HMV users were identified. Of these, 329 centres completed surveys between July 2001 and June 2002, representing up to 21,526 HMV users and a response rate of between 62% and 79%. The estimated prevalence of HMV in Europe was 6.6 per 100.000 people. The variation in prevalence between countries was only partially related to the median year of starting HMV services. In addition, there were marked differences between countries in the relative proportions of lung and neuromuscular patients using HMV, and the use of tracheostomies in lung and neuromuscular HMV users. Lung users were linked to a HMV duration of <1 yr, thoracic cage users with 6–10 yrs of ventilation and neuromuscular users with a duration of ≥ 6 yrs.

In conclusion, wide variations exist in the patterns of home mechanical ventilation provision throughout Europe. Further work is needed to monitor its use and ensure equality of provision and access.



Home mechanical ventilation in Australia and New Zealand

Daniel J. Garner, David J. Berlowitz, James Douglas, Nick Harkness, Mark Howard, Nigel McArdle, Matthew T. Naughton, Alister Neill, Amanda Piper, Aeneas Yeo and Alan Young

ABSTRACT: This study aims to describe the pattern of home mechanical ventilation (HMV) usage in Australia and New Zealand.

34 centres providing HMV in the region were identified and asked to complete a questionnaire regarding centre demographics, patient diagnoses, HMV equipment and settings, staffing levels and methods employed to implement and follow-up therapy.

28 (82%) centres responded, providing data on 2,725 patients. The minimum prevalence of HMV usage was 9.9 patients per 100,000 population in Australia and 12.0 patients per 100,000 population in New Zealand. Variation existed across Australian states (range 4–13 patients per 100,000 population) correlating with population density (r=0.82; p<0.05). The commonest indications for treatment were obesity hypoventilation syndrome (OHS) (31%) and neuromuscular disease (NMD) (30%). OHS was more likely to be treated in New Zealand, in smaller, newer centres, whilst NMD was more likely to be treated in Australia, in larger, older centres. Chronic obstructive pulmonary disease was an uncommon indication (8.0%). No consensus on indications for commencing treatment was found.

In conclusion, the prevalence of HMV usage varies across Australia and New Zealand according to centre location, size and experience. These findings can assist HMV service planning locally and highlight trends in usage that may be relevant in other countries.

Long-Term Home Mechanical Ventilation in the United States

Angela C King RPFT RRT-NPS

Unfortunately, there are no comprehensive databases or national registry of home ventilator patients therefore the number of home ventilator patients is unknown. There are real challenges to providing mechanical ventilation in the home, which include caregiver training, adequacy of respiratory care, and reimbursement

Ideally, the preferred location for long-term mechanical ventilation is in the home, because costs are reduced (hospital costs= \$21,570, homecare costs= \$7,050, dollar saving per patient, per month \$14,520), quality of life is enhanced, and integration into the community is maximized

NNIV support in the home are increasing as technology and infrastructure support improves;however, reimbursement constraints make it challenging to provide home ventilator patients with the optimal equipment and services required

A central registry would allow for the development and monitoring of national home mechanical ventilator patient outcomes

Hindawi Publishing Corporation Canadian Respiratory Journal Volume 2016, Article ID 6547180, 10 pages http://dx.doi.org/10.1155/2016/6547180

Review Article

NMD: Neuromuscular disorders

HRQL: Health related quality of life

FCG: Family caregiver

VAI: Ventilator assisted individual

Clinical Outcomes Associated with Home Mechanical Ventilation: A Systematic Review

Erika J. MacIntyre,¹ Leyla Asadi,² Doug A. Mckim,³ and Sean M. Bagshaw^{1,2}

4.1. Summary of Major Findings. We found that HMV generally had a favorable impact on HRQL. Not surprisingly, improvement was more prominent and consistent for mental domains compared with physical domains across HRQL measures, particularly in those with NMD. We found HMV may be associated with an initially low and a subsequently reduced rate of hospitalization and days in hospital following implementation. While poorly described, FCG burden appears quite high. This likely relates in part to the financial strain associated with keeping VAIs in a home environment and may contribute to the unwillingness of some caregivers to choose HMV for their loved one again if given a second opportunity.

Indications for Home Noninvasive Ventilation

- Individual has chronic stable or slowly progressive respiratory failure, as evidenced by:
 - Daytime CO₂ retention ≥ 50 mm Hg, with appropriately compensated pH, or
 - Mild daytime or nocturnal CO₂ retention (45–50 mm Hg) with symptoms attributable to hypoventilation (eg, morning headaches, restless sleep, nightmares, enuresis, daytime hypersomnolence)

Nocturnal hypoventilation or oxygen desaturation

And the following conditions have been met:

- Individual has had optimal medical therapy for underlying respiratory disorders
- Individual is able to protect airway and clear secretions adequately
- Individual's reversible contributing factors have been treated (eg, obstructive sleep apnea, hypothyroidism, congestive heart failure, electrolyte disturbance)
- The diagnosis is appropriate and may include:
 - Neuromuscular disease
 - Chest wall deformity
 - Central hypoventilation or obesity hypoventilation
 - Obstructive sleep apnea, and a failure to improve with nasal CPAP
 - COPD with severe hypercapnia or nocturnal desaturation

Medical conditions for HMV

CNS disorders	
Arnold-Chiari malfor	mation
CNS trauma	
Cerebrovascular diso	rders
Congenital and acqui	ired central control of breathing disorders
Myelomeningocele	-
Neuromuscular disord	lers
ALS	
Congenital childhoo	od hypotonias
Guillain-Barré synd	rome
Infant botulism	
Muscular dystrophi	es
Myasthenia gravis	
Phrenic nerve paral	lysis
Polio and postpolio	sequelae
Spinal muscular atr	ophy
Myotonic dystrophy	1
Skeletal disorde	ers
Kyphoscolios	is
Thoracic wall	deformities
Thoracoplast	y.
Cardiovascular	disorders
Acquired hea	urt diseases
Congenital h	eart diseases

