

## Appendix F. Results from the Included Studies

KQ1. What are the patient characteristics and/or laboratory criteria and/or target level measurable improvements considered for the initiation and continuation of noninvasive positive pressure ventilation supplied by a Home Mechanical Ventilator (HMV), Bilevel Positive Airway Pressure device (BPAP), and Continuous Positive Airway Pressure device (CPAP)?

**Table F.1. COPD - New initiation of home device**

Author, Year, Study Design	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
Murphy, 2017 <sup>46</sup> RCT	BPAP ST	-COPD (FEV1 < 50%) -NIPPV during hospital admission	-PaCO <sub>2</sub> >53 mmHg -PaO <sub>2</sub> <55 mmHg or PaO <sub>2</sub> < 60 mmHg with polycythemia, pulmonary hypertension or cor pulmonale -ST 90<30% -pH >7.30 (daytime, room air)	"High pressure ventilation strategy" titrated during polysomnography
Oscroft, 2014 <sup>50</sup> RCT	BPAP IVAPS versus BPAP ST	-COPD (FEV1 < 50%) -Mixed stable disease or following AECOPD	-PaCO <sub>2</sub> >7 kPa (53 mmHg) -pH >7.35 or PtcCO <sub>2</sub> >9 kPa (68 mmHg) (daytime)	BPAP IVAPS: Target minute ventilation and target back up respiratory rates were the mean minute ventilation and rates that the patients had during a one hour trial of pressure support ventilation at 15 cmH <sub>2</sub> O while awake. The device then attempted to reproduce target minute ventilation overnight by automatically adjusting the inspiratory pressures in the range 7-25 cmH <sub>2</sub> O. (Titration took on average 3.3 [SD 1.6] days)  BPAP ST: IPAP and backup rate were adjusted to optimize ventilation with the aim of reducing PtcCO <sub>2</sub> . EPAP set at 5cmH <sub>2</sub> O. (Titration took on average 5.2 [SD 2.8] days)
Paone, 2014 <sup>51</sup> Observational	BPAP ST	-COPD (FEV1 < 50%) -NIPPV during hospital admission	-PaCO <sub>2</sub> > 50 mmHg (after awakening from a night without NIPPV)	Maximum tolerated IPAP to target tidal volume of 6 mL/kg (measured body weight). EPAP set at 2-8 cmH <sub>2</sub> O. Backup rate set

Author, Year, Study Design	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
				at 12 breaths/min.
Galli, 2014 <sup>29</sup> Observational	BPAP NOS	-COPD (ICD-9) -NIPPV during hospital admission	-PaCO <sub>2</sub> > 45 mmHg	
Bhatt, 2013 <sup>4</sup> RCT	BPAP NOS	-COPD (FEV1 NOS) -Stable (no AECOPD in prior 4 weeks)	-PaCO <sub>2</sub> <52 mmHg	IPAP set at 15 cmH <sub>2</sub> O. EPAP set at 5 cmH <sub>2</sub> O. Initiation performed in home by respiratory therapist over 1 week.
Duiverman <sup>22, 23</sup> , 2011 RCT	BPAP ST	-COPD (FEV1 <50%) -Stable (no AECOPD in prior 4 weeks)	-PaCO <sub>2</sub> >6.0 kPa (45 mmHg) -pH >7.35 (daytime, room air)	Maximum tolerated IPAP to target PaCO <sub>2</sub> <6.0 kPa and PaO <sub>2</sub> > 8.0 kPa.
Oscroft, 2010 <sup>48</sup> Observational	BPAP ST started in AECOPD	-COPD (FEV1 <50%) -NIPPV during hospital admission for AECOPD	-PaCO <sub>2</sub> >7.5 kPa (56 mmHg) -pH 7.35-7.45 (daytime) or -PaCO <sub>2</sub> >6.5 kPa (49 mmHg) -pH 7.35-7.45 + PtcCO <sub>2</sub> >9 kPa (68 mmHg) (daytime)	
	BPAP ST started in stable COPD	-COPD (FEV1 <50%) -Stable (no current AECOPD)	-PaCO <sub>2</sub> >7.5 kPa (56 mmHg) -pH 7.35-7.45 (daytime) or -PaCO <sub>2</sub> >6.5 kPa (49 mmHg) -pH 7.35-7.45 + PtcCO <sub>2</sub> >9 kPa (68 mmHg) (daytime)	
Cheung, 2010 <sup>12</sup> RCT	CPAP versus BPAP ST	-NIPPV during hospital admission for AECOPD	-PaCO <sub>2</sub> > 6 kPa (45 mmHg) -pH <7.35	CPAP: CPAP set at 5 cmH <sub>2</sub> O  BPAP ST: Maximum tolerated IPAP (range 10 to 20 cmH <sub>2</sub> O) to target tidal volume 7-10 mL/kg. EPAP set at 5 cmH <sub>2</sub> O. Backup rate set at 14 breaths/min.

Author, Year, Study Design	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
Casanova, 2000 <sup>10</sup> RCT	BPAP S	-COPD (FEV1 <45%) -Stable (no AECOPD in prior 3 months)		Maximum tolerated IPAP ( $\geq 8$ cmH <sub>2</sub> O above EPAP) to target 20% decrease in respiratory rate and visible decrease in accessory muscle use and dyspnea. EPAP set at 4 cmH <sub>2</sub> O. (Titrated in hospital for 1 week).
Garrod, 2000 <sup>30</sup> RCT	BPAP S	-COPD (FEV1 <50%) -Stable (no AECOPD in prior 4 weeks) -exercise intolerance due to dyspnea		Maximum tolerated IPAP and EPAP. (Titrated over 1 week).
Clini, 1998 <sup>15</sup> Observational	BPAP ST	-COPD (FEV1 <50%) -Stable (no AECOPD in prior 4 weeks) -LTOT $\geq 12$ months - $\geq 1$ ICU admission due to AECOPD in prior 2 years	-PaCO <sub>2</sub> >6 kPa (45 mmHg) -pH >7.35 -PaO <sub>2</sub> <8 kPa (60 mmHg) (daytime, room air, rest)	Minimal IPAP to achieve an expiratory tidal volume > 8ml/kg. EPAP was set in order not to overcome the intrinsic PEEP. Backup rate set at 10 breaths/min.
Clini, 1996 <sup>14</sup> Observational	BPAP ST	-COPD (FEV1 30-49%) -LTOT $\geq 18$ months - $\geq 1$ hospital admission due to AECOPD in prior 18 months	-PaCO <sub>2</sub> >6.7 kPa (50 mmHg)	Minimal IPAP to achieve an expiratory tidal volume > 8ml/kg. Rate set at 10 breaths/min (Titration over 15 days in hospital).
Zhou, 2017 <sup>70</sup> RCT	BPAP ST	-COPD (FEV1 <50%) -Stable (no AECOPD in prior 4 weeks)	-Hypercapnia (daytime, rest) NOS	Maximum tolerated IPAP ( $\geq 10$ cmH <sub>2</sub> O). EPAP set at 4 cmH <sub>2</sub> O. Backup rate set at 16 breaths/min.
Marquez-Martin, 2014 <sup>38</sup> RCT	BPAP ST	-COPD (FEV1 <50%) -Stable (no AECOPD in prior 3 months)	-PaCO <sub>2</sub> > 45 mmHg -PaO <sub>2</sub> < 60 mmHg	Maximum tolerated IPAP (10-20 cmH <sub>2</sub> O) to target good clinical response and SaO <sub>2</sub> . EPAP set at 4 cmH <sub>2</sub> O. Backup rate set at 12 breaths/min.
Köhnlein, 2014 <sup>37</sup> RCT	BPAP ST	-COPD (FEV1 <30%) -Stable (no AECOPD in prior 4 weeks)	-PaCO <sub>2</sub> $\geq 7$ kPa (53 mmHg) -pH $\geq 7.35$ (daytime, rest)	Targeted to reduce baseline PaCO <sub>2</sub> by $\geq 20\%$ or achieve PaCO <sub>2</sub> <6.5 kPa (49 mmHg).
De Backer, 2011 <sup>19</sup> RCT	BPAP NOS	-COPD (FEV1 <50%) -AECOPD requiring hospitalization	-PaCO <sub>2</sub> >45 mmHg on day 5-12 of hospitalization	Targeted SaO <sub>2</sub> >90% during 90% of time and reduction in PaCO <sub>2</sub> $\geq 5\%$ in 1 hour.

Author, Year, Study Design	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
Dreher, 2010 <sup>21</sup> RCT	HMV (pressure assist/control) versus HMV (PSV ST)	-COPD (Gold stage IV) -Stable (no current AECOPD).	-PaCO <sub>2</sub> >45 mmHg (daytime) and PaCO <sub>2</sub> >50 mmHg (nocturnal)	HMV (pressure assist/control): Maximum tolerated IPAP to target maximum reduction in PaCO <sub>2</sub> (normocapnia if possible). EPAP set to avoid dynamic hyperinflation (3-6 cmH <sub>2</sub> O). I:E ratio set at 1:2 and modified per patient tolerance. Inspiratory flow trigger set to 3 l/min.  HMV (PSV ST): IPAP set to 14-16 mbar. Backup rate set to 8 breaths/minute. Inspiratory flow trigger set to 3 l/min. Expiratory trigger set to 70% of maximal inspiratory flow.
McEvoy, 2009 <sup>43</sup> RCT	BPAP S	-COPD (FEV <sub>1</sub> <50% or <1.5L) -Stable disease -LTOT for ≥3 months	-PaCO <sub>2</sub> >46 mmHg (at least twice in prior 6 months during stability)	Maximum tolerated IPAP-EPAP difference (≥5 cmH <sub>2</sub> O). EPAP set at 3 cmH <sub>2</sub> O and titrated up to target reduction of snoring and obstructive hypopneas/apneas in polysomnogram. (Titration performed in elective hospital admission for 3-4 days.)
Tsolaki, 2008 <sup>65</sup> Observational	BPAP ST	-COPD (FEV <sub>1</sub> <50%) -Stable (no AECOPD in prior 4 weeks)	-PaCO <sub>2</sub> >50 mmHg -PaO <sub>2</sub> <60 mmHg (room air)	IPAP and EPAP to target patient comfort, decreased accessory muscle use, lower respiratory rate, and decrease in PaCO <sub>2</sub> >5% after 1 hour. (Titration in hospital).
Windisch, 2006 <sup>69</sup> Observational	HMV with pressure controlled ventilation (PCV) mode	-COPD NOS -Stable (no worsening symptoms in prior 2 weeks, respiratory rate <30 breaths/minute, no signs of current respiratory infection, no changes in symptoms or medications in prior 3 months) -NIPPV in hospital admission	-pH≥7.35	Maximum tolerated IPAP to target a maximum decrease in PaCO <sub>2</sub>
Gay, 1996 <sup>31</sup> RCT	BPAP ST versus sham CPAP lowest setting	-COPD (FEV <sub>1</sub> < 40%) -Stable disease	-PaCO <sub>2</sub> > 45 mmHg (daytime, rest)	IPAP set to 10 cmH <sub>2</sub> O. EPAP set to lowest possible. Backup rate to target patient comfort.
Gad, 2014 <sup>28</sup> Observational	BPAP ST	-COPD (FEV <sub>1</sub> < 50%) -Stable (no AECOPD in prior 4 weeks) PaCO <sub>2</sub> >50 mmHg	-PaCO <sub>2</sub> >50 mmHg -pH > 7.35 (daytime)	Maximum tolerated IPAP (targeting 15-20 cmH <sub>2</sub> O). EPAP 3-6 cmH <sub>2</sub> O. (Titration occurred in hospital over 2-3 day period.)
Sin, 2007 <sup>62</sup> RCT	BPAP NOS versus sham	-COPD (FEV <sub>1</sub> NOS) -Stable disease		Maximum tolerated IPAP (maximum of 20 cmH <sub>2</sub> O). EPAP set at 4 cmH <sub>2</sub> O.

Author, Year, Study Design	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
	CPAP 4 cmH <sub>2</sub> O			
Heinemann, 2011 <sup>34</sup> Observational	BPAP (pressure controlled ventilation)	-COPD (FEV1 NOS) -invasive mechanical ventilation for AECOPD, pneumonia, or postoperative respiratory failure -prolonged weaning from invasive mechanical ventilation	-PaCO <sub>2</sub> >52.5mmHg or -pH<7.35 (recurrent acidosis)	
Budweiser, 2007 <sup>8</sup> Observational	BPAP (pressure controlled ventilation)	-COPD (FEV1 <50%) -Stable and unstable disease	-PaCO <sub>2</sub> >55mmHg -pH<7.35 (recurrent acidosis)	Maximum tolerated IPAP to achieve maximum reduction in PaCO <sub>2</sub> .
Clini, 2002 <sup>16</sup> RCT	BPAP ST	-COPD (FEV1 NOS) -Stable (no AECOPD in prior 4 weeks)	-PaCO <sub>2</sub> >6.6 kPa (50 mmHg) -pH>7.35 (daytime, room air)	Maximum tolerated IPAP with goal decrease in PaCO <sub>2</sub> >5% after 1 hour; and nocturnal SaO <sub>2</sub> ≥90% for 90% of time. (Titration in hospital).
Struik, 2014 <sup>64</sup> RCT	BPAP ST	-COPD (FEV1 <50%) -NIPPV or invasive mechanical ventilation in hospital admission	-PaCO <sub>2</sub> >6 kPa (45 mmHg)	Maximum tolerated IPAP to achieve normal PaCO <sub>2</sub> . Respiratory rate was set to match respiratory rate of patient, I:E set to 1:3 with a short rise time and then titrated on comfort.
Duroo, 2018 <sup>25</sup> Observational	HMV/BPAP mix	-COPD (NOS) -AECOPD		Maximum tolerated IPAP to achieve maximum reduction in PaCO <sub>2</sub> . Backup respiratory rate was increased above resting respiratory rate if persistent hypercapnia. Pressure support ventilation was switched to pressure controlled ventilation if persistent hypercapnia. Volume assured pressure assisted/controlled ventilation was used if prolonged ventilation (>12 hours/day) or intolerant to IPAP >25 cmH <sub>2</sub> O)
	HMV/BPAP mix	-COPD (NOS) -Stable (no current AECOPD)		
Duiverman, 2017 <sup>24</sup> RCT	HMV /BPAP mix (pressure controlled ventilation versus pressure support ventilation)	-COPD (FEV1 NOS) -Stable (no AECOPD in prior 4 weeks) -≥ 2 AECOPD with acute hypercapnic respiratory failure (pH<7.35) per year	-PaCO <sub>2</sub> ≥6.7 kPa (50 mmHg) (daytime) or -PaCO <sub>2</sub> ≥7.3 kPa (55 mmHg) (nighttime) or -Nighttime rise in PtCO <sub>2</sub> ≥1.3 kPa (10 mmHg)	Pressure controlled ventilation: Maximum tolerated IPAP to achieve maximum reduction in PaCO <sub>2</sub> . Backup rate set just above spontaneous breathing frequency. EPAP set at 4-6cm H <sub>2</sub> O.  Pressure support ventilation: Maximum tolerated IPAP, with maximum IPAP of 18 cmH <sub>2</sub> O and maximum backup rate of 14

Author, Year, Study Design	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
Blankenburg, 2017 <sup>5</sup> Observational	HMV (pressure controlled ventilation or pressure support ventilation)	-COPD (FEV1 NOS) -Stable (no AECOPD in prior 2 weeks)	-PaCO <sub>2</sub> >7.0kPa (53mmHg) -pH>7.35	breaths/minute. Goal of titration was normal PaCO <sub>2</sub> as well as patient tolerability of NIPPV. Titration started in pressure controlled ventilation mode. If pressure controlled ventilation was not achievable, pressure support ventilation was used. Inspiratory pressure was set to relieve “air hunger” on inspiration or to reach a tidal volume ≥800mL. PEEP was increased to maximally tolerated. Respiratory rate was set at 2 breaths/minute above the spontaneous respiratory rate.
Tsolaki, 2011 <sup>66</sup> , Observational	BPAP ST	-COPD (FEV1 NOS) -Stable (no AECOPD in prior 4 weeks) -Symptoms of nocturnal hypoventilation (fatigue, dyspnea, morning headaches)	-PaCO <sub>2</sub> ≥50mmHg -pH>7.35	Patients were hospitalized for 2–3 days during the initial application of NIV, in order to ensure maximal compliance. The modality of NIV was pressure support ventilation delivered by a variable positive airway pressure device (VPAP III ST, ResMed, Sydney, N.S.W., Australia), set in the spontaneous/timed mode with a backup respiratory rate of 8–14 breaths / min. Inspiratory and expiratory positive airway pressures (IPAP and EPAP, respectively) were adjusted according to the patient’s comfort and synchrony with the ventilator and a marked reduction in the use of accessory muscles. For the first 2 h from the initiation of ventilation the patient was observed by a pulmonologist and a fully trained nurse, in order to assure a decrease in accessory respiratory muscle use and respiratory rate, check the patient’s comfort of mask ventilation and possible air leaks of the system. Supplemental oxygen was added as needed in order to maintain oxygen saturation between 88 and 92%. Arterial blood gases were measured 1 h after the initiation of ventilation. A 5% decrease in Pa CO <sub>2</sub> values was considered as adequate ventilatory support.