Respiratory disorders Upper airway Pierre-Robin syndrome Tracheomalacia Vocal cord paralysis Lower respiratory tract BPD COPD Complications of acute lung injury Cystic fibrosis Complications of infectious pneumonias Pulmonary fibrotic diseases







Ventilation centre

Ventilation centre

- Weaning centre (Recommendation according to the German Guideline "Prolonged Weaning") Main focus: Weaning intensive care unit
- weaning unit
- specialised ward for home mechanical ventilation



Home mechanical ventilation centre (new)

- Main focus: Home mechanical ventilation - (intensive care unit)
- specialised ward for invasive/non-invasive home mechanical ventilation



Noninvasive Ventilation









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Trigger

- The trigger variable defines how a mechanical breath is initiated. It can be initiated by:
- 1) **Pressure** (assisted breath pressure trigger initiated by the patient)
- 2) Flow (assisted breath flow-trigger initiated by the patient)
- 3) Volume (assisted breath volume-trigger initiated by the patient)
- 4) Combining Pressure+ Flow +Volume
- 5) Waveform algorithms (assisted breaths algorithms trigger initiated by the patient)
- 6) Time (timed mandatory breaths initiated by the ventilator)

"Assisted "breath the pressure applied to the respiratory system is generated by patient's respiratory muscles and by the ventilator. This makes it different from a "spontaneous" breath (eg CPAP =pressure is negative during the inspiratory phase and positive during the expiratory phase)

"Mandatory" breath defines a breath that is always initiated by the ventilator after a given time determined by the respiratory rate set by the operator and cycled by the ventilator

Trigger sensitivities (inspiratory and expiratory trigger) differ significantly amongst individual ventilation machines and may influence ventilation quality; this particularly pertains to synchronisation between the patient and the ventilator Patient-ventilator asynchrony (PVA)

- Significant proportion of patients are unable to adhere to their HMV prescription
- PVA describes the poor interaction between patient and ventilator
- Ramsay et al using parasternal electromyography during HMV set-up, found PVA is frequent and, in contrast to other studies not associated with an adverse impact on nocturnal gas exchange
- Controlled ventilation (Pressure and Volume Controlled) unload respiratory muscles most intensively, as the ventilator takes over the complete work of breathing BUT synchronicity of the patients' breathing rhythm with the ventilator may impact efficacy and tolerance.

Ventilation Interfaces

- Nasal masks offer greater patient comfort fewer pressure ulcers, but there is oral leakage during sleep, application of a chin band can be useful in individual cases
- Oronasal mask ameliorate oral leakage
- The use of oronasal masks in patients with obstructive sleep apnoea may lead to a deterioration in nocturnal sleep patterns
- When an oronasal mask or chin band cannot be tolerated, the use of humidification can lead to an improvement
- The use of oronasal masks compared to nasal masks has increased in parallel with inspiratory pressure application (studies in COPD)
- Customised masks can become necessary if high ventilation pressures and long ventilation times are applied, if the ready-made mask fits poorly, or if the patient has sensitive skin
- Mouth Piece Mask for Mouth Piece Ventialation in NMD (mouthpieces enable Intermittent Positive Pressure Ventilation (IPPV)

Comparison of the different types of masks	Commercial nasal masks	Commercial oronasal masks	Customised masks		
Dead space Oral leakage	Small Likely	Larger Unlikely	Minimal Type-dependent		
Pressure ulcer risk	Medium	Higher	Optimised	all and the	
Cost	Lower	Lower	Higher	Rec	514

Type-specific characteristics of different mask types for non-invasive ventilation.

Noninvasive Ventilation: What Is in the Box?

Ventilator-lung system in NIV



CPAP or APAP

Inhalation (breathing in)

CPAP blows constant pressure while you breathe in. Techn developed

Bi-Level PAP (BiPAP)

Inhalation (breathing in)

Bi-level blows higher pressure while you breathe in. Technologies for NIV were

developed earlier than AUTO CPAP

Exhalation (breathing out)





total and the second se

CPAP

- Improve oxygenation by increasing FRC and recruiting collapsed alveoli
- It provides certain positive airway pressure throughout all phases of spontaneous ventilation
- It is similar to breathing with your head stuck out of a moving car

♦ CPAP ≈ PEEP

BiPAP



Pressure Waveform CPAP

- Differential in pressure between inspiration and expiration allows for better patient-ventilator synchrony and thus more confort
- EPAP \approx CPAP \approx PEEP
- IPAP ≈ PS

0

- -Augments TV
- -Reduces Atelectasis
- -Reduces WOB

The EPAP is set to maintain upper airway patency and the IPAP/ EPAP difference provides PS to sustain/augment the patient's VT





Volume Ventilator More complicated to use Wide range of alarms Constant tidal volume Breath stacking possible No leak compensation Can be used without PEEP Rebreathing minimized Pressure Ventilator Simple to use Limited alarms Variable tidal volume Breath stacking not possible Leak compensation PEEP (expiratory positive airway pressure) always present Rebreathing possible



Device Trigger

← >60/X ---->

Time (sec)

Spontaneous Trigger

<60/X -+

Back up rate (BUR) = X

0

The device senses the patient's inspiratory effort and triggers IPAP in response to an increase in flow, and cycles into EPAP at the end of inspiration. In this mode, the breath will last as long as inspiratory flow is noted.

The breath rate and the respiratory pattern will be determined by the patient.

Clinical: applicable to any clinical scenario in which the proper minute ventilation can be sustained based on the patient's natural respiration rate

The device augments (provides PS) any breath initiated by the patient (spontaneous [S]), but will also apply to device-delivered breaths (timed [T] breath) should the patient's breath rate fall to less than the clinician's set "backup" rate

In contrast to S breaths, IPAP in T breaths will persist for the entire Ti time programmed as part of the device settings. This may cause the tidal volume to be reduced or uneven between S and T breaths. *This may vary by manufacturer.*

Clinical: in the setting of COPD, this uneven tidal volume may be beneficial by assisting ventilation without facilitating air trapping/

Clinical: in the setting of COPD, this uneven tidal volume may be beneficial by assisting ventilation without facilitating air trapping/ hyperinflation that may limit tolerance for NIV



The fixed breath rate and the fixed inspiration time set by the clinician are applied regardless of patient effort. Clinical: rarely used in clinical practice

Clinical: rarely used in clinical practice



The distinction between ST and PC mode centers on the programmed Ti Although the inspiration can be triggered spontaneously (S) or timed (T), there is no flow cycling out of IPAP. As opposed to ST, when Ti is applied only in timed breaths, in PC mode the programmed Ti time is applied to every breath. This will even out tidal volume on a breath-to-breath basis.

Clinical: a consistently applied fixed Ti is adventitious in the setting of NMD when diaphragmatic fatigue will drive otherwise short Ti time



- It is designed to maintain a preset target ventilation by monitoring ventilation, adjusting the PS, and providing a backup breath automatically
- VAPS technology is add-on software, which can be used with NIV in either ST or PC mode
- The settings include a range of PS values that are automatically adjusted to reach a targeted tidal volume. The EPAP is also programed using a range of settings to allow for automatic adjustments, targeting EPAP to prevent upper airway obstruction.
- Regarding backup rate, iVAPS "intelligent" backup shifts between twothirds of set rate during spontaneous breathing and set rate during period of apnea. In AVAPS devices, backup rate can be fixed or autoset, which is 2 bpm less than the average rate of the most recent six spontaneous breaths.

Clinical: not inferior to BPAP-ST in management of OHS, COPD and NMD.

Treating Chronic Hypoventilation With Automatic Adjustable Versus Fixed EPAP Intelligent Volume-Assured Positive Airway Pressure Support (iVAPS): A Randomized Controlled Trial SLEEP, Vol. 40, No. 10, 2017

Nigel McArdle, MD^{1,2}; Clare Rea BSc, BA²; Stuart King, BAppSc^{1,2}; Kathleen Maddison, PhD^{1,2}; Dinesh Ramanan, M Biomed Eng³; Sahisha Ketheeswaran, BBiomedSc³; Lisa Erikli, DAppSc (Nursing)³; Vanessa Baker, BHSc^{1,2}; Jeff Armitstead, PhD³; Glenn Richards, MB ChB³; Bhajan Singh, PhD^{1,2}; David Hillman, FANZCA^{1,2}; Peter Eastwood, PhD^{1,2}

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CONCLUSIONS

A night of AutoEPAP iVAPS in clinically stable patients on long-term NIV for chronic hypoventilation was noninferior to a night of FixedEPAP iVAPS for control of OSA (AHI). Other PSG sleep breathing and sleep quality and self-reported sleep quality outcomes were similar between the two iVAPS modes. Further studies are required to assess clinically relevant outcomes during treatment using the different modes both in NIVnaïve patients and over longer treatment durations.

Table 1. Patient's View of the Environment, ICU Versus Home

ICU	Home			
Noise	Relative quiet			
Light	Day/night cycles			
Limited view of the world	Outdoors easily visible and probably accessible			
Crowded/cramped	Relatively roomy			
Limited visitation allowed	Supportive visitors encouraged			
Immobilized	Mobility increased			
Sterile surroundings	Personal objects			
Little control	More independence			
Communication limited (or not a priority)	More time for speech development			
High reliance on technology	More reliance on family supervision			

Limited staff nurturing time Family nurturing

Trial design and size	RCT (n = 72) NIV (n = 72) control; analyzed on per protocol basis, not intention to treat	RCT (n = 102) NIV (n = 93) control	RCT (n = 101) NIV (n = 102) control
Primary end point	2-yr survival	1-yr survival	Time to readmission for respiratory cause, or survival at 1 yr
Inclusion criteria	GOLD ≥ 3 stable; Pa _{CO₂} > 46 mm Hg	GOLD = 4 stable; Pa _{CO₂} > 51.9 mm Hg	GOLD ≥ 3; > 48 h off ventilatory support for ARF; Pa _{CO₂} > 45 mm Ho
LTOT recipients, % IPAP/EPAP setting, mean, cmH ₂ O Back-up rate, mean (SD) ventilator mode	100 IPAP: 12.9 (95% CI 12.5-13.4); EPAP: 5.1 (95% CI 4.8-5.3) Nil, spontaneous mode	65 IPAP: 21 (SD 4.7); EPAP: 4.8 (SD 1.6) 16.1 (3.6), spontaneous/timed	77 IPAP: 19.2 (SD 3.4); EPAP: 4.8 (SD 1.0) 15 (3), spontaneous/timed
Adherence, mean (SD), h	4.5 (3.2) No difference from baseline at 6 and 12 mo	5.9 (3.1) Decreased in NIV group; change in baseline, -7.4% NIV, -2.4% control	 6.3 (2.4) Decreased in NIV group; change in Pa_{CO2}, -9.7 mm Hg NIV vs. 6 mm Hg control
12 mo	Improved on NIV, 80% NIV, 71% control	Improved on NIV, 88% NIV, 66% control	No difference in time to readmission or death: 30%
24 mo Other end points	68% NIV, 53% control	—	<u></u>
HRQOL	Reduced HRQOL in general and mental vigor domains in NIV group	Improvement in NIV group in disease-specific SGRQ and SBI summary score	Insignificant trend to improvement in HRQOL in NIV group (P = 0.054)
Sleep	Small but significant decrease in AHI and increase in REM sleep when receiving NIV	No detailed sleep architecture measurement	No detailed sleep architecture measurement
Comments	Stable patients	Stable patients; improvement in survival; significant reduction in Pace, achieved on NIV	Non-steady-state patients after exacerbation
	Improvement in survival	Slow recruitment	No improvement in survival or reduction of admissions; significant reduction of Pa _{CO2}
	No significant reduction in Pa _{CO2} on NIV	AnnalsATS Volur	ne 13 Number 11 November 2016