AECOPD: acute exacerbation of chronic obstructive pulmonary disease, BPAP: Bilevel Positive Airway Pressure, cmH2O: centimeters of water (pressure), COPD; chronic obstructive pulmonary disease, CPAP: Continuous Positive Airway Pressure, EPAP: expiratory positive airway pressure, FEV1: Forced expiratory volume in one second, HMV: Home Mechanical Ventilation, ICU: Intensive Care Unit, IPAP: inspiratory positive airway pressure, IVAPS: intelligent volume assured pressure support, kPa: kilopascal, LTOT: long term oxygen therapy, mmHg: millimeters of mercury (pressure), NIPPV: Noninvasive positive pressure Ventilation, NOS: Not otherwise Specified, PaO2: partial pressure of arterial oxygen, PaCO2: partial pressure of arterial carbon dioxide, PEEP: positive end expiratory pressure, pH: potential of hydrogen, PSV: Pressure support ventilation, RCT: randomized controlled trial, S: spontaneous mode, SaO2: arterial blood oxygen saturation, ST: spontaneous/timed breath mode

**Table F.2. COPD - Established home device use**

Author, Year, Study Design	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
Salturk, 2015 <sup>57</sup> Observational	BPAP ST	-COPD (FEV1 not specified) -post ICU admission -home NIPPV ≥ 4 hours/day		IPAP titrated to achieve “desired tidal volume” (maximum 30 mbar)
Hitzl, 2009 <sup>35</sup> Observational	HMV (pressure cycled assist control mode)	-Stable (no current AECOPD) -HMV initiated ≥3 months		
Funk, 2010 <sup>27</sup> RCT	BPAP NOS for 6 months	-COPD "standard criteria" NOS -AECOPD requiring NIPPV or invasive ventilation -chronic nocturnal NIPPV use at home for ≥ 6 months	-PaCO2 > 45 mmHg (stable, measured immediately after awakening from a night without mechanical ventilation)	Maximum tolerated IPAP (10-20 cmH2O). EPAP set to 5 cmH2O. Inspiratory time was limited to a maximum of 1.3 s to avoid leak-induced prolongation of inspiration.
	BPAP NOS more than 6 months			
Vasquez, 2017 <sup>67</sup> cohort	BPAP NOS versus CPAP NOS versus HMV NOS	-COPD (ICD-9)		
Oscroft, 2010 <sup>49</sup> RCT	BPAP ST (pressure controlled ventilation)	-COPD, FEV1<50%, FEV1/FVC<70%, TLC>80%, >20 pack year smoking history -Stable (no current AECOPD: no increased breathlessness, cough or sputum in the prior 4 weeks, no increase in PaCO2 and no decrease in FEV1 since study initiation) -Chronic nocturnal NIPPV use at home for ≥ 3 months	-pH 7.35-7.45 -PaCO2>7.5 kPa or PtcCo2>9kPa	
	No PAP			

AECOPD: acute exacerbation of chronic obstructive pulmonary disease, BPAP: Bilevel Positive Airway Pressure, cmH2O: centimeters of water (pressure), COPD; chronic obstructive pulmonary disease, EPAP: expiratory positive airway pressure, FEV1: Forced expiratory volume in one second, HMV: Home Mechanical Ventilation, ICU: Intensive Care Unit, IPAP: inspiratory positive airway pressure, mmHg: millimeters of mercury (pressure), NIPPV: Noninvasive positive pressure Ventilation, NOS: Not otherwise Specified, RCT: randomized controlled trial, ST: spontaneous/timed breath mode

**Table F.3. Thoracic Restrictive Disorders - New initiation of home device**

Author, Year, Study Design	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
Salturk, 2015 <sup>57</sup> Observational	BPAP ST	Kyphoscoliosis NOS		IPAP titrated to achieve “desired tidal volume” (maximum 30 mbar)
Domenéech-Clar, 2003 <sup>20</sup> Observational	BPAP NOS	-Kyphoscoliosis or fibrothorax or thoracoplasty -Stable (no infection in past 3 months) -Symptoms of hypercapnia (fatigue, dyspnea, morning headache)	-PaCO2 >45 mmHg or -FVC <40% or -MIP <60 cm H2O or -nocturnal SaO2 < 88% for ≥ 5 consecutive minutes	IPAP increased (minimum 10 cmH2O) to target a normal PaCO2 or a decrease of at least 10 mmHg. (Titration occurred in hospital)
Nauffal, 2002 <sup>47</sup> Observational	BPAP NOS	-Kyphoscoliosis NOS -stable (no infection in past 3 months) -symptoms (fatigue, dyspnea, morning headache)	-PaCO2 >45 mmHg or -FVC <50% or -MIP <60 cm H2O or -SaO2 < 88% for ≥ 5 consecutive minutes by nocturnal oximetry	IPAP and EPAP titrated to maximize change of arterial blood gases. (Titration occurred in hospital)
Masa, 2000 <sup>39</sup> Observational	HMV (volume controlled ventilation with change to pressure controlled ventilation if volume could not be tolerated)	-Kyphoscoliosis (scoliosis angle [Cobb] >90 degrees -FEV1/FVC ≥65% -Apnea-hypopnea index ≤ 20 events/hour	-PaCO2 >47 mmHg for at least 3 months	Ventilator parameters adjusted to target maximum reduction in PaCO2 as well as patient tolerance, air leakage, and nocturnal saturation >90%. Patient initially treated with volume-cycled ventilator. Patients with poor compliance to volume-cycled ventilator were switched to a bilevel pressure ventilator. (Titration occurred in hospital over 3-7 days)
Schonhofer, 2001 <sup>61</sup> Observational	HMV (volume controlled ventilation with change to BPAP ST if volume could not be tolerated)	-TRD (post-TB or scoliosis NOS) -Stable disease (stable PaCO2, no hospital admission in prior 1 month)	-Absence of severe acidosis -PaCO2 45-55 mmHg	



Author, Year, Study Design	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
Tsolaki, 2011 <sup>66</sup> Observational	BPAP ST	-TRD NOS -Stable (no acute exacerbation in prior 4 weeks) -Symptoms of nocturnal hypoventilation (fatigue, dyspnea, morning headaches)	-PaCO <sub>2</sub> ≥45mmHg -pH>7.35	Patients were hospitalized for 2–3 days during the initial application of NIV, in order to ensure maximal compliance. The modality of NIV was pressure support ventilation delivered by a variable positive airway pressure device (VPAP III ST, ResMed, Sydney, N.S.W., Australia), set in the spontaneous/timed mode with a backup respiratory rate of 8–14 breaths / min. Inspiratory and expiratory positive airway pressures (IPAP and EPAP, respectively) were adjusted according to the patient's comfort and synchrony with the ventilator and a marked reduction in the use of accessory muscles. For the first 2 h from the initiation of ventilation the patient was observed by a pulmonologist and a fully trained nurse, in order to assure a decrease in accessory respiratory muscle use and respiratory rate, check the patient's comfort of mask ventilation and possible air leaks of the system. Supplemental oxygen was added as needed in order to maintain oxygen saturation between 88 and 92%. Arterial blood gases were measured 1 h after the initiation of ventilation. A 5% decrease in Pa CO <sub>2</sub> values was considered as adequate ventilatory support.

BPAP: Bilevel Positive Airway Pressure, cmH<sub>2</sub>O: centimeters of water (pressure), EPAP: expiratory positive airway pressure, FEV<sub>1</sub>: Forced expiratory volume in one second, FVC: Forced vital capacity, HMV: Home Mechanical Ventilation, IPAP: inspiratory positive airway pressure, MIP: maximal static inspiratory pressure, mmHg: millimeters of mercury (pressure), NOS: Not otherwise Specified, PaCO<sub>2</sub>: partial pressure of arterial carbon dioxide, ST: spontaneous/timed breath mode, TB: tuberculosis, TRD: Thoracic Restrictive Disorder

**Table F.4. Thoracic Restrictive Disorders – Established home device use**

Author, Year, Study Design	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
Hitzl, 2009 <sup>35</sup> Observational	HMV (pressure cycled assist control mode) in restrictive thoracic disease	-TRD NOS -HMV initiated ≥3 months		
Buyse, 2003 <sup>9</sup> Observational	HMV (volume or pressure cycled ventilator NOS) + oxygen	-Kyphoscoliosis NOS -NIPPV use NOS		

HMV: Home Mechanical Ventilation, NIPPV: Noninvasive positive pressure Ventilation, NOS: Not otherwise Specified, TRD: Thoracic Restrictive Disorder

**Table F.5. Neuromuscular Disease - New initiation of home device**

Author, Year, Study Design	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
Sanjuan-López, 2014 <sup>60</sup> Observational	HMV (PSV or ST) started after outpatient pulmonary evaluation	-ALS (El Escorial criteria) -hospital admission -chronic respiratory failure by pulmonologist		Increase in IPAP to target symptom relief. Monitored with daytime and nocturnal oximetry and blood gases.
	HMV (PSV or ST) started in an emergency situation without prior outpatient pulmonary evaluation			
Pinto, 1995 <sup>53</sup> Observational	BPAP NOS	-ALS (El Escorial criteria) -bulbar features		
Doménech-Clar, 2003 <sup>20</sup> Observational	BPAP NOS	-NMD NOS -stable (no infection in past 3 months) -symptoms (fatigue, dyspnea, morning headache)	-PaCO <sub>2</sub> >45 mmHg or -FVC <50% or -MIP <60 cm H <sub>2</sub> O or -nocturnal SaO <sub>2</sub> < 88% for ≥ 5 consecutive minutes	IPAP increased (minimum 10 cmH <sub>2</sub> O) to target a normal PaCO <sub>2</sub> or a decrease of at least 10 mmHg. (Titration occurred in hospital)
Nauffal, 2002 <sup>47</sup> Observational	BPAP NOS	-NMD NOS -stable (no infection in past 3 months) -symptoms (fatigue, dyspnea, morning headache)	-PaCO <sub>2</sub> >45 mmHg or -FVC <50% or -MIP <60 cm H <sub>2</sub> O or -SaO <sub>2</sub> < 88% for ≥ 5 consecutive minutes by nocturnal oximetry	IPAP and EPAP titrated to maximize change of arterial blood gases. (Titration occurred in hospital)
Sancho, 2014 <sup>58</sup> Observational	HMV (volume cycled) versus BPAP ST	-ALS NOS -symptoms (fatigue, dyspnea, orthopnea, morning headache)	-PaCO <sub>2</sub> >45 mmHg or -FVC <50% or -MIP <60 cm H <sub>2</sub> O or -SaO <sub>2</sub> < 88% for ≥ 5 consecutive minutes by nocturnal oximetry	Titration occurred in the hospital.

<b>Author, Year, Study Design</b>	<b>Device/mode</b>	<b>Patient characteristics to start or continue device</b>	<b>Laboratory characteristics to start or continue device</b>	<b>Device titration</b>
Sivori, 2007 <sup>63</sup> Observational	BPAP NOS	-ALS (El Escorial criteria) -symptomatic ventilatory impairment (dyspnea, morning headache, fatigue)	-PaCO <sub>2</sub> >45 mmHg or -FVC <50% or -MIP <60 cm H <sub>2</sub> O or -nocturnal SaO <sub>2</sub> < 88% for ≥ 5 consecutive minutes	IPAP adjusted to maintain SpO <sub>2</sub> >92% (ranged 13-25 cmH <sub>2</sub> O). EPAP set from 5-9 cmH <sub>2</sub> O.
Coco, 2006 <sup>17</sup> Observational	BPAP ST	-ALS (El Escorial criteria) -symptomatic ventilatory impairment (dyspnea, morning headache, fatigue)	-PaCO <sub>2</sub> >45 mmHg or -FVC <50% or -MIP <60 cm H <sub>2</sub> O or -nocturnal SaO <sub>2</sub> < 88% for ≥ 5 consecutive minutes	Maximum tolerated IPAP and EPAP to target patient comfort, leaks, normal PaO <sub>2</sub> , PaCO <sub>2</sub> , SpO <sub>2</sub> , and symptom relief. IPAP started at 8-12 cmH <sub>2</sub> O and EPAP started at 3-4 cmH <sub>2</sub> O.
Bourke, 2006 <sup>7</sup> RCT	BPAP ST	-ALS NOS	-Orthopnea with Pimax <60% or -symptomatic daytime hypercapnia	IPAP and EPAP adjusted to optimize daytime arterial blood gases, nocturnal oximetry breathing room air, and increased use/duration of device.
Vitacca, 2017 <sup>68</sup> Observational	HMV/BPAP mix started in FVC ≥ 80% (early)	-ALS NOS -FVC ≥ 80%		Pressures adjusted to patient comfort, normalization of PaCO <sub>2</sub> , optimize nocturnal oximetry/polysomnography, and improve compliance. Backup rate set at 12 breaths/min. Preset tidal volume set at 5 ml/kg.
	HMV/BPAP mix started in FVC < 80% (late)	-ALS NOS -FVC < 80%		
Sancho, 2017 <sup>59</sup> Observational	HMV (volume assist control ventilation)	-ALS (El Escorial criteria) -symptomatic ventilatory impairment (dyspnea, orthopnea, fatigue, morning headache, daytime hypersomnolence, decreased cognitive function)	-PaCO <sub>2</sub> >45 mmHg and -FVC <50% and -nocturnal SaO <sub>2</sub> < 90% for ≥ 5% of time	Ventilator adjusted to target PaCO <sub>2</sub> < 45 mmHg, nocturnal SaO <sub>2</sub> < 90% for < 5% of time, optimize comfort, prevent air leaks.
Bertella, 2017 <sup>3</sup> RCT	BPAP volume assured pressure support ventilation	-ALS (definite via El Escorial Criteria) -Stable disease (no respiratory infection in prior 3 months)	-PaCO <sub>2</sub> > 45 mmHg, MIP < 70% predicted, subjective respiratory discomfort in any position, FVC < 70% predicted, or 20% decline in MIP or FVC over 3 months	Tidal volume was set, but to unclear settings. Respiratory rate set at 12 breaths/minute. IPAP set to maximal patient comfort. EPAP set to relieve obstructive events on polysomnogram. Settings adjusted to achieve maximal reduction in PaCO <sub>2</sub> . Device titrated in patient versus outpatient according to randomization.
Tsolaki, 2011 <sup>66</sup>	BPAP ST	-NMD NOS	-PaCO <sub>2</sub> ≥ 45 mmHg	Patients were hospitalized for 2–3 days during the initial

Author, Year, Study Design	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
Observational		<p>-Stable (no acute exacerbation in prior 4 weeks)</p> <p>-Symptoms of nocturnal hypoventilation (fatigue, dyspnea, morning headaches)</p>	-pH>7.35	<p>application of NIV, in order to ensure maximal compliance. The modality of NIV was pressure support ventilation delivered by a variable positive airway pressure device (VPAP III ST, ResMed, Sydney, N.S.W., Australia), set in the spontaneous/timed mode with a backup respiratory rate of 8–14 breaths / min. Inspiratory and expiratory positive airway pressures (IPAP and EPAP, respectively) were adjusted according to the patient's comfort and synchrony with the ventilator and a marked reduction in the use of accessory muscles. For the first 2 h from the initiation of ventilation the patient was observed by a pulmonologist and a fully trained nurse, in order to assure a decrease in accessory respiratory muscle use and respiratory rate, check the patient's comfort of mask ventilation and possible air leaks of the system. Supplemental oxygen was added as needed in order to maintain oxygen saturation between 88 and 92%. Arterial blood gases were measured 1 h after the initiation of ventilation. A 5% decrease in Pa CO<sub>2</sub> values was considered as adequate ventilatory support.</p>
Aboussouan, 1997 <sup>1</sup> Observational	HMV/BPAP mix	<p>-ALS via el Escorial criteria</p> <p>-dyspnea on exertion or orthopnea or FVC &lt; 60% predicted.</p>	- PaCO <sub>2</sub> ≥ 45 mmHg	<p>The devices used were a volume-controlled ventilator (PLV-100, Life Care Products, Lafayette, Colorado) in assist-control mode or a bilevel positive-pressure device (BiPAP, Respironics, Inc., Murrysville, Pennsylvania) in spontaneous-timed mode (the latter was added as an option after September 1994). Patients were ventilated in the supine position while in clinic. Tidal volume (for the volume-controlled ventilator) or pressure (for the bilevel positive-pressure device) were initially adjusted for chest rise, leaks, and patient comfort and were adjusted on subsequent visits to control hypercapnia and dyspnea. The ultimate choice of a device was made by the patient after the two devices had been sampled. Patients were instructed to use noninvasive positivepressure positivepressure ventilation nightly as tolerated and as necessary in the daytime. On subsequent visits, alternate interfaces were used for mask-related problems, nasal steroid sprays were used for nasal congestion, and suction machines or mechanical insufflation- exsufflation were used for clearance of secretions. Tolerance was defined as the ability to sleep nightly while receiving noninvasive</p>