Kaplan-Meier survival curves in patients with obesity hypoventilation syndrome using noninvasive ventilation (NIV)





Clinical Investigations

Respiration

Respiration 2011;81:402-410 DOI: 10.1159/000317138 Received: October 26, 2009 Accepted after revision: March 30, 2010 Published online: June 26, 2010

Noninvasive Ventilation in Chronic Respiratory Failure: Effects on Quality of Life

Vassiliki Tsolaki Chaido Pastaka Konstantinos Kostikas



TABLE 3 Contraindications for noninvasive ventilation

```
Inability to protect the airway
  Inefficient cough
  Disordered swallowing with chronic aspiration
  Excess bronchial secretions
  Need for continuous or nearly continuous ventilation
Anatomical features that prevent appropriate mask
  placement
Lack of patient or family motivation
Inability of the patient to cooperate or understand
  the procedure
```

Side effect	After 1 month,	After 12 months,
	%	%
Dry throat	37	26
Facial pain	33	25
Fragmented sleep	27	20
Impaired nasal breathing	22	24
Abdominal bloating	22	13
Flatulence	19	17
Sleep impairment	13	16
Eye irritation	12	11
Nasal bleeding	7	2
Nausea	1	2
Facial pressure sores	1	0
Vomiting	0	0

Side effects of non-invasive long-term ventilation





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OBESITY HYPOVENTILATION



RESTRICTIVE CHEST DISORDER



NEUROMUSCULAR DISEASE



The respiratory system. The disorders of the respiratory systems, their implication on blood gases, and treatment





COPD, NMD, OHS, RESTRICTIVE DISORDERS with border diurnal normal blood values

An attended titration PSG or a review PSG in those initiated on NIV empirically is recommended to optimize therapy

OSA is common in patients with chronic hypoventilation disorders

SLEEP HYPONTILATION

DIURNAL CHRONIC HYPOVENTILATION

SPECIAL ARTICLES

јсѕм Journal of Clinical Sleep Medicine

Best Clinical Practices for the Sleep Center Adjustment of Noninvasive Positive Pressure Ventilation (NPPV) in Stable Chronic Alveolar Hypoventilation Syndromes

NPPV Titration Task Force of the American Academy of Sleep Medicine

Task Force Members: Richard B. Berry, M.D. (Chair)¹; Alejandro Chediak, M.D. (Vice-Chair)²; Lee K. Brown, M.D.³; Jonathan Finder, M.D.⁴; David Gozal, M.D.⁵; Conrad Iber, M.D.⁶; Clete A. Kushida, M.D., Ph.D.⁷; Timothy Morgenthaler, M.D.⁸; James A. Rowley, M.D.⁹; Sally L. Davidson-Ward, M.D.¹⁰

Recommendations for Limits of IPAP, EPAP, and PS Settings:

- 1. The recommended starting min IPAP=8cm H2O and recommended starting min EPAP=4cm H2O
- 2. The recommended maximum IPAP should be 30 cm H2O for patients ≥ 12 years and 20 cm H2O for patients < 12 years
- 3. The recommended minPS= 4cm H2O and maxPS=20 cm H2O
- 4. The min and max incremental changes in PS should be 1 and 2 cm H2O, respectively.

Recommendations for Adjustment of IPAP, EPAP, and PS:

1. The pressure support (PS) should be increased every 5 minutes if the tidal volume is < 6 to 8 mL/kg

- 2. The PS should be increased if the arterial PCO2>10 mm Hg above the PCO2 goal at the current settings for >10 min. An acceptable goal for PCO2 is a value less than or equal to the awake PCO2.
- 5. The PS may be increased if the SpO2<90%> 5 min and tidal volume is < 6 to 8 mL/kg

SPECIAL ARTICLES



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Recommendations for Use and Adjustment of the Backup Rate/Respiratory Rate:

- 1. A backup rate (i.e., ST mode) should be used in all patients with central hypoventilation, those with a significant number of central apneas or an inappropriately low respiratory rate, and those who unreliably trigger IPAP/EPAP cycles due to muscle weakness
- 3. The starting backup rate should be equal to or slightly less than the spontaneous sleeping respiratory rate (minimum of 10 bpm)
- 4. The backup rate should be increased in 1 to 2 bpm increments every 10 minutes if the desired goal of the backup rate has not been attained
- 5. The IPAP time (inspiratory time) should be set based on the respiratory rate to provide an inspiratory time (IPAP time) between 30% and 40% of the cycle time (60/respiratory rate in breaths per minute).



Table 2 Consensus clinical indicators for home NIV use in COPD

Disease documentation

Indications for usage



Before considering a COPD patient for NIV, a physician with skills and experience in NIV must establish and document an appropriate diagnosis on the basis of history, physical examination, and results of diagnostic tests, and assure optimal management of COPD with such treatments as bronchodilators, oxygen when indicated, and optimal management of other underlying disorders (such as performing a multi-channel sleep study to exclude associated sleep apnoea if clinically indicated).

The most common obstructive lung diseases would include chronic bronchitis, emphysema, bronchiectasis, and cystic fibrosis.

Symptoms (fatigue, dyspnoea, morning headache, *etc.*) AND Physiological criteria (one of the following): a) $Pa_{1}CO_{2} \ge 55$ mmHg. b) $Pa_{1}CO_{2} \ge 55$ mmHg and nocturnal desaturation (oxygen saturation by pulse oximeter $\le 88\%$ for 5 continuous minutes while receiving oxygen therapy ≥ 2 L per min). c) $Pa_{1}CO_{2}$ 50–54 mmHg and hospitalisation related to recurrent (≥ 2 in a 12-month period) epirespiratory failure. Breathe I March 2010



Comparison of selected trials of domiciliary non-invasive ventilation in COPD.

COPD Phenotype: Who and When?

	Trial Design	NIV settings (IPAP/EPAP Mode Back Up Rate)	Home NIV Titration strategy	Reduction in PaCO2 > 0.5 kPa in first 6 weeks	Primary outcome	Other reported effects
Casanova, 2000 ¹⁷	RCT	12cmH ₂ O/4cmH ₂ O S mode No back up rate	To reduce WOB	No	Mortality (No benefit)	Reduced admission rate
Clini, 2002 ¹⁰	RCT	14cmH ₂ O/2cmH ₂ O S/T mode BUR 8 bpm	To improve nocturnal SpO ₂	No	PaCO ₂ (No benefit)	Reduced self-reported dyspnoea
McEvoy, 2009 ¹¹	RCT	13cmH ₂ O/5cmH ₂ O S mode No back up rate	To achieve 3 hours sleep	No	Mortality <mark>(</mark> Equivocal)	Worse health related quality of life in the home NIV group
Windisch, 2005 ¹⁶	Cohort	28cmH ₂ O/0cmH ₂ O S/T mode BUR 20 bpm	To maximally tolerated by patient	Yes	Mortality (Benefit)	Non-randomised
Kohnlein, 2014 ⁹	RCT	22cmH ₂ O/5cmH ₂ O S/T mode BUR 16 bpm	20% reduction in PaCO ₂	Yes	Mortality (Benefit ARR 21%)	Stable COPD with low admission rate and preserved exercise capacity

Home NIV and long-term oxygen therapy (n = 102) LTOT alone (n = 93)



The ventilator settings required were lower compared to earlier reports for 'high intensity' NIV

Kohnlein et al Lancet Respir Med 2014



NIV High Intensity Ventilation delivered (IPAP=24cmH2O(22-26), EPAP=4cmH2O (4-5), backup rate=14bpm

P.B. Murphy, N. Hart / Arch Bronconeumol. 2017;xxx(xx):xxx-xxx



Kaplan-Meier plot of time from randomisation to first event (readmission or death) for 1 year follow up in HOT-HMV UK study

Conclussion: Assessment of COPD patients admitted with an episode of decompensated respiratory failure requiring acute NIV at 2-4 weeks following resolution of respiratory acidosis. If at 2-4 weeks, the patient has persistent hypercapnia then home NIV should be added to home oxygen therapy

Murphy PB, et al. JAMA. 2017

Delivery of NIV in COPD: How

(+)'High intensity' NIV= High inspiratory positive airways pressure and high back up rate IPAP 22 ± 5 cmH2O, EPAP 5 ± 2 cmH2O, 16 ± 4 bpm (Tolerability, Adherence, Expiratory Airflow Limitation)?

'High intensity' NIV needs slower acclimatisation (admission of greater than 5 days

(-)'High intensity' NIV = negative short term cardiovascular consequences with the long-term cardiovascular consequences largely unknown

High Inspiratory Pressure only without high back up rate has demonstrated similar benefit as 'high intensity NIV

 Table 2 Ventilator settings used in long-term randomized controlled trials on Home-NIV therapy for chronic hypercapnic COPD patients

Study (year)	Patients ^a	Mean IPAP/EPAP	Mode; mean backup	Interface	Compliance
			rate		
Casanova et al ¹¹	N=44	12/4 cm H ₂ O	Spontaneous mode; n/a	Nasal mask	6.2 hours/day at 3 and 6 months;
		_			5.9 hours/day after 12 months
Clini et al ¹²	N=86	14/2 cm H ₂ O	Spontaneous/timed mode; n/a	Nasal mask	9 hours/day
McEvoy et al ¹³	N=144	13/5 cm H ₂ O	n/a	Nasal or full-face mask, according to patient comfort	n/a
Duiverman et al ⁸	N=72	23/6 cm H ₂ O	Spontaneous/timed mode;	Nasal, oronasal	6.9 hours/night at 24 months
			18 breaths/min		
Köhnlein et al ⁷	N=195	22/5 cm H ₂ O	Controlled or assisted pressure support; 16 breaths/min	n/a	5.9 hours/day
Struik et al ⁵	N=201	19/5 cm H ₂ O	Spontaneous/timed mode;	Full-face mask (exception for	6.3 hours/night
			15 breaths/min	I patient with total face mask)	
Murphy et al ⁶	N=116	24/4 cm H ₂ O	Spontaneous/timed mode;	Nasal, oronasal, or total face mask,	4.7 hours/night at 6 weeks;
		-	14 breaths/min	according to patient's comfort	7.6 hours/night after 12 months
				Storre et al Internationa	al Journal of COPD 2018