Author, Year, Study Design	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
				positive-pressure ventilation for at least 4 consecutive hours.
Farrero, 2005 <sup>26</sup> Observational	HMV/BPAP mix	-ALS NOS -Symptoms (orthopnea) -FVC $\leq$ 50% predicted or a decrease in FVC of $\geq$ 500 mL on two consecutive visits	-Desaturations in nocturnal pulse oximetry (arterial oxygen saturation, $<$ 90% during 5 consecutive min)  Or  PaCO <sub>2</sub> $>$ 45 mm Hg	A volume ventilator (LIFECARE PLV100; Respironics; Murrysville, PA) was used in all cases of invasive ventilation, whereas either a volume ventilator (LIFECARE PLV100; Respironics; and PV 501; BREAS Medical; Gothenburg, Sweden) or a bilevel pressure ventilator (BiPAP; Respironics; and Sullivan VPAP ST II; ResMed Ltd; Abingdon, UK) was used for NIV. Interfaces included nasal masks (customized or commercial) with a chinstrap (to minimize oral leaks), mouthpiece, or facemask. The choice of ventilator and interface was based on the adaptation of the patient and the number of hours of ventilation required. Treatment with HMV was initiated during a hospital admission, and ventilation parameters were adjusted to achieve comfort as well as adequate ventilation according to daytime arterial blood gas levels and nocturnal oximetry measurements.

ALS: amyotrophic lateral sclerosis, BPAP: Bilevel Positive Airway Pressure, cmH<sub>2</sub>O: centimeter of water (pressure), EPAP: expiratory positive airway pressure, FVC: Forced vital capacity, HMV: Home Mechanical Ventilation, IPAP: inspiratory airway pressure, MIP: maximal static inspiratory pressure, mmHg: millimeters of mercury (pressure), NMD: Neuromuscular Disease, NOS: Not otherwise Specified, PaO<sub>2</sub>: partial pressure of arterial oxygen, PaCO<sub>2</sub>: partial pressure of arterial carbon dioxide, Pimax: maximal inspiratory mouth pressures, PSV: Pressure support ventilation, RCT: randomized controlled trial, SaO<sub>2</sub>: arterial blood oxygen saturation, SpO<sub>2</sub>: peripheral capillary oxygen saturation, ST: spontaneous/timed breath mode

**Table F.6. Neuromuscular Disease – Established home device use**

Author, Year, Study Design	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
Pinto, 2010 <sup>54</sup> Observational	BPAP ST + weekly telemonitoring versus BPAP ST without weekly telemonitoring	-ALS NOS -home BPAP use -FVC $\geq$ 75%	-PaCO <sub>2</sub> $\leq$ 45 mmHg -PaO <sub>2</sub> $\geq$ 80 mmHg	Increase in IPAP to achieve normal breathing patterns, daytime and nocturnal SaO <sub>2</sub> $>$ 95%. Backup rate set slightly lower than the patient's own respiratory frequency. (Titration occurred in hospital or outpatient clinic)
Gonzalez-Bermejo, 2013 <sup>32</sup> Observational	BPAP ST	-ALS NOS -home BPAP with 4 hours/night minimal adherence		Maximum tolerated IPAP to target patient comfort, leaks, and efficiency of ventilation, relieve symptoms, and achieve normal daytime PaO <sub>2</sub> , PaCO <sub>2</sub> , and SpO <sub>2</sub> . EPAP ranged from 3-5 cmH <sub>2</sub> O.

ALS: amyotrophic lateral sclerosis, BPAP: Bilevel Positive Airway Pressure, cmH2O: centimeters of water (pressure), , EPAP: expiratory positive airway pressure, FVC: Forced vital capacity, IPAP: inspiratory positive airway pressure, mmHg: millimeters of mercury (pressure), PaO2: partial pressure of arterial oxygen, PaCO2: partial pressure of arterial carbon dioxide, NOS: Not otherwise Specified, SaO2: arterial blood oxygen saturation, SpO2: peripheral capillary oxygen saturation, ST: spontaneous/timed breath mode

**Table F.7. Obesity Hypoventilation Syndrome - New initiation of home device**

<b>Author, Year, Study Design</b>	<b>Device/mode</b>	<b>Patient characteristics to start or continue device</b>	<b>Laboratory characteristics to start or continue device</b>	<b>Device titration</b>
Howard, 2016 <sup>36</sup> RCT	BPAP ST versus CPAP	-OHS (BMI >30, daytime PaCO2 >45 mmHg, other causes of hypoventilation ruled out including NMD, chest wall abnormalities, respiratory depressant medications, COPD, FEV1/FVC <70% after bronchodilators)	-PaCO2 >45 mmHg (daytime) -pH 7.35-7.45	BPAP ST: IPAP and EPAP titrated to overcome obstructive events and nocturnal hypoventilation CPAP: Fixed pressure titrated to overcome obstructive events in polysomnography
Salturk, 2015 <sup>57</sup> Observational	BPAP ST	-OHS (BMI>30, daytime PaCO2 ≥ 45 mmHg and symptoms of hypercapnia, no other cause of hypoventilation)	-PaCO2 >45 mmHg (daytime)	IPAP titrated to achieve “desired tidal volume” (maximum 30 mbar)
Masa, 2000 <sup>39</sup> Observational	HMV (volume cycled or pressure cycled)	-OHS (BMI>33; PaCO2 >47 mmHg for 3 months; weight loss failure; refusal for weight loss surgery) -FEV1/FVC ≥65% -Apnea-hypopnea index ≤ 20 events/hour	-PaCO2 >47 mmHg for at least 3 months	Ventilator parameters adjusted to target maximum reduction in PaCO2 as well as patient tolerance, air leakage, and nocturnal saturation >90%. Patient initially treated with volume-cycled ventilator. Patients with poor compliance to volume-cycled ventilator were switched to a bilevel pressure ventilator. (Titration occurred in hospital over 3-7 days)
Castillejo, 2014 <sup>11</sup> Observational	BPAP ST in OHS without OSA compared to BPAP ST in OHS with OSA	-OHS (BMI >30, daytime PaCO2 >45 mmHg, nighttime PaCO2 > 50 mmHg, with or without associated OSA, other causes of hypoventilation excluded (FEV/FVC ratio <70%, NMD with respiratory involvement, respiratory disease other than OHS)	-PaCO2 >45 mmHg (daytime, PaCO2 > 50 mmHg (nighttime)	IPAP adjusted during daytime to target PaCO2 < 45 mmHg or a decrease from baseline by 5 mmHg with a mean SaO2 > 90% (IPAP range 16-24 cmH2o). EPAP 6-10 cmH2O. Pressures further adjusted at nighttime via polysomnography.

Author, Year, Study Design	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
Masa, 2015 <sup>40, 41</sup> RCT	HMV/BPAP mix (all with bilevel pressure with assured volume) versus CPAP (fixed pressure)	-OHS (BMI $\geq$ 30; stable PaCO <sub>2</sub> $\geq$ 45 mmHg; pH $\geq$ 7.35; no clinical worsening in prior 2 months; other causes of hypoventilation ruled out including no evidence of COPD, NMD, narcolepsy) -Severe OSA (apnea-hypopnea index $\geq$ 30) -Correctly executed 30min CPAP/NIPPV treatment trial test	-PaCO <sub>2</sub> $\geq$ 45 mmHg -pH $\geq$ 7.35	HMV/BPAP mix: IPAP maximum tolerated to target reduction in PaCO <sub>2</sub> , normal SaO <sub>2</sub> , patient tolerance, target volume of 5-6 ml/kg of actual body weight. IPAP range 18-22 mmHg. EPAP range 4-8 mmHg. Pressures further adjusted in polysomnography to treat apneas and hypopneas.  CPAP: Polysomnography to eliminate apneas, hypopneas, thoracoabdominal paradoxical movement, flow limitation, and snoring.
Borel, 2011 <sup>6</sup> RCT	BPAP ST	-OHS (BMI >30; daytime PaCO <sub>2</sub> $\geq$ 45 mmHg, other causes of hypoventilation ruled out including airway obstruction, scoliosis, cardiac failure, progressive NMD)	-PaCO <sub>2</sub> $\geq$ 45 mmHg (daytime)	(Titration occurred in hospital over 3-4 nights)
Murphy, 2012 <sup>45</sup> RCT	BPAP (volume assured pressure support ventilation) versus BPAP ST	-OHS (BMI >40, daytime chronic PaCO <sub>2</sub> >6 kPa, pH >7.35), absence of other identifiable hypoventilation cause, FEV <sub>1</sub> /FVC >70%, FVC <70% -Stable disease	-PaCO <sub>2</sub> >6 kPa (45 mmHg) -pH >7.35 (daytime)	Titration according to a protocol with goal to abolish apneas, snoring, and "to achieve adequate nocturnal respiratory control" (See online data supplement of primary article)
Piper, 2008 <sup>55</sup> RCT	BPAP S versus CPAP	-OHS (BMI $\geq$ 30, PaCO <sub>2</sub> $\geq$ 45 mmHg [awake, stable], absence of another cause for hypercapnia, FEV <sub>1</sub> /FVC $\geq$ 70%)	-PaCO <sub>2</sub> $\geq$ 45 mmHg -pH $\geq$ 7.34 (daytime, stable)  Excluded during CPAP titration study: -SaO <sub>2</sub> <80% for 10 minutes in absence of apnea -TcCO <sub>2</sub> during REM $\geq$ 10mmHg -increase in afternoon to morning PaCO <sub>2</sub> $\geq$ 10mmHg in patients with awake PaCO <sub>2</sub> >55 mmHg	

Author, Year, Study Design	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
Blankenburg, 2017 <sup>5</sup>	HMV (pressure controlled ventilation or pressure support ventilation)	-OHS (BMI>30, PaCO <sub>2</sub> >6.7kPa[50mmHg], symptoms of hypercapnia NOS), absence of another cause for hypercapnia	-PaCO <sub>2</sub> >7.0kPa (53mmHg) -pH>7.35	Goal of titration was normal PaCO <sub>2</sub> as well as patient tolerability of NIPPV. Titration started in pressure controlled ventilation mode. If pressure controlled ventilation was not achievable, pressure support ventilation was used. Inspiratory pressure was set to relieve “air hunger” on inspiration or to reach a tidal volume ≥800mL. PEEP was increased to maximally tolerated. Respiratory rate was set at 2 breaths/minute above the spontaneous respiratory rate.
Tsolaki, 2011 <sup>66</sup> Observational	BPAP ST	-OHS NOS -Stable (no acute exacerbation in prior 4 weeks) -Symptoms of nocturnal hypoventilation (fatigue, dyspnea, morning headaches) -BMI > 30 -persistent hypoventilation despite overnight trial of CPAP	-PaCO <sub>2</sub> >45mmHg -PaO <sub>2</sub> <70mmHg -pH>7.35	Patients were hospitalized for 2–3 days during the initial application of NIV, in order to ensure maximal compliance. The modality of NIV was pressure support ventilation delivered by a variable positive airway pressure device (VPAP III ST, ResMed, Sydney, N.S.W., Australia), set in the spontaneous/timed mode with a backup respiratory rate of 8–14 breaths / min. Inspiratory and expiratory positive airway pressures (IPAP and EPAP, respectively) were adjusted according to the patient’s comfort and synchrony with the ventilator and a marked reduction in the use of accessory muscles. For the first 2 h from the initiation of ventilation the patient was observed by a pulmonologist and a fully trained nurse, in order to assure a decrease in accessory respiratory muscle use and respiratory rate, check the patient’s comfort of mask ventilation and possible air leaks of the system. Supplemental oxygen was added as needed in order to maintain oxygen saturation between 88 and 92%. Arterial blood gases were measured 1 h after the initiation of ventilation. A 5% decrease in Pa CO <sub>2</sub> values was considered as adequate ventilatory support.
Masa, 2016 <sup>42</sup> RCT	BPAP volume assured pressure support ventilation	-OHS (BMI ≥ 30 kg/m <sup>2</sup> , no COPD, no NMD, no TRD, no narcolepsy, no restless leg syndrome) -Stable disease	-PaCO <sub>2</sub> ≥ 45 mmHg, (daytime, awake) -pH ≥ 7.35	The ventilator mode was set at bilevel pressure with assured volume (ie, volume targeted pressure support). While the patient was awake, the expiratory positive airway pressure (EPAP) was initially set between 4 and 8 cm H <sub>2</sub> O and the inspiratory positive airway pressure (IPAP) was set between 18 and 22 cm H <sub>2</sub> O (EPAP included). The pressures were adjusted to obtain normal oxygen saturation, if possible, as measured by pulse oximetry and patient tolerance. The backup respiratory



Author, Year, Study Design	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
				<p>rate was initially adjusted to 12–15 breaths/min (close to the spontaneous respiratory rate, if possible) and the target volume was set at between 5 and 6 mL/kg of actual weight, allowing for an increase in the maximum pressure over the previously minimum IPAP, if necessary. A check of mechanical ventilation phases (trigger, pressurisation and ending) was also performed to avoid asynchronies and to refine the setting. After 30 min of continuous use with patient adaptation and an adequate patient–ventilator interaction, an ABG analysis was performed. The PaCO<sub>2</sub> result was used to adjust the ventilator parameters. The final adjustment was performed by means of conventional PSG, with an increase in EPAP for obstructive apnoeas and an increase in IPAP for hypopnoeas, flow limitation, snoring or non-apnoeic hypoventilation, with the goal of achieving normalisation of oxygen saturation or the maximal pressure tolerated was reached. No changes were made in the assured volume during this nocturnal titration</p>
Perez de Llano, 2005 <sup>52</sup> , Observational	HMV/BPAP mix	-OHS, BMI > 30, , FEV1/FVc < 70%, absence of other respiratory disorder such as kyphoscoliosis or diaphragmatic paralysis	-PaCO <sub>2</sub> ≥ 50 mmHg	<p>Treatment with NIPPV was started in all patients who experienced respiratory failure presumed to be secondary to OHS. Patients were treated initially with bilevel pressure devices (DP-90 and Eclipse Delta; Taema; Antony, France; and PV-102; BREAS; Gothenburg, Sweden), but, in those patients who did not achieve sufficient improvement with this system, we subsequently changed over to a volume-cycled ventilator (Home 2; Airox; Pau, France). The interface used in all patients was a commercially available nasal mask that was secured with head straps. Initially, positive expiratory pressure (PEP) was set at 6 cm H<sub>2</sub>O, and the positive inspiratory pressure (PIP) was set at 10 cm H<sub>2</sub>O. PIP was gradually adjusted upward as tolerated. Oxygen was administered, when needed, through the mask until the arterial oxygen saturation (Sao<sub>2</sub>) was ≥ 90%. Daytime sessions lasted from 3 to 6 h with pauses of 3 h to allow the administration of conventional medication and feeding. Nighttime sessions were continuous, provided that patient tolerance permitted. When arterial blood gas levels were stable (<i>ie</i>, pH &gt; 7.35), daytime NIPPV therapy was stopped. We employed daytime arterial blood gas measurements and overnight pulse oximetry to determine</p>

Author, Year, Study Design	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
				the NIPPV settings. We gradually increased PEP until the disappearance of repetitive dips in Sao <sub>2</sub> was achieved. PIP was then increased until an acceptable level of steady saturation was obtained. We considered treatment with NIPPV to be successful if orotracheal intubation had been avoided in patients with an initial pH of < 7.34 and, for the entire group, when the mean Sao <sub>2</sub> during overnight oximetry was ≥ 88% and diurnal Paco <sub>2</sub> was ≤ 65 mm Hg with a normal pH. Then, the patients could be discharged from the hospital, and they were instructed to employ NIPPV during the night with the final settings obtained.
Priou, 2010, <sup>56</sup> Observational	BPAP	-OHS (PaCO <sub>2</sub> > 45 mm Hg in the absence of any other cause of hypoventilation on the basis of clinical examination, chest radiograph, and pulmonary function tests (eg, COPD [FEV <sub>1</sub> to vital capacity ratio, 70%]).	PaCO <sub>2</sub> > 45 mm Hg	Expiratory positive airway pressure (EPAP) and inspiratory positive airway pressure (IPAP) were adapted by 2 cm H <sub>2</sub> O steps using repeated oximetry and arterial blood gases (ABG) to alleviate OSA-related desaturations, to improve mean nocturnal oxygen desaturation index (SaO <sub>2</sub> ), and to achieve a maximal reduction in daytime PaCO <sub>2</sub> . Supplemental oxygen was added to NPPV in patients with persistent nocturnal hypoxia (as defined arbitrarily by ≥ 20% of time with SaO <sub>2</sub> , < 90%) despite a delta between EPAP and IPAP of at least 10 cm H <sub>2</sub> O as tolerated by the patient.