COPD	COPD/OSA Overlap	Severe COPD	COPD Exacerbation	COPD "Plus"
Clinical presentation	Baseline PaO ₂ and Paco ₂ are normal PSG demonstrates sleep apnea Spirometry: mild to moderate obstruction	Chronic need for supplemental oxygen Chronic Paco ₂ > 52 mm Hg Would benefit from the use of NIV in both the ICU and at home	At baseline <i>may</i> not have hypercapnia or hypoxemia Is in the midst of an acute episode with high WOB/CO ₂ and hypoxemia, with need for inpatient NIV	COPD plus means that NIV may be more helpful for hospitalized patients with: COPD + MI COPD + HF COPD + CAP
Clinical risks	Pulmonary hypertension	Readmission mortality	Intubation risk	Worsening of underlying disease (eg, HF)
Device	CPAP therapy	Bilevel (S/ST) or VAPS Oxygen supplementation	Bilevel (S/ST) or VAPS Oxygen bleed in	Bilevel (ST/S) Oxygen bleed in

TABLE 4] Clinical Spectrum of COPD for NIV Intervention

Pathophysiology/ Device Settings	Chronic COPD (Compensated)		
Respiratory mechanics	 ↑ Muscle load (↑ Lower airway resistance in COPD) ↓ Muscle capacity (diaphragm atrophy, mechanical disadvantage) 	<mark>Ti (ms)</mark>	Short Ti or short Ti max to increase expiratory time and minimize iPEEP (I:E) Ti/Ttot 25% in patients with BMI > 30
		Cycle Sensitivity ^a	Default or high cycle sensitivity (early cycle) to provide a longer exhalation time (↓ I:E) Respironics: Auto-Trak or manual at 30%-50% of
Target volume (cc) Respiration To ad rate (bpm) IPAP (cm H ₂ O)	Target tidal volume 8 cc/kg ideal body weight just to goal minute ventilation based on ABGs or TcCO ₂ , or both High IPAP (or best tolerated)		peak flow ResMed: Cycle sensitivity medium to high
	BPAP: adjust IPAP to a PS for goal tidal volume (or best tolerated) Allow large IPAP max/IPAP min difference to reach target expiratory tidal volume or Va as tolerated		
<mark>EPAP (cm H</mark> ₂O)	Adjust to eliminate obstructive apneas if present If ineffective trigger, increase EPAP to overcome high iPEEP (first-line therapy)		
Trigger sensitivity ^a	Respironics: Auto-Trak or flow trigger 4-5 L/min ResMed: trigger medium		
<mark>Rise time (ms)</mark>	Fast rise time		

To adjust PS (BPAP-ST), expiratory tidal volume (AVAPS), or Va (iVAPS) based on ABG (pH, Paco2), TcCO2, or a combination

Table 7. Common thoracic-restrictive diseases with an indication for home mechanical ventilation

Kyphoscoliosis Kyphosis Pigeon chest Funnel chest Bechterew syndrome Restrictive pleural diseases Post-tuberculosis syndrome Post-traumatic thoracic deformities Post-operative thoracic deformities (thoracoplastic)



Indication for Non-Invasive Ventilation Exist in Thoracic-Restrictive Disease Patients

- The most important criterion for NIV is chronic hypercapnia (PaCO2 of ≥45 mm Hg)
- Normocapnia during the day with a rise in PtcCO2 of ≥10 mm Hg during the night
- VC<50% of that predicted, or with typical symptoms of hypercapnia, have shown that the development of daytime respiratory failure can be favourably influenced by early implementation of NIV when hypoventiation is exclusively nocturnal
- Patients without manifest hypercapnia, but with severe restrictive ventilatory dysfunction
- (VC < 50% of the predicted value), should undergo short-term (within 3 months) clinical monitoring including polygraphy

*

Significant nocturnal hypoventilation, first during the REM sleep phase, and also later during non-REM sleep phases, which can even worsen the patient's prognosis



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Effects of ventilation on thoracic restrictive diseases

- Improvements in gas exchange, quality of life, sleep profile, physical resilience, pulmonary haemodynamics (decresed pulmonary hypertension), maximal oxygen uptake, lung function, and inspiratory muscle strength
- Reduction in the rate of hopitalisation
- Elderly patients (> 75 years) can also benefit from NIV
- Reduction both in dyspnoea and daytime sleepiness as assessed by the Epworth sleepiness scale
- Respiratory muscle unloading, restoration of central chemosensitivity
- Improvement in pulmonary or thoracic compliance with a reduction in restrictive ventilatory dysfunction
- In comparison to patients receiving long-term oxygen therapy (LTOT), a clear survival advantage exists in patients undergoing NIV, especially in those with severe kyphoscoliosis or post-tuberculosis syndrome



OBESITY HYPOVENTILATION SYNDROME (OHS)

- OHS brings about an 8-fold increase in yearly costs
- OHS has 15% higher 5-year mortality and morbidity rate, even when NIV therapy is applied
- Untreated OHS leads to a 23% increase in mortality

DEFINITION OHS

-presence of obesity (BMI ≥30 kg/m2)

-chronic alveolar hypoventilation

-subsequent diurnal hypercapnia (PaCO2 ≥45 mm Hg) during normal tioai preating after ruling out other causes of hypoventilation

Right ventricular decompensation, pulmonary hypertension, polyglobulia

Coexistence OHS and OSAS in up to 90% of cases

Diagnostic Test: POLYSOMNOGRAPHY



OBESITY IS NOW A

GLOBAL EPIDEMIC!