BPAP: Bilevel Positive Airway Pressure, COPD; chronic obstructive pulmonary disease, CPAP: Continuous Positive Airway Pressure, BMI: Body Mass Index, EPAP: expiratory positive airway pressure, FEV<sub>1</sub>: Forced expiratory volume in one second, FVC: Forced vital capacity, HMV: Home Mechanical Ventilation, IPAP: inspiratory positive airway pressure, kPa: kilopascal, mmHg: millimeters of mercury (pressure), NIPPV: Noninvasive Positive Pressure Ventilation, NMD: Neuromuscular Disease, OHS: Obesity hypoventilation syndrome, OSA: Obstructive sleep apnea, PaCO<sub>2</sub>: partial pressure of arterial carbon dioxide, S: spontaneous mode, SaO<sub>2</sub>: arterial blood oxygen saturation, ST: spontaneous/timed breath mode, TcCO<sub>2</sub>: transcutaneous carbon dioxide

**Table F.8. Other Respiratory Diseases - New initiation of home device**

Author, Year, Study Design	Disease	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
Salturk, 2015 <sup>57</sup> Observational	Diffuse parenchymal lung disease	BPAP ST	-Diffuse parenchymal lung disease (sequela of TB or bronchiectasis with hypoxemia and hypercapnia)		IPAP titrated to achieve "desired tidal volume" (maximum 30 mbar)

BPAP: Bilevel Positive Airway Pressure, IPAP: inspiratory positive airway pressure, ST: spontaneous/timed breath mode, TB: tuberculosis

**Table F.9. Other Respiratory Diseases – Established home device use**

Author, Year, Study Design	Disease	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
Benhamou, 1997 <sup>2</sup> Observational	Diffuse bronchiectasis	HMV (volume cycled)	-Diffuse bronchiectasis -Home HMV -LTOT		Target PaO <sub>2</sub> > 9kPa (67 mmHg) without deterioration in PaCO <sub>2</sub> .

HMV: Home Mechanical Ventilation, kPa: kilopascal, LTOT: long term oxygen therapy, mmHg: millimeters of mercury (pressure), PaO<sub>2</sub>: partial pressure of arterial oxygen, PaCO<sub>2</sub>: partial pressure of arterial carbon dioxide

**Table F.10. Mixed diseases – New initiation of home device**

Author, Year, Study Design	Disease	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
Windisch, 2006 <sup>69</sup> Observational	TRD, OHS	HMV with pressure controlled ventilation (PCV) mode	-COPD NOS -NIPPV in hospital admission -Stable (no worsening symptoms in prior 2 weeks, respiratory rate <30 breaths/minute, no signs of current respiratory infection, no changes in symptoms or medications in prior 3 months)	-pH≥7.35	Maximum tolerated IPAP to target a maximum decrease in PaCO <sub>2</sub>
Hazenberg, 2014 <sup>33</sup>	NMD, TRD	HMV (pressure or volume)	-NMD or thoracic cage disorder -Stable disease without acute respiratory failure	-PaCO <sub>2</sub> >6.0 kPa (>45 mmHg) (daytime)	Maximum tolerated IPAP to target a target tidal volume of 8-10 ml/kg and a

Author, Year, Study Design	Disease	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
RCT		control) started at home			respiratory rate close to the baseline respiratory rate, reduce snoring, patient comfort. Titration of ventilator parameters to achieve normal PaCO <sub>2</sub> and PaO <sub>2</sub> . (Titration occurred at home)
		HMV (pressure or volume control) started in the hospital	-NMD or thoracic cage disorder -Stable disease without acute respiratory failure	-PaCO <sub>2</sub> >6.0 kPa (>45 mmHg) (daytime)	Maximum tolerated IPAP to target a target tidal volume of 8-10 ml/kg and a respiratory rate close to the baseline respiratory rate, reduce snoring, patient comfort. Titration of ventilator parameters to achieve normal PaCO <sub>2</sub> and PaO <sub>2</sub> . (Titration occurred at the hospital)
Munoz, 2005 <sup>44</sup> Observational	NMD, TRD	HMV volume assist/control mode versus HMV volume control mode	-Hospital admission with chronic hypercapnic respiratory failure to NMD (ALS excluded) or kyphoscoliosis or post TB sequelae	-PaCO <sub>2</sub> > 45 mmHg (daytime, stable)	The tidal volume, respiratory frequency, and the I/E ratio were adjusted individually according to tolerance, air leaks, and ventilatory response.
Chiang, 2003 <sup>13</sup> RCT	COPD, Other	BPAP NOS	-COPD or asthma or bronchiectasis -hospital readmission due to respiratory cause -Daytime sleepiness or morning headache	-PaCO <sub>2</sub> > 50 mmHg (daytime rest) -SpO <sub>2</sub> < 88% for more than 5 consecutive minutes while on usual oxygen during polysomnography	IPAP and EPAP and volumes set to target optimal daytime PaCO <sub>2</sub>

BPAP: Bilevel Positive Airway Pressure, COPD; chronic obstructive pulmonary disease, EPAP: expiratory positive airway pressure, HMV: Home Mechanical Ventilation, IPAP: inspiratory positive airway pressure, mmHg: millimeters of mercury (pressure), NIPPV: Noninvasive Positive Pressure Ventilation, NMD: Neuromuscular Disease, NOS: Not otherwise Specified, OHS: Obesity hypoventilation syndrome, PaO<sub>2</sub>: partial pressure of arterial oxygen, PaCO<sub>2</sub>: partial pressure of arterial carbon dioxide, RCT: randomized controlled trial, SpO<sub>2</sub>: peripheral capillary oxygen saturation, TB: tuberculosis, TRD: Thoracic Restrictive Disorder

**Table F.11. Mixed diseases – Established home device use**

Author, Year, Study Design	Disease	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
Crespo, 2009 <sup>18</sup> Observational	COPD, TRD, NMD, OHS, Other	HMV (pressure or volume NOS)	-home HMV use -stable respiratory disease (all cause)		

COPD: chronic obstructive pulmonary disease, HMV: Home Mechanical Ventilation, NMD: Neuromuscular Disease, NOS: Not otherwise Specified, OHS: Obesity hypoventilation syndrome, TRD: Thoracic Restrictive Disorder

KQ2. In each of the disease groups, what is the effect of HMV, a BPAP, or a CPAP use on patient outcomes?

**Table F.12. COPD – Effectiveness of home devices**

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
Vasquez, 2017 <sup>67</sup> Observational	COPD	Inclusion: COPD (ICD-9); age ≥40 years	1) BPAP NOS	Longest duration: 6 months	The HMV group had significantly more reduction of mortality than those with CPAP (p<0.001) or BPAP (p<0.001), and more reduction on COPD-related hospitalization than the CPAP group (p=0.01).
			2) CPAP NOS		
			3) HMV NOS		
Murphy, 2017 <sup>46</sup> , RCT	COPD	Inclusion: COPD (FEV1 < 50%, FEV1/FVC ratio <60%, smoking history >20 pack	1) BPAP ST + home oxygen	Longest duration: 12 months	The BPAP ST group had significantly fewer AECOPD than the home oxygen alone group (rate ratio, 0.66;

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
		<p>years); AECOPD requiring hospital admission and acute NIPPV; PaCO<sub>2</sub> &gt;53 mmHg; PaO<sub>2</sub> &lt;55 mmHg or PaO<sub>2</sub> &lt; 60 mmHg with polycythemia, pulmonary hypertension or cor pulmonale; &gt;30% sleep time with SaO<sub>2</sub> &lt;90%; pH &gt;7.30 room air</p> <p>Exclusion: intubated during AECOPD, current home NIPPV use, cognitive impairment, unstable psychiatric morbidity, undergoing renal replacement therapy, unstable coronary artery syndrome, age &lt; 18 years, homeless, BMI &gt;35, OSA.</p>	2) Home oxygen		<p>95%CI, 0.46-0.95, p = 0.03). Twelve month mortality was not significantly different between the two groups (HR, 0.67; 95%CI, 0.34- 1.30, p = 0.23). Quality of life at 12 months was not significantly different between the groups.</p>
Oscroft, 2014 <sup>50</sup> , RCT	COPD	<p>Inclusion: COPD (FEV1 &lt;50%, FEV1/FVC ratio &lt;70%, TLC&gt;80%, smoking history &gt; 20 pack years); daytime PaCO<sub>2</sub> &gt;7 kPa and pH &gt;7.35</p>	1) BPAP volume assured pressure support ventilation	3 months	<p>The BPAP volume assured pressure support ventilation group had significantly shorter hospital stay than the BPAP ST group (3.3 days vs. 5.2 days, p=0.02).</p>

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
		<p>or PtcCO<sub>2</sub> &gt;9 kPa</p> <p>Exclusion: Age&gt;80 years, other respiratory disease, BMI&gt;40, significant OSA.</p>	2) BPAP ST	3 months	There was no significant difference on mortality (OR=0.47, 95% CI: 0.04 to 5.69; p=0.56), exercise tolerance, dyspnea, quality of life, or sleep quality after 3-month followup.
Paone, 2014 <sup>51</sup> , Observational	COPD	<p>Inclusion: COPD (FEV<sub>1</sub> &lt;50%, FEV<sub>1</sub>/FVC ratio &lt;70%, &lt;20% improvement bronchodilator response); NIPPV during hospital stay; PaCO<sub>2</sub> &gt; 50 mmHg immediately after awakening from a night without NIPPV</p> <p>Exclusion: Significant comorbidities affecting survival (cancer, left ventricular heart failure, unstable angina), psychiatric disorders affecting ability to undergo NIPPV, other chronic respiratory disease, history of OSA, BMI&gt;40, systemic corticosteroids.</p>	<p>1) BPAP ST + Home oxygen</p> <hr/> <p>2) Home oxygen</p>	24 months	The BPAP ST + home oxygen group had significantly less hospital admissions (Rate Ratio= 0.50; 95% CI: 0.35 to 0.71; p<0.01). There was no significant difference on mortality (27.1% vs. 22.2%; p=0.59).



Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
Galli, 2014 <sup>29</sup> , Observational	COPD	Inclusion: AECOPD (ICD-9); PaCO <sub>2</sub> > 45 mmHg; NIPPV during hospital stay  Exclusion: discharged to hospice.	1) BPAP NOS	Longest duration: 6 months	BPAP was associated with significantly fewer hospital readmissions (p < 0.0001) and ICU readmissions. There was no significant difference on mortality at 6-month followup (10% vs. 19%, p=0.13).
			2) No BPAP		
Bhatt, 2013 <sup>4</sup> , RCT	COPD	Inclusion: COPD (FEV/FVC < 70%, smoking >10 pack years); no exacerbations in past 4 weeks; low clinical probability of OSA  Exclusion: Congestive heart failure, OSA, chronic respiratory conditions other than COPD, age < 35 years, diseases limiting life expectancy < 2 years, active malignancies in previous 2 years, process precluding a nasal mask.	1) BPAP NOS	Longest duration: 6 months	BPAP was associated with significantly higher quality of life scale (measured by Chronic Respiratory disease Questionnaire) than the no BPAP group (p=0.04). There was no significant difference on exacerbations, exercise tolerance (6-minute walk distance test), dyspnea, and sleep quality.
			2) No BPAP		

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
Duiverman, 2011 <sup>22, 23</sup> , RCT	COPD	<p>Inclusion: COPD (FEV1 &lt;50%, FEV1/FVC &lt; 70%, GOLD stage III/IV); age 40-76 years; no exacerbation in past 4 weeks; daytime PaCO2 &gt;6.0 kPa</p> <p>Exclusion: cardiac/neuromuscular disease limiting exercise tolerance, pulmonary rehabilitation in past 18 months, prior NIPPV, apnea-hypopnea index ≥10h.</p>	1) BPAP ST + Pulmonary rehabilitation	Longest duration: 24 months	<p>At 24 months, BPAP was associated with significantly better outcomes, including dyspnea (Medical Research Council -0.4; 95% CI: -0.8 to -0.0), 6-minute walk distance test (77.3 meters, 95% CI: 46.4 to 108.0), and activities of daily living (Groningen Activity and Restriction Scale, -3.8, 95% CI: -7.4 to -0.4). No significant difference was found on mortality (OR= 0.94, 95% CI: 0.25 to 3.57), quality of life (Chronic Respiratory Questionnaire) (-1.3; 95% CI: -9.7 to 7.4), exacerbation frequency, and hospitalization rate.</p>
			2) Pulmonary rehabilitation alone		
Oscroft, 2010 <sup>48</sup> , Observational	COPD	<p>Inclusion: COPD (FEV1 &lt;50%, FEV1/FVC ratio &lt;70%, smoking history &gt;20 pack years); AECOPD requiring hospital admission; daytime PaCO2 &gt;7.5 kPa with pH 7.35-7.45 or daytime PaCO2 &gt;6.5 kPa with pH</p>	1) BPAP ST started in AECOPD	28.6 months, 95% CI 10.9-46.8 months, Median 52.4 months	<p>The BPAP ST started in AECOPD group had significantly shorter median survival time than the stable group (28.6 months vs. 52.6 months, p=0.03).</p>

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
		<p>7.35-7.45 with PtcCO<sub>2</sub> &gt;9 kPa</p> <p>Exclusion: Age&gt;80 years, other respiratory disease, BMI&gt;35, significant OSA, tracheostomy, impaired left ventricular function.</p>	2) BPAP ST started in stable COPD		
Cheung, 2010 <sup>12</sup> RCT	COPD	<p>Inclusion: AECOPD requiring hospital admission and NIPPV, pH &lt;7.35, PaCO<sub>2</sub> &gt; 6 kPa</p> <p>Exclusion: Active smokers, RF from other cause, pneumonia, transmissible infections, long-term corticosteroid use, comorbidity giving life expectancy &lt;1 year, significant OSA, already on home NIPPV</p>	<p>1) CPAP</p> <p>2) BPAP ST</p>	Longest duration: 12 months	<p>7 out of 23 patients in the BPAP group developed severe COPD exacerbation with AHRF while 14 out of 26 patients in the COPD group had severe exacerbation with AHRF (OR= 0.38, 95% CI: 0.12 to 1.22; p=0.10). 8 patients in the BPAP group withdrew from the study, compared to 4 patients in the CPAP group (OR= 2.93; 95% CI: 0.75 to 11.52; p=0.12). No significant difference of number of adverse events were found between the two groups (p=0.29).</p>

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
McEvoy, 2009 <sup>43</sup> RCT	COPD	<p>Inclusion: COPD (FEV1&lt;50% or &lt;1.5L, bronchodilator response &lt;20%, FEV1/FVC ratio &lt;60%); PaCO2 &lt;46 mmHg at least twice in the prior 6 months during clinical stability; LTOT for ≥3 month; Age&lt;80 years</p> <p>Exclusion: current smokers, significant comorbidities (malignancies, left ventricular HF, unstable angina) likely affecting 2 year survival, severe psychiatric disorder impairing ability to comply to NIPPV, BMI&gt;40, evidence of sleep apnea.</p>	1) BPAP S + Oxygen	Longest duration: 12 months	No significant difference was found on survival (unadjusted HR: 0.82; 95% CI 0.53 to 1.25, OR= 0.71; 95% CI: 0.36 to 1.38), quality of life and hospitalization rates.
			2) Oxygen alone		
Casanova <sup>10</sup> , 2000 RCT	COPD	Inclusion: COPD (FEV1 <45%, FEV1/FVC <70%, smoking >20 pack years, TLC ≥80%); stable disease (no AECOPD in past 3 months); age 45-75	1) BPAP S + Standard care	Longest duration: 12 months	There were no significant differences on mortality, the number of acute exacerbations, hospital admissions, intubations, dyspnea (Medical Research