Pathophysiology/ Device Settings	Chronic OHS (Compensated)		
Respiratory mechanics	spiratory hechanics		Respironics: Auto-Trak or flow trigger 2-3 L/min ResMed: trigger from medium to low
	 FRC due to obesity (expiratory flow limitation, airway closure, V/Q mismatch) Respiratory drive (leptin resistance, 10% OHS) 	<mark>Rise time (ms)</mark>	Default or slow rise time Respironics: 3 (300 ms)-6 (600 ms) ResMed: 500-900 ms
Target volume (cc)	Target tidal volume 8 cc/kg ideal body weight	Pathophysiology/ Device Settings	Chronic OHS (Compensated)
	To adjust PS (BPAP-ST), expiratory tidal v	Ti (ms)	Long Ti or long Ti min to maximize tidal
IPAP (cm H ₂ O)	High IPAP BPAP-ST: adjust IPAP to a PS for goal		volume and gas exchange by († I:E) Ti/Ttot 50%
	tidal volume (average PS, 8-10 cm H ₂ O) VAPS: allow a large IPAP max/IPAP min difference to reach target expiratory tidal volume or Va	<mark>Cycle</mark> Sensitivity ^a	Default or low cycle sensitivity Respironics: Auto-Trak or manual at 10%-15% of peak flow ResMed: Cycle medium to low
EPAP (cm H _z O)	High EPAP in OHS/OSA Adjust to eliminate obstructive apneas (average 8-12 cm H ₂ O) or snoring		
Respiration rate (bpm)	To adjust to goal minute ventilation based o	on ABGs or TcCO	2, or both
To adjust PS (BPAP-S	ST), expiratory tidal volume (AVAPS), or Va (iVAP	S) based on ABG	(pH, Paco ₂), TcCO ₂ , or a combination

	Spinal cord/neuropathic	Neuromuscular junction	Myopathic
Onset	disorders	disorders	disorders
Acute	 Guillain–Barre syndrome Cervical spinal cord injury Critical illness neuropathy Multiple sclerosis Transverse myelitis Epidural abscess Acute poliomyelitis Paralytic rabies 	 Myasthenia gravis Lambert-Eaton myasthenic syndrome Congenital myasthenic syndrome Botulism Venoms (snake, scorpions, ticks) Neuromuscular junction blockers Organophosphorus poisoning 	
Chronic	 Spinal cord injury Motor neuron disease Amyotrophic lateral sclerosis Spinal muscular atrophy Post-polio syndrome Chronic inflammatory demyelinating polyneuropathy Charcot–Marie–tooth disease 		 Muscular dystrophies Myotonic dystrophy Inflammatory myopathies Congenital and metabolic myopathies

Table 9.1 Neuromuscular diseases causing respiratory dysfunction

Indications NIV in NMD

- PaCO2 \geq 45 mmHg
- Nocturnal desaturation to ≤88% for≥ 5 consecutive minutes or nocturnal desaturation <90% for one cumulative minute
- Pimax <60 cmH2O or FVC < 50% predicted or FVC is < 70% and there is a reduction in FVC of > 10% in 3 months
- Peak Cough Flow >270L/MIN Elevated or rising serum bicarbonate level



Figure 2 Suggested pathway for investigating and managing patients with a neuromuscular disorder and respiratory insufficiency.

Goals of NIV in NMDs

1. Improve and maintain pulmonary compliance by reducing alveolar atelectasis

2. Optimize alveolar ventilation

Contraindications for NIV

1. Bulbar affection

- 2. Inability to protect airway
- 3. Severely impaired mental status (GCS of ≤8)
- 4. Inability to clear respiratory secretions with usual methods
- 5. Hypotension or shock
- 6. Massive upper gastrointestinal bleeding or actively vomiting
- 7. Multi-organ failure
- 8. Inability to fit mask

Tracheotomy is indicated for the following situations

Patient's wish and consent Inability to fit an appropriate ventilation interface NIV intolerance NIV inefficiency Severe bulbar symptoms with recurrent aspiration Inefficiency of non-invasive secretion management Failure to switch to NIV after intubation and invasive ventilation

Ο βαθμός του νυχτερινού αποκορεσμού συσχετίζεται με την μυική αδυναμία του διαφράγματος

Sawtooth desaturation

- 1. In patients without bulbar affection, non-invasive ventilation (NIV) can be used as an alternative to ventilation, usually via tracheostomy.
- With the increased awareness of <u>nocturnal hypoventilation in patients with</u> <u>NMDs</u>, use of <u>nocturnal non-invasive ventilation (NIV)</u> has gained impetus.
- Hypoventilation from poor effort and resultant lung restriction (reduced compliance), inability to effectively clear respiratory secretions, and failure of upper or lower airway patency are the ma Key Points of respiratory failure in neuromuscular disorders.
- While dyspnea/tachypnea, exhaustion, diaphoresis, and the inability to speak in complete sentences are all obvious manifestations of respiratory failure, in someone with suspected NMD, such overt signs may not be always present.
- An elevated or rising serum bicarbonate level may be an early sign of undiagnosed nocturnal hypercapnia and has been predictive of mortality in patients with ALS.
- When <u>NIV</u> is judiciously <u>combined with cough assist devices (mechanical and</u> other modalities to clear airway secretions), the number and duration of hospitalizations is reduced.
- Concomitant chronic obstructive pulmonary disease (COPD) and/or congestive heart failure may be another indication for successful use of NIV in neuromuscular disorders.