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
		years  Exclusion: current smoker, OSA, apnea-hypopnea index >10/hour, other etiologies of chronic airway obstruction, significant comorbidities.	2) Standard care		Council).
Garrod, 2000 <sup>30</sup> RCT	COPD	Inclusion: COPD (FEV1 <50%, bronchodilator response <15%); exercise intolerance due to dyspnea, no prior NIPPV  Exclusion: unstable angina, intermittent claudication, other mobility-limiting conditions.	1) BPAP S + Pulmonary rehabilitation  2) Pulmonary rehabilitation		The BPAP S plus pulmonary rehabilitation had significantly better outcomes on quality of life (Chronic Respiratory Disease Questionnaire, 12.3; 95% CI: 1.19 to 23.4; p=0.03), and shuttle walk test (72 meters, 95% CI: 12.9 to 131 meters). There was no difference on activities of daily living, and dyspnea.
Clini, 1998 <sup>15</sup> Observational	COPD	Inclusion: COPD, prior smokers, LTOT ≥12 month; stable disease (no AECOPD in prior 4 weeks); stable PaCO <sub>2</sub> ; pH>7.35; PaO <sub>2</sub> < 8 kPa (daytime room air),	1) BPAP ST + Oxygen	Longest duration: 2 months	The BPAP plus oxygen group was found to have significantly more changes in 6-minute walk distance test than the oxygen group

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
		PaCO <sub>2</sub> >6 kPa (daytime room air); ≥1 ICU admission due to AECOPD in prior 2 years  Exclusion: other organ failure, cancer, suspected OSA.	2) Oxygen		(p<0.01). There were no significant differences on mortality (OR=0.79, 95% CI; 0.25 to 2.45); or changes in dyspnea (American Thoracic Society).
Clini, 1996 <sup>14</sup> , Observational	COPD	Inclusion: COPD, LTOT ≥18 mo.; chronic PaCO <sub>2</sub> >6.7 kPa (50 mmHg); ≥1 hospital admission due to AECOPD in prior 18 months  Exclusion: suspected OSA, ≥15% bronchodilator response, comorbidities making patients unsuitable for long-term trials.	1) BPAP ST + Home care + Oxygen	Longest duration: 18 months	During the 18 month followup, there was no difference on mortality (23% vs. 18%), ICU admissions (rate ratio: 0.29; 95% CI: 0.06 to 1.38) and hospital admissions (rate ratio: 0.88, 95% CI: 0.44 to 1.77).
			2) Home care + Oxygen		
Zhou, 2017 <sup>70</sup> RCT	COPD	Inclusion: COPD (Gold Stage III/IV); chronic hypercapnia (measured during daytime at rest with no oxygen or NIPPV); age ≥40 years  Exclusion:	1) BPAP ST	3 months	Significantly more patients in the BPAP ST group achieved the minimum clinical improvement on 6-minute walk distance test (38.2% vs. 18.2%, p=0.02) than the

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
		Abnormalities of lung/thorax other than COPD, previously treated on NIPPV, OSA, severe HF, severe arrhythmias, unstable angina, malignant comorbidities, COPD w/ OSA overlap syndrome, impairments that could affect ability for followup.	2) Standard care		standard care group.  No significant difference was found on mortality, and quality of life (Severe Respiratory Insufficiency Questionnaire).
Marquez-Martin, 2014 <sup>38</sup> RCT	COPD	Inclusion: COPD (FEV1 <50%); PaO2 < 60 mmHg (chronic); PaCO2 > 45 mmHg (chronic).	1) BPAP ST	3 months	In 6-minute walk distance test, patients in the BPAP ST group increased by 40 meters (p=0.01); 32 meters in the exercise group (p=0.01) and 83 meters in the

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
			2) Exercise program		combined group (p<0.001). No significant difference was found between the groups on 6-minute walk distance test, and dyspnea (Medical Research Council, 1 vs.1.5 vs.1, p=0.6), and quality of life (Chronic Respiratory Disease Questionnaire, 4.6 vs. 5.61 vs.5.26, p=0.06).
Köhnlein, 2014 <sup>37</sup> RCT	COPD	Inclusion: COPD (GOLD IV); clinically stable (no AECOPD in prior 4 weeks); PaCO <sub>2</sub> ≥ 7	1) BPAP ST + Standard care	12 months	The BPAP group was found to have significantly less mortality rate at 1



Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
		<p>kPa (51.9 mmHg); pH <math>\geq</math> 7.35 (rest)</p> <p>Exclusion: Thorax/lung abnormalities other than COPD, BMI<math>\geq</math>35, other conditions resulting in hypercapnia, previously initiated NPPV, malignant comorbidities, severe HF, unstable angina, severe arrhythmias.</p>	2) Standard care		<p>year (HR=0.24, 95% CI: 0.11 to 0.49). The difference was significant after 1 year. The BPAP group had better outcomes on quality of life (Saint George's Respiratory Questionnaire, 6.2, 95% CI: 0.7 to 11.8). Patients were electively admitted to hospital for 2.0 (0.1) days in the standard care group and 3.1 (0.9) days in the BPAP group. No significant difference was found on 6-minute walk distance test.</p>
De Backer, 2011 <sup>19</sup> RCT	COPD	<p>Inclusion: COPD (FEV1&lt;50%, FEV1/FVC &lt;70%), AECOPD requiring hospitalization, PaCO<sub>2</sub> &gt;45 mmHg, stopped smoking</p>	1) BPAP NOS	At least 6 months	<p>The 6-minute walk distance increased significantly in the BPAP group (232 <math>\pm</math> 151 m to 282 <math>\pm</math> 146 m, p = 0.01), while there was</p>

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
		Exclusion: home NIPPV prior to admission, invasive ventilation, asthma, restrictive lung disease, malignancy, HF, OSA.	2) Standard care	At least 6 months	no change in the control group (408 ± 34 m to 401 ± 78 m, p = 0.09). No significant difference was found between the groups.
Funk, 2010 <sup>27</sup> RCT	COPD	Inclusion: COPD; AECOPD requiring NIPPV or invasive ventilation; chronic nocturnal NIPPV use at home for ≥ 6 months; clinically stable, PaCO <sub>2</sub> > 45 mmHg immediately after awakening from night without NIPPV  Exclusion: Severe psychiatric disorder likely to impair NIPPV compliance, other severe pulmonary diseases not COPD, other severe non-pulmonary diseases limiting prognosis, noncompliance to NIPPV, women of childbearing age, evidence of sleep apnea.	1) BPAP NOS for 6 months  2) BPAP NOS for more than 6 months	Longest duration: 12 months	Patients who received BPAP more than 9 months had significantly increases (43%) in the 6-minute walk distance test while the group with 6-month treatment decreased by 11% (p =0.04). No significant difference was found on quality of life (the Saint George's Respiratory Questionnaire).

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
Dreher, 2010 <sup>21</sup> RCT	COPD	Inclusion: COPD (Gold stage IV); daytime PaCO <sub>2</sub> > 45 mmHg; nocturnal PaCO <sub>2</sub> > 50 mmHg  Exclusion: Acute RF, invasive ventilation via tracheostomy, weaned from invasive ventilation, intubated during prior 3 months, other ventilatory support prior to study.	1) HMV (pressure controlled ventilation) (time period 1)	1.5 months	Treatment compliance was higher in the HMV (pressure controlled ventilation) group than the HMV (pressure support ventilation) group (10.8 hours per day vs. 7.7 hours per day, p=0.02). The HMV (pressure controlled ventilation) group had higher Borg dyspnea scale after 6-minute walk distance test (2.4, 95% CI: 0.4 to 4.3, p=0.03). There were no significant difference on quality of life (Severe Respiratory Insufficiency Questionnaire Summary Score), and 6-minute walk distance test.
			2) HMV (pressure support ventilation)(time period 1)		
			2) Pulmonary rehabilitation alone		
Tsolaki, 2008 <sup>65</sup> Observational	COPD	Inclusion: COPD (FEV <sub>1</sub> <50%, FEV/FVC <70%); smoking >20 pack	1) BPAP ST	12 months	Compared to standard care, the BPAP group was found to have significantly better

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
		<p>years; Age≤75 years; PaO<sub>2</sub> &lt; 60 mmHg (room air); PaCO<sub>2</sub> &gt;50 mmHg (room air)</p> <p>Exclusion: Significant comorbidities (OSA, OHS, RF from disease other than COPD), important concomitant chronic systemic disorders, poor ventilator compliance, apnea-hypopnea index ≥10 episodes/hour.</p>	2) Standard care		<p>outcomes on Medical Research Council dyspnea score, Epworth Sleepiness Scale, SF-36 Physical Component Summary score, and SF-36 Mental Component Summary score. Patients in the BPAP group spent significantly less days in hospital (6.6 days vs. 16.0 days, p=0.02).</p> <p>There was no significant difference on number of exacerbations, hospitalization due to exacerbations, endotracheal intubation, or mortality.</p>
Chiang, 2003 <sup>13</sup> RCT	COPD, other	Inclusion: COPD or asthma or bronchiectasis; hospital readmission due to respiratory cause; daytime sleepiness or morning	1) BPAP NOS	6 months	Compared to the standard care group, the BPAP group had significantly better outcomes on 6-minute walk

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
		<p>headache; PaCO<sub>2</sub> &gt; 50 mmHg (daytime rest); SpO<sub>2</sub> &lt; 88% for more than 5 consecutive minutes while on usual oxygen during polysomnography</p> <p>Exclusion: Unable to tolerate nocturnal nasal positive pressure ventilation, OSA, unable to perform 6-minute walk distance test due to other disease.</p>	2) Standard care		<p>distance test group (101.2 meters vs. - 33.8 meters, p&lt;0.05), number of hospitalization, and total hospital stay. No significant difference was found on resting Borg score and Borg score at end of 6-minute walk distance test.</p>
Gay, 1996 <sup>31</sup> RCT	COPD	<p>Inclusion: COPD (FEV<sub>1</sub> &lt; 40%); PaCO<sub>2</sub> &gt;45 mmHg (daytime, rest); Age&lt;80 years, BMI≤30</p>	1) BPAP ST	3 months	<p>No difference was found on 6-minute walk distance test, total sleep time, sleep efficiency,</p>

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
		Exclusion: activated for lung transplantation, active psychiatric disease that necessitated sedative or hypnotic meds, current use of nocturnal ventilation or continuous PAP, major illness likely to preclude completion of prolonged trial.	2) Sham BPAP ST/no device		REM sleep, and multiple sleep latency tests.
Gad, 2014 <sup>28</sup> Observational	COPD	Inclusion: COPD (FEV1 < 50%, FEV1/FVC < 70%); clinically stable (no exacerbation in prior 4 weeks); PaCO2 ≥ 50 mmHg (daytime)  Exclusion: invasive MV, OSA, cardiac disease limiting exercise tolerance, NMDs, orthopedic impairment of shoulder girdle.	1) BPAP ST + Exercise program  2) Exercise program	3 months	After 3 month, compared to the exercise group, the BPAP group had significantly better outcomes on quality of life (COPD Assessment Test, 20.2 vs. 23, p=0.01).
Sin, 2007 <sup>62</sup> RCT	COPD	Inclusion: COPD (FEV1/FVC < 70%, post-bronchodilator	1) BPAP NOS + Standard care	3 months	After 3 months, the changes in 6-minute walk distance test was

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
		FEV1 <70%, smoking ≥10 pack years); age ≥40 years  Exclusion: Comorbidities making survival <6mo. Unlikely, clinical history of left ventricular HF, apnea-hypopnea index >20.	2) Sham BPAP/no device		significant in the BPAP group (30 meters, 95% CI, 2 to 57) while not significant in sham group (4 meters; 95% CI, - 38 to 47 m). However, the difference between the groups was not significant.
Heinemann, 2011 <sup>34</sup> Observational	COPD	Inclusion: COPD; prolonged weaning from invasive mechanical ventilation Exclusion: intubated due to cardiogenic edema or cardiopulmonary resuscitation	1) HMV pressure controlled ventilation	12 months	Patients received HMV were more likely to survive after 1-year followup than patients received standard care (HR=3.63, 95% CI: 1.23 to 10.75, p=0.02).
			2) No device		
Budweiser, 2007 <sup>8</sup> Observational	COPD	Inclusion: severe COPD (Global Initiative of Chronic Obstructive Lung Disease (GOLD) IV, FEV1/VC < 70% and FEV1 < 50% predicted, PaCO2 ≥50 mmHg after optimization of	1) BPAP ST	48 months	The BPAP ST group (mean followup: 19.8 months) had significantly lower mortality than those in the standard care group (mean followup: 12.9 months) (HR=0.48; 95% CI: 0.24 to 0.93, p<0.05).

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
		therapy or treatment of exacerbation), age<80 years Exclusion: prior diagnosis of a malignancy within 5 years, underwent intubation or tracheostomy prior to BPAP ST	2) Standard care/no device		
Clini, 2002 <sup>16</sup> RCT	COPD	Inclusion: severe COPD (he American Thoracic Society criteria), CVF, stable clinical condition (arterial pH>7.35, free from exacerbation in the 4 weeks), age≤75 years, LTOT at least 6 months, MRC dyspnea score≥2, FEV1<1.5 L, FEV1/FVC<60%, total lung capacity ≥90% predicted, PaCO2>6.6 kPa, PaO2<7.8 kPa Exclusion: 15% increase in FEV1 after inhaled	1) BPAP ST plus LTOT	24 months	Compared to the LTOT group, the BPAP ST plus LTOT group had significantly better outcomes on dyspnea (measured by the MRC scale, -0.60, 95% CI: -1.05 to -0.15), and sleep quality (measured by a semi-qualitative multipoint scale with a range 1 (best) to 4 (worst), -0.31, 95% CI: -1.0 to -0.1). There was no significantly difference on mortality (17% in both groups), exercise tolerance (measured by 6-minute walking



Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
		salbutamol (200 mg), pH $\leq$ 7.34, active smoking, history of obstructive sleep apnea syndrome, therapy with systemic steroids, important concomitant chronic systemic diseases (e.g. significant fibrothorax, bronchiectasis, cystic fibrosis), concomitant NPPV, other home care program apart from LTOT	2) LTOT		distance test), quality of life (measured by Saint George's Respiratory Questionnaire, p=0.55), hospital admissions (0.9 per patient per year vs. 1.4 per patient per year), length of hospitalization, and ICU admissions (0.2 per patient per year vs. 0.4 per patient per year).
Struik, 2014 <sup>64</sup> RCT	COPD	Inclusion: Severe COPD (GOLD stage 3 and 4), >48 hours independence from ventilatory support (invasive or non-invasive) for	1) BPAP ST	12 months	There was no significant difference between the BPAP ST group and the Standard Care group on mortality (30 vs. 29), survival time (mean: 299 days vs.

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
		ARF, prolonged hypercapnia(PaCO <sub>2</sub> >6.0 kPa) during daytime at rest without oxygen or ventilatory support	2) Standard care		291 days, p=0.99), number of hospital admissions (1.0 per person per year vs. 1.0 per person per year), number of patients with hospital readmissions due to respiratory causes (56% vs. 57%), length of hospitalization (7.0 days vs. 3.5 days, p=0.09), annual number of exacerbations at home (median: 1.0 vs. 2.0, p=0.26), quality of life (measured by Chronic Respiratory Questionnaire, 0.01, 95% CI: -0.4 to 0.4), dyspnea scale (measured by MRC dyspnea, -0.05, 95% CI: -0.6 to 0.5), and activity of daily living (measured by Groninger Activity Restriction Scale, 0.4, 95% CI: -2.3 to 3.0).
Duraio, 2018 <sup>25</sup> Observational	COPD	Inclusion: COPD NOS  Exclusion: No clinical assessment in prior 6 months, OSA with a history	1) HMV/BPAP mix started in AECOPD	>1 year	There were no difference on number of hospital admission for respiratory causes (changes before and after NIPPV per year: -0.6 vs. -0.3, p=0.46)

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
		of noncompliance with CPAP	2) HMV/BPAP mix started in stable disease		and length of hospital stay for respiratory causes (changes before and after NIPPV per year: -9.8 days vs. -1.7 days, p=0.09).
Duiverman, 2017 <sup>24</sup> RCT	COPD	Inclusion: COPD (GOLD III or IV), ≥ 2 AECOPD with acute hypercapnic respiratory failure (pH<7.35) per year, daytime Inclusion: PaCO <sub>2</sub> ≥6.7 kPa (50 mmHg) or nocturnal PaCO <sub>2</sub> ≥7.3 kPa (55 mmHg) or nighttime rise in PtCO <sub>2</sub> ≥1.3 kPa (10 mmHg), stable (no AECOPD in prior 4 weeks, pH>7.35).	1) HMV/BPAP mix (pressure controlled ventilation) (high intensity)	1.5 months	There was no statistical difference between two groups on quality of life (the COPD assessment test, WMD: 2.30, 95% CI: -2.35 to 6.95).
			2) HMV (pressure controlled ventilation) (low intensity)		
Oscroft, 2010 <sup>49</sup>	COPD	COPD, FEV <sub>1</sub> <50%,	1) BPAP (pressure controlled ventilation)	6 months	There was no significant difference

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
RCT		FEV1/FVC<70%, TLC>80%, >20 pack year smoking history, pH 7.35-7.45, PaCO2>7.5 kPa or PtcCo2>9kPa, treated with NIPPV for at least 3 months with compliance at least 4 hours/day, clinical stability (no increased breathlessness, cough or sputum in the prior 4 weeks, no increase in PaCO2 and no decrease in FEV1 since study initiation)	2) No device		between the two groups on quality of life (St. Georges Respiratory Questionnaire, p=0.10).

Note: ± denotes standard deviation.

AECOPD: acute exacerbation of chronic obstructive pulmonary disease, AHRF: acute hypoxemic respiratory failure, ATS: American Thoracic Society, BPAP: Bilevel Positive Airway Pressure, CI: confidence interval, COPD; chronic obstructive pulmonary disease, CPAP: Continuous Positive Airway Pressure, CRF: Chronic Respiratory Failure, BMI: Body Mass Index, FEV1: Forced expiratory volume in one second, FVC: Forced vital capacity, HF: heart failure, HMV: Home Mechanical Ventilation, HR: hazard ratio, ICU: Intensive Care Unit, IVAPS: intelligent volume assured pressure support, kPa: kilopascal, LTOT: long term oxygen therapy, mmHg: millimeters of mercury (pressure), NIPPV: Noninvasive positive pressure ventilation, NMD: Neuromuscular Disease, NOS: Not otherwise Specified, OHS: Obesity hypoventilation syndrome, OR: odds ratio, OSA: Obstructive sleep apnea, PaO2: partial pressure of arterial oxygen, PaCO2: partial pressure of arterial carbon dioxide, PAP: positive airway pressure, pH: potential of hydrogen, PSV: Pressure support ventilation, PtcCO2: transcutaneous pressure of carbon dioxide, RCT: randomized controlled trial, REM: rapid eye movement, RF: Respiratory Failure, S: spontaneous mode, SaO2: arterial blood oxygen saturation, SF-36: Medical Outcomes Study Questionnaire Short Form, SpO2: peripheral capillary oxygen saturation, ST: spontaneous/timed breath mode, TLC: total lung capacity, WMD: weighted mean difference

**Table F.13. Thoracic Restrictive Disorders – Effectiveness of home devices**

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group) (add n per arm)	Duration Of Device Use In Home Setting	Conclusion
Buyse, 2003 <sup>9</sup> Observational	TRD	Inclusion: Kyphoscoliosis with respiratory insufficiency who started LTOT and/or NIPPV.	1) HMV (volume cycled or pressure cycled) + oxygen	10 months	Survival rate was significantly higher in patients treated with HMV plus long-term oxygen than patients with long-term oxygen alone (p<0.05)
			2) Oxygen alone		
Schonhofer, 2001 <sup>61</sup> Observational	TRD	Inclusion: TRD (post-TB or scoliosis); PaCO <sub>2</sub> 45-55 mmHg; stable PaCO <sub>2</sub> compared to baseline; stable disease (no hospital admission 1 month prior)  Exclusion: Rapidly progressive NMD, OHS, COPD, acute RF, severe acidosis.	1) Mixed: HMV (volume assist control ventilation) with change to BPAP ST if not tolerated	3 months	HMV: significant improvements before and after 3-month treatment in inspiratory threshold loading test (278%), cycle ergometer test (176%), and shuttle walking test (32%). Standard care: no significant changes before and after 3-month treatment.
			2) Standard care without HMV/BPAP device		

COPD: chronic obstructive pulmonary disease, HMV: Home Mechanical Ventilation, LTOT: long term oxygen therapy, mmHg: millimeters of mercury (pressure), NIPPV: Noninvasive Positive Pressure Ventilation, NMD: Neuromuscular Disease, NOS: Not otherwise Specified, OHS: obesity hypoventilation syndrome, PaCO<sub>2</sub>: partial pressure of arterial carbon dioxide, RF: Respiratory Failure, TB: tuberculosis, TRD: thoracic restrictive disorder

**Table F.14. Neuromuscular Disorder – Effectiveness of Home Devices**

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
Sanjuan-López, 2014 <sup>60</sup> Observational	NMD	<p>Inclusion: ALS; hospital admission; chronic RF by pulmonologist</p> <p>Exclusion: Neuromuscular processes other than ALS, treatment in social welfare palliative center.</p>	1) HMV (pressure support ventilation mode or BPAP ST mode) started after outpatient pulmonary evaluation	23.3 months (95% CI, 16.7–28.8)	<p>Patients received HMV after pulmonary evaluation have longer length of survival than those without pulmonary evaluation (mean survival: 12.3 months vs. 2.8 months, p&lt;0.004).</p>
			2) HMV (pressure support ventilation mode or BPAP ST mode) started in an emergency situation without prior outpatient pulmonary evaluation	26.7 months	
Pinto, 2010 <sup>54</sup> Observational	NMD	<p>Inclusion: ALS; home BPAP use; FVC ≥75%; PaO2 ≥80 mmHg; PaCO2 ≤ 45 mmHg; age 18-75 years</p> <p>Exclusion: Gastrostomy, cognitive impairment, other significant disorders.</p>	1) BPAP ST + Weekly telemonitoring + Standard care	36 months	<p>The BPAP ST + weekly telemonitoring group had significantly lower number of office visits (IRR: 0.34, 95% CI: 0.29 to 0.38); ER visits (IRR: 0.19; 95% CI: 0.10 to 0.37); hospital admission (IRR: 0.17; 95% CI: 0.07 to 0.41). There was no significant difference on mortality (OR= 1.00; 95% CI: 0.24 to 4.18) or median survival time (from BPAP adoption to death) (865 days vs. 334 days, p=0.13)</p>
			2) BPAP ST + Standard care		
Gonzalez-Bermejo, 2013 <sup>32</sup> Observational	NMD	<p>Inclusion: ALS on home BPAP with 4 hour/night minimal adherence</p> <p>Exclusion: Use of other ventilator types, without integrated SpO2</p>	1) BPAP ST "correctly ventilated patients"	12 months	<p>The "correctly ventilated" patients had significantly lower mortality than those "insufficiently ventilated" patients (OR= 0.25; 95% CI: 0.10 to 0.64).</p>
			2) BPAP ST "insufficiently ventilated patients"	12 months	

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
		monitoring.			
Sancho, 2014 <sup>58</sup> Observational	NMD	<p>Inclusion: ALS; symptoms (fatigue, dyspnea, orthopnea, morning headache) plus one of the following 1) PaCO<sub>2</sub> &gt;45 mmHg or 2) FVC &lt;50% or 3) MIP &lt;60 cm H<sub>2</sub>O or 4) SaO<sub>2</sub> &lt; 88% for ≥ 5 consecutive minutes by nocturnal oximetry</p> <p>Exclusion: Presence of previous pulmonary/airway disease, rapidly progressing disease w/ survival expectancy &lt;1 month, severe frontotemporal dementia, NIPPV tolerance &lt;4 consecutive hour/night.</p>	<p>1) HMV (volume assist control ventilation)</p> <hr/> <p>2) BPAP ST</p>	15 months	No significant difference was found on length of survival (median 15.00 months (95% CI: 7.48 to 22.41) vs. median 15.00 months (95% CI; 95% CI 10.25 to 19.75), p=0.53)
Sivori, 2007 <sup>63</sup> Observational	NMD	<p>Inclusion: ALS; symptomatic ventilatory impairment (dyspnea, morning headache, fatigue) plus 1) PaCo<sub>2</sub> &gt; 45 mmHg or 2) nocturnal oxygen saturation by pulse</p>	<p>1)BPAP, Riluzole</p> <hr/> <p>2) BPAP NOS</p> <hr/> <p>3) No BPAP, No Riluzole</p>	Longest Duration: 60 months	With a 30-month followup, 9 out of 11 patients died in the BPAP group; while 42 out of 42 patients in the no BPAP group (OR=0.04, 95% CI: 0.00 to 1.01, p=0.05).

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
		oximeter $\leq$ 88% for 5 continuous minutes or 3) MIP < 60 cmH <sub>2</sub> O or 4) FVC < 50%.			
Coco, 2006 <sup>17</sup> Observational	NMD	Inclusion: ALS; symptomatic ventilatory impairment (dyspnea, morning headache, hypersomnolence, fatigue) plus 1) PaCO <sub>2</sub> $\geq$ 45 mmHg or 2) nocturnal oxygen saturation by pulse oximeter $\leq$ 88% for 5 continuous minutes or 3) MIP < 60 cmH <sub>2</sub> O or 4) FVC < 50%  Exclusion: Primary lateral sclerosis, diagnosis other than ALS during followup.	1) BPAP ST (use $\geq$ 4 hours/day)	Longest Duration: 30 months	The group with $\geq$ 4 hours/days use had significantly longer survival time from BPAP start to death (median: 18 months (interquartile range: 7 to 28) vs. 6 months (interquartile range: 3 to 12), p<0.001). No patient was lost to followup
			2) BPAP ST < 4 hours/day)		
Bourke, 2006 <sup>7</sup> RCT	NMD	Inclusion: ALS; orthopnea with Pimax <60% or symptomatic daytime hypercapnia	1) BPAP ST (full cohort)	12 months	Patients with BPAP were also found to have better median survival length (216 days vs. 11 days, p=0.01) and quality of



Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
		Exclusion: Current or previous NIPPV use, significant co-morbidities, age>75 years, inability to complete quality of life assessment	2) no BPAP ST (full cohort)		life measured by SF-36 mental components (168 vs. 99, p<0.01) and physical component (150 vs. 81, p<0.01).
Pinto, 1995 <sup>53</sup> Observational	NMD	Inclusion: ALS; bulbar features  Exclusion: Tracheotomised, refusal of attempts to prolong survival.	1) BPAP NOS	Longest Duration: 42 months	With a 3-year followup, patients treated with BPAP were found to have significantly higher overall survival than patients with palliative management (p=0.004).
			2) No BPAP NOS		
Vitacca, 2017 <sup>68</sup> Observational	NMD	Inclusion: ALS NOS admitted to hospital, NIPPV use  Exclusion: dementia confirmed by Mini-Mental State Examination score <20, refusal of NIPPV	1) HMV/BPAP mix started in FVC≥ 80% (early)	36 months	The patients started in FVC≥ 80% (early) were found to have significantly longer survival time (31.33 months vs. 27.51 months, p=0.01) and lower mortality (HR: 0.46, 95% CI: 0.29 to 0.74; p=0.001) than the patients started in FVC <80% (late).
			2) HMV/BPAP mix started in FVC <80% (late)		
Sancho, 2018 <sup>59</sup> Observational	NMD	Inclusion: ALS (Escorial criteria), hospital admission  Exclusion: lung disease, <1 year life expectancy, NIV use <4 consecutive hours/night, slow disease progression (>3	1) HMV (volume assist control ventilation)	Longest Duration: 36 months	The HMV group had significantly longer survival time than the group not treated with any device (mean: 18.50 months vs. 3.00 months, p=0.001). The significant difference was also found in patients with no or moderate bulbar dysfunction (mean:
			2) No device		

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
		yrs), severe frontotemporal dementia			20.00 months vs. 3.00 months, p=0.0001) and in patients with severe bulbar dysfunction (mean: 13.00 months vs. 3.00 months, p=0.001).
Bertella, 2017 <sup>3</sup> RCT	NMD	Inclusion: ALS (definite via El Escorial Criteria), stable disease (no respiratory infection in prior 3 months)  Exclusion: cognitive impairment, severe comorbidity, contraindications to NIV, distance from hospital >40 km.	1) BPAP volume assured pressure support ventilation outpatient initiation 2) BPAP volume assured pressure support ventilation inpatient initiation	3 months	There was no statistically significant difference on dyspnea (measured by VAS score), sleep quality (measured by VAS score). No adverse events were reported in both groups.
Aboussouan, 1997 <sup>1</sup> Observational	NMD	Inclusion: ALS via el Escorial criteria; dyspnea on exertion or PaCO <sub>2</sub> ≥ 45 mmHg or orthopnea or FVC < 60% predicted.	1) HMV/BPAP mix tolerant 2) HMV/BPAP mix intolerant	Longest Duration: 25 months	The intolerant patients had significantly higher mortality than the tolerant patients (OR: 20.00, 95% CI: 2.19 to 182.44, p<0.01).
Farrero, 2005 <sup>26</sup> Observational	NMD	ALS NOS	1) HMV/BPAP mix in pre-protocol group 2) HMV/BPAP mix in post-protocol group	Longest Duration: 48 months	No significant difference on survival time was observed between the two groups (p=0.84).

ALS: amyotrophic lateral sclerosis, BPAP: Bilevel Positive Airway Pressure, CI: confidence interval, cmH<sub>2</sub>O: centimeters of water (pressure), CPAP: Continuous Positive Airway Pressure, BMI: Body Mass Index, FEV<sub>1</sub>: Forced expiratory volume in one second, FVC: Forced vital capacity, HMV: Home Mechanical Ventilation, HR: hazard ratio, IRR: incidence rate ratio, MIP: maximum inspiratory pressure, mmHg: millimeters of mercury (pressure), NIV: noninvasive ventilation, NIPPV: Noninvasive Positive Pressure Ventilation, NMD: Neuromuscular Disease, NOS: Not otherwise Specified, OR: odds ratio, PaO<sub>2</sub>: partial pressure of arterial oxygen, PaCO<sub>2</sub>: partial pressure of arterial carbon dioxide, Pimax: maximal inspiratory mouth pressures, PSV: Pressure support ventilation, RCT: randomized controlled trial, RF: Respiratory Failure, SaO<sub>2</sub>: arterial blood oxygen saturation, SF-36: Medical Outcomes Study Questionnaire Short Form, SpO<sub>2</sub>: peripheral capillary oxygen saturation, ST: spontaneous/timed breath mode, VAS: visual analog scale

**Table F.15. Obesity Hypoventilation Syndrome – Effectiveness of home devices**

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group) (add n per arm)	Duration Of Device Use In Home Setting	Conclusion
Howard, 2016 <sup>36</sup> RCT	OHS	Inclusion: OHS (BMI >30, daytime PaCO <sub>2</sub> >45 mmHg)  Exclusion: Other conditions contributing to hypoventilation.	1) BPAP ST	3 months	No significant difference was found between groups on Epworth Sleepiness Scale scores (p=0.86), SF-36 Physical Component (p=0.37), SF-36 mental Component (p=0.57), Severe Respiratory Insufficiency Questionnaire (p=0.54) and physical activities (sedentary time awake min/day, moderate-to-vigorous physical activity, steps/day).
			2) CPAP	3 months	
Masa, 2015 <sup>40</sup> , <sup>41</sup> RCT	OHS	Inclusion: OHS (BMI ≥ 30; stable PaCO <sub>2</sub> ≥ 45 mmHg; pH ≥ 7.35; no clinical worsening in prior 2 months); severe OSA (apnea-hypopnea index ≥30); correctly executed 30min CPAP/NIPPV treatment test; age 15-80 years  Exclusion: COPD (FEV <sub>1</sub> /FVC <70%), NMD, narcolepsy, restless legs syndrome, psychophysical,	1) HMV/BPAP mix (all with bilevel pressure with assured volume) + lifestyle modification	2 months	The HMV/BPAP group and the CPAP group reported significantly better sleep quality measured by Epworth Sleepiness Scale than the lifestyle modification group. No significant difference between the HMV/BPAP and CPAP group. Patients treated by HMV/BPAP were found to have significant better outcomes on 6-minute walk distance test than CPAP (p=0.01). There was no difference between
			2) CPAP + Lifestyle modification	2 months	
			3) Lifestyle modification	2 months	