Pathophysiology/ Device Settings	Chronic NMD (Compensated)		
Respiratory mechanics	↓ Muscle capacity ↑ Chest wall resistance	<mark>Ti (ms)</mark>	Long Ti or long Ti min to maximize tidal volume and gas exchange (†I:E) Ti/Ttot 50%
		<mark>Cycle</mark> Sensitivity ^a	Default or low cycle sensitivity (late cycle) to provide a longer inhalation time (maximize tidal volume and gas exchange by high I:E)
Target volume	Target tidal volume 8 cc/kg ideal body weight		Respironics: Auto-Trak or manual at 10%-15% of peak
(CC) Respira	tion To adjust to goal minute ventilation based on ABGs or	TcCO ₂ , or both	flow
rate (bpm)		ResMed: Cvcle low
o adjust PS (BPAP-ST	Adjust IPAP to a PS for tidal volume goal in BPAP-ST. (average PS, 6 cm H ₂ O) Allow IPAP min at a higher baseline), expiratory tidal volume (AVAPS), or Va (iVAPS) base	d on ABG (pH	I, Paco _z), TcCO _z , or a combination
EPAP (cm H _z O)	Low EPAP to reduce work of breathing and improve triggering		
Trigger sensitivity ^a	High trigger sensitivity to support a weak respiratory muscular effort Respironics: flow trigger at 1-3 L/min ResMed: trigger high or very high		
<mark>Rise time (ms)</mark>	Default or slow rise time Respironics: 3 (300 ms)-6 (600 ms) ResMed: 500-900 ms		

Benefits of NIV in NMD

- Improvement in blood gases
- – Resetting of the respiratory centre with an increase in
- the hypercapnic-ventilatory response
- – Increase in pulmonary compliance
- – Improved sleep quality
- Improvement in health-related quality of life
- – Regression of hypoventilation symptoms
- – Improvement of depression
- – Reduction in the rate of rehospitalisation
- – Prolonged survival
- – Improvement in neurocognition

Improvement of the survival by approximately 7 months in ALS with orthopnea or daytime hypercapnia

Figure 1 Survival in hypercapnic patients with Duchenne muscular dystrophy using nasal intermittent positive pressure ventilation.

The medium age of death in Duchenne's muscle dystrophy has ameliorated from 18-20 years to nearly 30 years under establishment of NIV

Role of Non-Invasive Ventilation in Patients with Pulmonary Fibrosis

- The insecure data situation does not currently allow the formulation of a general recommendation for NIV in pulmonary fibrosis patients
- Poor prognosis in IPF, there are no long-term data in relation to NIV in this disease group
- NIV can be considered here for palliative care purposes
- NIV initiation can exist in cases of long-term interstitial lung diseases
- (e.g., before a scheduled lung transplantation) and possibly in the context of rehabilitation (benefit needs to be examined on a case by case basis)

PAP/DEVICES CRISIS

In conclusion, patients who had heart failure with a reduced ejection fraction and predominantly central sleep apnea, the addition of adaptive servo-ventilation to guideline-based medical treatment did not improve the outcome. The risk of cardiovascular death was increased by 34%, which was sustained through-out the trial, and there was no beneficial effect on quality of life or symptoms of heart failure.

 JOURNAL of MEDICINE

 ESTABLISHED IN 1812
 SEPTEMBER 17, 2015
 VOL. 373
 NO. 12

Adaptive Servo-Ventilation for Central Sleep Apnea in Systolic Heart Failure

CONCLUSSIONS

- In clinical practice, non-invasive pressure support ventilation has become the most favorite therapeutical approach
- Assisted ventilation allows the patient to spontaneously trigger pressure or volume support, which may improve synchronicity and may damage muscle fibers in less extent
- While patients with neuromuscular or thoraco-skeletal diseases can often be treated sufficiently with low tidal volumes, high pressure support may be needed in patients with COPD
- In both NMD, COPD the treatment target is normalization of carbon dioxide so that the term "high pressure ventilation" is misleading
- High intensity pressure in COPD is associated with better compliance (mean difference of 3.6 h/d) and was superior in terms of controlling nocturnal hypoventilation
- Volume-targeted algorithms may be favorable in NMD. These patients may suffer from impaired coughing causing mucoid bronchus obliteration so that pressure-targeted systems may not guarantee minimum minute ventilation
- There are no differences between automated volume assured pressure ventilation (AVAPS) and fixed-level pressure support in super obese patients (BMI 50±7 kg/m2) and COPD
- The addition of NIV has proven to improve quality of sleep, nocturnal oxygen saturation, diurnal and nocturnal PaCO2, survival and quality of life in a broad spectrum of various chronic hypoventilation disorders

NIV the art of science

The success of NIV in a given patient population depends on selection of an appropriate patient, selection of an appropriate interface, selection of an appropriate ventilator and ventilator settings, the skills of the clinician, the motivation of the patient, and the support of the family

ΕΝΩΣΗ ΠΝΕΥΜΟΝΟΛΟΓΩΝ ΕΛΛΑΔΑΣ

ετήσιο σύνεδριο

30 Μαΐου - 2 Ιουνίου 2019 Αθήνα, Ξενοδοχείο Royal Olympic 09.00-10.30 ΣΤΡΟΓΓΥΛΗ ΤΡΑΠΕΖΑ ΜΗ ΕΠΕΜΒΑΤΙΚΟΣ ΜΗΧΑΝΙΚΟΣ ΑΕΡΙΣΜΟΣ (ΜΕΜΑ) Προεδρείο: Α. Μαραδιά, Ι. Σιγάλα, Α. Δέρβας Βασικές αρχές ΜΕΜΑ Ε. Δήμα Ενδείξεις εφαρμογής ΜΕΜΑ στα ΤΕΠ Α. Κυριακούδη ΜΕΜΑ στο σπίτι Α. Βλάμη Παρακολούδηση ασδενών με κατ' οίκον ΜΕΜΑ: Τι πληροφορίες μου δίνει η κάρτα μνήμης των συσκευών Ε. Περράκη

Ευχαριστώ για την Προσοχή σας