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group) (add n per arm)	Duration Of Device Use In Home Setting	Conclusion
		severe chronic debilitating illness, severe chronic nasal obstruction.			groups on quality of life and number of dropouts.
Borel, 2011 <sup>6</sup> , RCT	OHS	Inclusion: OHS (BMI >30; daytime PaCO <sub>2</sub> ≥ 45 mmHg); age 20-75 years  Exclusion: Declined or presented any significant airway obstruction, scoliosis, cardiac failure, progressive NMD.	1) BPAP ST	Longest Duration: 1 month	No significant difference were found on sleep quality measured by Epworth Sleepiness Scale (p=0.49).
			2) Lifestyle counseling		
Murphy, 2012 <sup>45</sup> , RCT	OHS	Inclusion: OHS (BMI>40, daytime chronic PaCO <sub>2</sub> >6 kPa, pH >7.35), absence of other identifiable hypoventilation cause, FEV1/FVC >70%, FVC <70%  Exclusion: Inability to provide written consent.	1) BPAP AVAPS	Longest Duration: 3 months	There was no statistically significant difference on quality of life (Severe Respiratory Insufficiency Questionnaire summary score, mean difference: 5, p=0.21), sleep quality (Epworth Sleepiness Score; 1, p=0.43).
			2) BPAP ST	Longest Duration: 3 months	
Piper, 2008 <sup>55</sup> , RCT	OHS	Inclusion: OHS (BMI≥30, PaCO <sub>2</sub> ≥ 45 mmHg (awake, stable), absence of another cause for hypercapnia,	1) CPAP	Longest Duration: 3 months	No significant difference was found between the groups on Epworth Sleepiness Scale (p=0.59),

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group) (add n per arm)	Duration Of Device Use In Home Setting	Conclusion
		FEV1/FVC $\geq$ 70%)  Exclusion: psychiatric illness, current home NIPPV use, PtcCO2 during REM $\geq$ 10mmHg, increase in afternoon to morning PaCO2 $\geq$ 10mmHg in patients with awake PaCO2 >55 mmHg.	2) BPAP S		SF-36 Physical Component (p=0.22), and SF-36 mental Component (p=0.28).
Masa, 2016, <sup>42</sup> RCT	OHS	Inclusion: OHS (BMI $\geq$ 30 kg/m <sup>2</sup> , no COPD, no NMD, no TRD, no narcooepsy, no restless leg syndrome), stable hypercapnic respiratory failure (daytime awake PaCO2 $\geq$ 45 mmHg, pH $\geq$ 7.35 and no clinical worsening in prior 2 months), ability to use NIPPV in 30 minute trial period  Exclusion: Psychophysical inability to complete questionnaires, severe chronic debilitating illness, severe chronic nasal obstruction, lack of informed	1) BPAP  2) Lifestyle modification	2 months	Patients in the BPAP group had significantly better improvements on Epworth Sleepiness Scale (p=0.02) and SF-36 Mental Component (p=0.04) than those in the lifestyle modification group. There was no significant difference on 6-minute walk distance test and SF-36 Physical Component.

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group) (add n per arm)	Duration Of Device Use In Home Setting	Conclusion
		consent.			
Perez de Llano, 2005 <sup>52</sup> Observational	OHS	Inclusion: OHS, BMI > 30, PaCO <sub>2</sub> ≥ 50 mmHg, FEV <sub>1</sub> /FVC < 70%, absence of other respiratory disorder such as kyphoscoliosis or diaphragmatic paralysis	1) HMV/BPAP mix 2) no device	Longest duration: 105 months	Patients treated without any device had significantly higher mortality rate (OR= 14.88, 95% CI: 3.18 to 69.68, p= 0.001) than those treated by HMV/BPAP mix.
Priou, 2010 <sup>56</sup> Observational	OHS	Inclusions: BMI ≥ 30 kg/m <sup>2</sup> and daytime hypercapnia (PaCO <sub>2</sub> > 45 mm Hg) in the absence of any other cause of hypoventilation on the basis of clinical examination, chest radiograph, and pulmonary function tests (eg, COPD [FEV <sub>1</sub> to vital capacity ratio , 70%]).	1) BPAP in acute exacerbation 2) BPAP in stable hypercapnia	50 months	There was no significant difference on mortality rate (OR= 1.27, 95% CI:0.49 to 3.27, p=0.63).

AVAPS: average volume assured pressure support, BPAP: Bilevel Positive Airway Pressure, COPD: chronic obstructive pulmonary disease, CPAP: Continuous Positive Airway Pressure, BMI: Body Mass Index, FEV<sub>1</sub>: Forced expiratory volume in one second, FVC: Forced vital capacity, HMV: Home Mechanical Ventilation, kPa: kilopascal, LTOT: long term oxygen therapy, mmHg: millimeters of mercury (pressure), NIPPV: Noninvasive Positive Pressure Ventilation, NMD: Neuromuscular Disease, NOS: Not otherwise Specified, OHS: obesity hypoventilation syndrome, OSA: Obstructive sleep apnea, PaCO<sub>2</sub>: partial pressure of arterial carbon dioxide, pH: potential of hydrogen, RCT: randomized controlled trial, REM: Rapid eye movement, RF: Respiratory Failure, S: Spontaneous mode, SF-36: Medical Outcomes Study Questionnaire Short Form, ST: spontaneous/timed breath mode, PtcCO<sub>2</sub>: transcutaneous pressure of carbon dioxide

**Table F.16. Other Respiratory Diseases – Effectiveness of home devices**

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
Benhamou, 1997 <sup>2</sup> , Observational	Other	Inclusion: Bronchiectasis; home nasal mask ventilation; LTOT.	1) HMV (volume assist control ventilation) + Oxygen	Longest Duration: 89 months	No significant difference was found on survival between the HMV and oxygen therapy group and the oxygen therapy group (median 45 months vs. 48 months, p>0.05).
			2) Oxygen alone		

HMV: Home Mechanical Ventilation, LTOT: long term oxygen therapy

**Table F.17. Mixed Diseases – Effectiveness of Home Devices**

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
Hazenberg, 2014 <sup>33</sup> , RCT	NMD, TRD	Inclusion: NMD or thoracic cage disorder; PaCO <sub>2</sub> >45 mmHg with respiratory symptoms  Exclusion: COPD, not mask naïve, acute RF, age < 18 years, invasive ventilation, nursing home resident.	1) HMV started at home pressure controlled ventilation with change to volume assist control ventilation if not tolerated	Longest Duration: 6 months	Compared to HMV started in the hospital, HMV started at home was not significantly better on mortality (OR=2.80, 95% CI: 0.51 to 15.43), withdrawals (OR= 1.03, 95% CI: 0.34 to 3.11), quality of life (Severe Respiratory Insufficiency, SF-36).
			2) HMV started in the hospital pressure controlled ventilation) with change to volume assist control ventilation if not tolerated	Longest Duration: 6 months	
Munoz, 2005 <sup>44</sup> , Observational	NMD, TRD	Inclusion: Hospital admission with CHRF secondary to NMD (ALS excluded) or kyphoscoliosis or post TB sequelae; PaCO <sub>2</sub> > 45 mmHg; HMV	1) HMV volume assist control ventilation	Longest Duration: 12 months	There was no statistically significant difference on mortality (OR= 0.91, 95% CI: 0.28 to 2.96, p=0.88), or number of hospital admissions (0.17 per patient in HMV volume assist/control
			2) HMV volume control	Longest Duration: 3 months	

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
		started in stable phase of disease  Exclusion: BiPAP users, ALS.			mode vs. 0.04 per patient in HMV volume control mode, p=0.11).  Adverse events were similar between the two groups.
Chiang, 2003 <sup>13</sup> , RCT	COPD, other	Inclusion: COPD or asthma or bronchiectasis; hospital readmission due to respiratory cause; daytime sleepiness or morning headache; PaCO <sub>2</sub> > 50 mmHg (daytime rest); SpO <sub>2</sub> < 88% for more than 5 consecutive minutes while on usual oxygen during polysomnography  Exclusion: Unable to tolerate nocturnal nasal positive pressure ventilation, OSA, unable to perform 6-minute walk distance test due to other disease.	1) BPAP NOS	6 months	Patients in the BPAP group was found to have significantly better outcomes on 6-minute walk distance test (WMD: 99.80; 95% CI: 34.14 to 165.46; p<0.01), number of hospitalization per patient (WMD: -2.30; 95% CI: -3.36 to -1.24; p<0.001), and length of hospital stay (WMD: -37.70; 95% CI: -57.68 to -17.72; p<0.001). There was no statistical difference between the two groups on resting Borg score and Borg score at end of 6-minute walk distance test.
			2) No BPAP NOS		

ALS: amyotrophic lateral sclerosis, BPAP: Bilevel Positive Airway Pressure, CHRF: chronic hypercapnic respiratory failure, CI: confidence interval, COPD: chronic obstructive pulmonary disease, mmHg: millimeters of mercury (pressure), NMD: Neuromuscular Disease, NOS: Not otherwise Specified, OR: odds ratio, OSA: Obstructive sleep apnea, PaCO<sub>2</sub>: partial pressure of arterial carbon dioxide, RCT: randomized controlled trial, RF: Respiratory Failure, SF-36: Medical Outcomes Study Questionnaire Short Form, SpO<sub>2</sub>: peripheral capillary oxygen saturation, TB: tuberculosis, TRD: thoracic restrictive disorder, WMD: weighted mean difference



### KQ3. What are the equipment parameters that are used in each of the above groups?

**Table F.18. COPD – Equipment parameters**

Author, Year, Study Design	Device/mode	Model (Manufacturer; Location of Manufacturer)	Device Characteristics	Prescribed Usage (frequency and duration)	Actual Usage
Vasquez, 2017 <sup>67</sup> Observational	BPAP NOS	NR	IPAP, EPAP	NR	NR
	CPAP NOS	NR	CPAP	NR	NR
	HMV NOS	NR	NR	NR	NR
Murphy, 2017 <sup>46</sup> RCT	BPAP ST	Harmony (Philips Respironics; USA)  VPAP III STa (ResMed; Bella Vista, Australia)	IPAP, EPAP, rate	≥ 6 hours nightly	-4.7 (2.5-5.6) hours/day (6 weeks) -7.6(3.6-8.4) hours/day (12 months). -IPAP: 24 (22-26) cm H2O -EPAP: 4 (4-5) cmH2O -Rate: 14 (14-16) breaths/minute
Salturk, 2015 <sup>57</sup> Observational	BPAP ST	NR	IPAP, EPAP, rate	NR	-6.7 ± 1.9 hours/day -IPAP: 24 ± 3 cm H2O -EPAP: 5.3 ± 0.7 cmH2O
Oscroft, 2014 <sup>50</sup> RCT	BPAP volume assured pressure support ventilation	Intelligent volume assured pressure support (iVAPS) (ResMed; Bella Vista, Australia)	IPAP, EPAP, rate, target minute ventilation	NR	-Target minute ventilation 8.4 [5.7-9.8] L/minute -EPAP: 4 (4-4) cmH2O -Rate: 15 (13.3-19.4) breaths/minute
	BPAP ST	NIPPY 3 (B and D Electromedical; Stratford, United Kingdom)	IPAP, EPAP, rate	NR	-IPAP: 28 (27.3-30) cmH2O -EPAP: 5 (5-5) cmH2O -Rate: 15.0 (15-15) breaths/minute
Paone, 2014 <sup>51</sup> Observational	BPAP ST	Synchrony (Philips Respironics; Andover, MA)  Neftis (Linde; Munich, Germany)	IPAP, EPAP, rate	NR	-IPAP: 18.5 ± 2.66 cm H2O -EPAP: 3.9 ± 1 cm H2O -Rate: 12 breaths/minute
Galli, 2014 <sup>29</sup> Observational	BPAP NOS	NR	IPAP, EPAP	NR	-IPAP: 22.1 ± 6.2 cm H2O -EPAP: 5.9 ± 1.8 cm H2O
Bhatt, 2013 <sup>4</sup>	BPAP NOS	BiPAP Synchrony	IPAP, EPAP	≥ 6 hours daily for 6 months	-IPAP: 15 cm H2O

Author, Year, Study Design	Device/mode	Model (Manufacturer; Location of Manufacturer)	Device Characteristics	Prescribed Usage (frequency and duration)	Actual Usage
RCT		(Respironics Inc.; Murrysville, USA)			-EPAP: 5 cm H <sub>2</sub> O
Duiverman, 2011 <sup>22, 23</sup> RCT	BPAP ST	BiPAP Synchrony (Respironics Inc.; Murrysville, USA)	IPAP, EPAP, rate	NR	Followup #1: -IPAP: 23 ± 4 cm H <sub>2</sub> O -EPAP: 6 ± 2 cm H <sub>2</sub> O -Rate: 18(3) breaths/minute  Followup #2: -7.7 (5.8-8.5) hours/day -IPAP: 20 ± 4 cm H <sub>2</sub> O -EPAP: 6 ± 2 cm H <sub>2</sub> O -Rate: 18 ± 3 breaths/minute
Oscroft, 2010 <sup>48</sup> Observational	BPAP ST	NIPPY I, 2 or 3 (B & D Electromedical; Stratford, United Kingdom)	IPAP, EPAP, rate	NR	-IPAP: 26 ± 3 cm H <sub>2</sub> O -EPAP: 4 ± 1 cm H <sub>2</sub> O -Short inspiratory (0.8- 1 s) -Long expiratory time (2.5-3.5 s).
Cheung, 2010 <sup>12</sup> RCT	CPAP	NR	CPAP	>8 hours nightly for 12 months	NR
	BPAP ST	NR	IPAP, EPAP, rate	>8 hours nightly for 12 months	-7-9 hours/night -IPAP: 14.8 ± 1.1 cm H <sub>2</sub> O -EPAP: 5 ± 0 cm H <sub>2</sub> O
Hitzl, 2009 <sup>35</sup> Observational	HMV (pressure controlled ventilation)	NR	inspiratory pressure, PEEP, inspiratory time, rate	NR	-IPAP: 20.9 ± 4.0 cm H <sub>2</sub> O -EPAP: 4.2 ± 1.9 cm H <sub>2</sub> O -Rate: 19.1 ± 3.8 breaths/minute
McEvoy, 2009 <sup>43</sup> RCT	BPAP S	VPAP S mode (ResMed; Sydney, Australia)	IPAP, EPAP	NR	-4.5 (3.2) hours/day -IPAP: 12.9 (12.5-13) cm H <sub>2</sub> O -EPAP: 5.1 (4.8-5.3) cm H <sub>2</sub> O
Windisch, 2006 <sup>69</sup> Observational	HMV (pressure controlled ventilation)	PV401 (Breas Medical AB; Moelnlycke, Sweden)	inspiratory pressure, PEEP, inspiratory time, rate	NR	-IPAP: 31.0 ± 6.6 mbar -Rate: 20.7 ± 2.1 breaths/minute -Inspiratory time 1.0 ±

Author, Year, Study Design	Device/mode	Model (Manufacturer; Location of Manufacturer)	Device Characteristics	Prescribed Usage (frequency and duration)	Actual Usage
					0.1 seconds
Casanova, 2000 <sup>10</sup> RCT	BPAP S	DP-90 (Taema; Paris, France)	IPAP, EPAP	NR	-6.2 hours/day (at 3 months) -5.9 hours/day (at 6 months) -IPAP: 12 ± 2 cm H2O
Garrod, 2000 <sup>30</sup> RCT	BPAP S	BiPAP ST 30 (Respironics Inc.; Murrysville, USA)	IPAP, EPAP	≥ 8 hours daily	-IPAP: 16 (13-24) cm H2O -EPAP: 4 (4-6) cm H2O
Clini, 1998 <sup>15</sup> Observational	BPAP ST	BiPAP (Respironics Inc. ; Murrysville, USA)	IPAP, EPAP, rate	NR	-7.4 ± 1.3 hours/day -IPAP: 10-16 cm H2O -EPAP: 2-4 cm H2O
Clini, 1996 <sup>14</sup> Observational	BPAP ST	BiPAP (Respironics Inc. ; Murrysville, USA)	IPAP, EPAP, rate	NR	NR
Zhou, 2017 RCT	BPAP ST	Flexo ST 30 NIV (Curative Co.; SuZhou, China)	IPAP, EPAP, rate	NR	-5.6 ± 1.4 hours/day -IPAP: 17.8 ± 2.08 cm H2O -EPAP: 4.2 ± 0.1 cm H2O
Marquez-Martin, 2014 <sup>38</sup> RCT	BPAP ST	BiPAP (Respironics Inc. ; Murrysville, USA)	IPAP, EPAP, rate	NR	-7 (6.5-9) hours nightly -IPAP: 16 cm H2O (median) (both NIPPV groups) -EPAP: 4 cm H2O (median, both NIPPV groups)
Köhnlein, 2014 <sup>37</sup> RCT	BPAP ST	Models not reported. Manufacturers: ResMed (Martinsried, Germany), Weinmann (Hamburg, Germany, or Tyco Healthcare (Neubrug, Germany)	IPAP, EPAP, rate	≥ 6 hours daily	-IPAP: 21.6 ± 4.7 cm H2O -EPAP: 4.8 ± 1.6 cm H2O -Rate: 16.1 ± 3.6 breaths/minute -Ventilator use measured in 48 (47%) of patients. In these 48 patients, 65% exceeded the prescribed usage of ≥ 6 hours daily)
De Backer, 2011 <sup>19</sup>	BPAP NOS	BiPAP Synchrony	IPAP, EPAP	> 5 hours daily	NR

Author, Year, Study Design	Device/mode	Model (Manufacturer; Location of Manufacturer)	Device Characteristics	Prescribed Usage (frequency and duration)	Actual Usage
RCT		(Respironics Inc., Murrysville, USA)			
Funk, 2010 <sup>27</sup> RCT	BPAP NOS	NR	IPAP, EPAP	NR	NR
Dreher, 2010 <sup>21</sup> RCT	HMV (pressure controlled ventilation)	Breas Vivo 40 (Breas Medical AB; Molnlycke, Sweden)  Smart Air (Airox; Pau Cedex, France)	inspiratory pressure, PEEP, inspiratory time, rate, inspiratory flow trigger	Entire night while sleeping and during daytime naps.	-IPAP: 28.6 ± 1.9 cm H2O -EPAP: 4.5 ± 0.7 cm H2O -Rate: 17.5 ± 0.7 breaths/minute
	HMV (pressure support ventilation)	Breas Vivo 40 (Breas Medical AB; Molnlycke, Sweden)  Smart Air (Airox; Pau Cedex, France)	inspiratory pressure, PEEP, inspiratory flow trigger, expiratory flow trigger	Use the entire night while sleeping and during daytime naps.	-IPAP: 14.6 ± 0.8 cm H2O -EPAP: 4 ± 0 cm H2O -Rate: 8.0 ± 0 breaths/minute
Tsolaki, 2008 <sup>65</sup> Observational	BPAP ST	VPAP III ST (ResMed; Sydney, Australia)	IPAP, EPAP, rate	≥ 5 hours daily	-9 ± 2.2 hours/day -IPAP: 15.3 ± 2 cm H2O -EPAP: 5.4 ± 0.7 (4-8) cm H2O
Gay, 1996 <sup>31</sup> RCT	BPAP ST	BiPAP (Respironics Inc. ; Murrysville, USA)	IPAP, EPAP, rate	NR	-5.1 ± 3.8 hours/day
Gad, 2014 <sup>28</sup> Observational	BPAP ST	NR	IPAP, EPAP, rate	NR	-IPAP: 15.5 ± 4.2 cm H2O -EPAP: 4.0 ± 0 cm H2O -9 ± 2 hours/day
Sin, 2007 <sup>62</sup> RCT	BPAP NOS	VPAP II (ResMed; Sydney, Australia)	IPAP, EPAP	NR	NR
Heinemann, 2011 <sup>34</sup> Observational	BPAP (pressure controlled ventilation)	NR	IPAP, EPAP, rate, inspiratory time	NR	-IPAP: 22.7 ± 4.3 mbar -EPAP: 5.0 ± 1.3 mbar -Rate: 16.8 ± 3.0 breaths/minute
Budweiser, 2007 <sup>8</sup> Observational	BPAP (pressure controlled ventilation)	Twin Air (Airox Inc.; Pau, France)  Smart Air (Airox Inc.; Pau, France)	IPAP, EPAP, rate, inspiratory time	NR	-6.5 ± 2.5 hours/day -IPAP: 21.0 ± 4.0 cm H2O -EPAP: 4.5 ± 1.4 cm H2O -Rate: 17.3 ± 2.5

Author, Year, Study Design	Device/mode	Model (Manufacturer; Location of Manufacturer)	Device Characteristics	Prescribed Usage (frequency and duration)	Actual Usage
		BiPAP Synchrony (Respironics Inc.; Murrysville, USA)			breaths/minute
Clini, 2002 <sup>16</sup> RCT	BPAP ST	BiPAP ST 30 (Respironics Inc.; Murrysville, USA)	IPAP, EPAP, rate	NR	-9 ± 2 hours/day -IPAP: 14 ± 3 cm H <sub>2</sub> O -EPAP: 2 ± 1 cm H <sub>2</sub> O
Struik, 2014 <sup>64</sup> RCT	BPAP ST	BiPAP Synchrony (Respironics Inc.; Murrysville, USA)	IPAP, EPAP, rate	NR	-6.3 ± 2.4 hours/day -IPAP: 19.2 ± 3.4 cm H <sub>2</sub> O -EPAP: 4.8 ± 1.0 cm H <sub>2</sub> O -Rate: 15 ± 3 breaths/minute -Inspiratory time 1.1 ± 0.3 s
Durao, 2018 <sup>25</sup> Observational	HMV/BPAP mix. HMV mode: pressure controlled ventilation. BPAP modes: ST and volume assured pressure support ventilation	VPAP ST S9 (Resmed) VPAP ST STA (Resmed) BiPAP PR1 (Philips Respironics) BiPAP A30 (Philips Respironics) BiPAP A40 (Philips Respironics) Trilogy 100 (Philips Respironics)	IPAP, EPAP, rate, inspiratory time, target tidal volume	NR	-8.7 ± 3.6 hours/day -IPAP: 23.7 ± 5.3 cm H <sub>2</sub> O -Rate: 15.2 ± 1.4 breaths/minute
Duiverman, 2017 <sup>24</sup> RCT	HMV/BPAP mix (pressure controlled ventilation) (high intensity)	Breas Vivo 50 (Breas Medical AB; Molnlycke, Sweden) Stellar 100; Resmed (Martinsried, Germany)	inspiratory pressure, PEEP, inspiratory time, rate, inspiratory flow trigger		-4.6 (0.11-9.2) hours/day -IPAP: 23.6 ± 3.1 cm H <sub>2</sub> O -EPAP: 5.4 ± 0.9 cm H <sub>2</sub> O -Rate: 15.4 ± 0.8 breaths/minute
	HMV/BPAP mix (pressure support ventilation)	Breas Vivo 50 (Breas Medical AB; Molnlycke, Sweden)	inspiratory pressure, PEEP, inspiratory flow trigger, expiratory flow trigger		-4.2 (0.04-7.5) hours/day -IPAP: 15.5 ± 1.1 cm H <sub>2</sub> O -EPAP: 5.2 ± 0.6 cm

Author, Year, Study Design	Device/mode	Model (Manufacturer; Location of Manufacturer)	Device Characteristics	Prescribed Usage (frequency and duration)	Actual Usage
	(low intensity)	Stellar 100; Resmed (Martinsried, Germany)			H2O -Rate: 11.6 ± 1.5 breaths/minute
Blankenburg, 2017 <sup>5</sup> Observational	HMV (pressure controlled ventilation or pressure support ventilation)	VS III; ResMed (Saime SA, France)	inspiratory pressure, PEEP, inspiratory time, rate, inspiratory flow trigger	12 hours/day	-5.6 ± 4.4 hours/day -Inspiratory pressure 22 ± 3.7 cm H2O -PEEP: 2.3 ± 2.5 cm H2O -Rate: 15.8 ± 3.3 breaths/minute
Oscroft, 2010 <sup>49</sup> Observational	BPAP (pressure controlled ventilation)	NIPPY 2; B and D Electromedical (Stratford, United Kingdom)	IPAP, EPAP, rate	At least 4 hours daily	-7.4 ± 1.7 hours/day -IPAP: 30 ± 6 cm H2O -EPAP: 4 ± 1 cm H2O -Rate: 16 breaths/minute
Tsolaki, 2011 <sup>66</sup> Observational	BPAP ST	VPAP III ST (ResMed; Sydney, Australia)	IPAP, EPAP, rate	≥ 4 hours daily	-IPAP: 15.4 ± 1.9 cm H2O -EPAP: 5.4 ± 0.9 cm H2O

Note: ± denotes standard deviation. Equipment parameters not reported: mask type, supplemental oxygen, heat and moisture exchanger

BPAP: Bilevel Positive Airway Pressure, cmH2O: centimeters of water (pressure), COPD: Chronic obstructive pulmonary diseases, CPAP: Continuous Positive Airway Pressure, EPAP: Expiratory positive airway pressure, FDA: Food and Drug Administration, H2O: Hydrogen dioxide, HMV: Home Mechanical Ventilation, IPAP: inspiratory positive airway pressure, IVAPS: Intelligent volume assured pressure support, NOS: Not Otherwise Specified, NR: Not Reported, PEEP: positive end expiratory pressure. S: spontaneous mode, ST: Spontaneous/timed, USA: United States of America, VPAP: Variable positive airway pressure.

**Table F.19. Thoracic Restrictive Disorders – Equipment parameters**

Author, Year, Study Design	Device/mode	Model; Manufacturer (Location of manufacturer)	Device Characteristics	Prescribed Usage (frequency and duration)	Actual Usage
Salturk, 2015 <sup>57</sup> Observational	BPAP ST	NR	IPAP, EPAP, rate	NR	-5.9 ± 1.8 hours/day -IPAP: 22 ± 5 cm H2O -EPAP: 5.3 ± 0.6 cmH2O

Author, Year, Study Design	Device/mode	Model; Manufacturer (Location of manufacturer)	Device Characteristics	Prescribed Usage (frequency and duration)	Actual Usage
Hitzl, 2009 <sup>35</sup> Observational	HMV (pressure controlled ventilation)	NR	inspiratory pressure, PEEP, inspiratory time, rate	NR	-IPAP: 20.9 ± 4.0 cm H <sub>2</sub> O -EPAP: 4.2 ± 1.9 cm H <sub>2</sub> O -Rate: 19.1 ± 3.8 breaths/minute
Doménech-Clar, 2003 <sup>20</sup> Observational	BPAP NOS	DP-90 (Taema; Paris, France)	IPAP, EPAP	≥7 hours/night	-mean 6 hours/night
Buyse, 2003 <sup>9</sup> Observational	HMV (volume cycled or pressure cycled)	Eole 3 (Saime; Savigny-Le-Temple, France)  O'nyx (Nellcor Puritan Bennet; Villers-les-Nancy, France)	NR	NR	
Nauffal, 2002 <sup>47</sup> Observational	BPAP NOS	DP-90 (Taema; Paris, France)	IPAP, EPAP	NR	-mean 7 hours/night
Schonhofer, 2001 <sup>61</sup> Observational	Mixed: HMV (volume assist control ventilation) with change to BPAP ST if not tolerated	HMV: Drager EV 800 (Drager; Lubeck, Germany) or PLV 100 (Respironics; Murrysville, USA)  BPAP ST: BP-T (Respironics Inc.; Murrysville, USA)	HMV: tidal volume, PEEP, rate  BPAP: IPAP, EPAP, rate	NR	NR
Masa, 2000 <sup>39</sup> Observational	HMV (volume cycled or pressure cycled)	Monal DCC (Taema; Paris, France).  If could not tolerate Monal DCC, then patients were switched to a Onyx Plus (Mallinckrodt SEFAM; Nancy, France).	NR	NR	-7.3 ± 0.7 hours/day
Tsolaki, 2011 <sup>66</sup> Observational	BPAP ST	VPAP III ST (ResMed; Sydney, Australia)	IPAP, EPAP, rate	≥ 4 hours daily	-IPAP: 14.7 ± 2.4 cm H <sub>2</sub> O -EPAP: 5.0 ± 1.1 cm H <sub>2</sub> O

Note: ± denotes standard deviation.

BPAP: Bilevel Positive Airway Pressure, cm: Centimeter, EPAP: Expiratory positive airway pressure, FDA: Food and Drug Administration, H<sub>2</sub>O: Hydrogen dioxide, HMV: Home Mechanical Ventilation, IPAP: Inspiratory positive airway pressure, NOS: Not Otherwise Specified, NR: Not reported, PEEP: positive end expiratory pressure, ST: Spontaneous/timed, TRD: Thoracic Restrictive Disorder, USA: United States of America.

**Table F.20. Neuromuscular Disease – Equipment parameters**

Author, Year, Study Design	Device/mode	Model and Manufacturer	Device Characteristics	Prescribed Usage (frequency and duration)	Actual Usage
Sanjuan-López, 2014 <sup>60</sup> Observational	HMV (pressure support ventilation mode or BPAP ST mode) started after outpatient pulmonary evaluation versus HMV (pressure support ventilation mode or BPAP ST mode) started in an emergency situation without prior outpatient pulmonary evaluation	VS ultra and VS III (ResMed)	HMV device set to pressure support ventilation mode: Inspiratory pressure, PEEP, inspiratory flow trigger, expiratory flow trigger  HMV device set to BPAP ST mode: IPAP, EPAP, rate	NR	NR
Pinto, 2010 <sup>54</sup> Observational	BPAP ST + weekly telemonitoring	Goodknight 425ST bi-level device (Tyco Healthcare Group LP; California)	-IPAP, EPAP, rate -FlowSens technology (allows the physician “to customize the inspiratory and expiratory settings for greater patient comfort and synchronicity”) -Telemonitoring (wireless telemetry to remotely monitor settings and change ventilator settings and to detect alarms. “The bidirectionality of the system allowed us not only to register compliance data but also to introduce modifications in parameter settings, thus permitting real time evaluation of its impact on ventilatory mechanics.” Patients were instructed to activate the system once a week or when difficulties arose.	≥6 hours/day	NR
	BPAP ST (no telemonitoring)	Goodknight 425ST bi-level device (Tyco Healthcare Group LP; California)	-IPAP, EPAP, rate -FlowSens technology (allows the physician “to customize the inspiratory and expiratory settings for greater patient comfort and synchronicity”)	≥6 hours/day	NR
Doménech-Clar, 2003 <sup>20</sup> Observational	BPAP NOS	DP-90 (Taema; Paris, France)	IPAP, EPAP	≥7 hours/night	-Mean 6 hours/night
Nauffal, 2002 <sup>47</sup> Observational	BPAP NOS	DP-90 (Taema; Paris, France)	IPAP, EPAP	NR	-Mean 7 hours/night
Gonzalez-	BPAP ST	VPAP-III or VPAP-	IPAP, EPAP, rate	As long as possible	-IPAP: 13 ± 2 cm



Author, Year, Study Design	Device/mode	Model and Manufacturer	Device Characteristics	Prescribed Usage (frequency and duration)	Actual Usage
Bermejo, 2013 <sup>32</sup> Observational		IV plus Reslink automatic ventilatory signal analysis (Resmed; Sydney, Australia)		at night and during daytime as needed	H2O -EPAP: 5 ± 2 cm H2O -Rate: 12 ± 1 breaths/minute
Sancho, 2014 <sup>58</sup> Observational	HMV, volume assist control ventilation	PV 501 (Breas Medical; Molndal, Sweden)  Legendair (Airox; Pau, France)	Tidal volume, PEEP, rate		-Tidal volume: 782.37 ± 107.57 ml -Rate: 14.31 ± 1.14 breaths/minute
	BPAP ST	VPAP-III or VPAP-IV plus Reslink automatic ventilatory signal analysis (Resmed; Sydney, Australia)	IPAP, EPAP, rate		-IPAP: 12.01 ± 2.38 cm H2O -EPAP: 4.43 ± 1.14 cm H2O -Tidal volume: 417.84 ± 136.62 ml -Rate: 11.66 ± 0.99 breaths/minute
Sivori, 2007 <sup>63</sup> Observational	BPAP NOS	NR	IPAP, EPAP	NR	NR
Coco, 2006 <sup>17</sup> Observational	BPAP ST	BiPAP (Respironics Inc.; Vitalaire, Italy)	IPAP, EPAP, rate	Use ≥ 4 or < 4 hours/day	NR
Bourke, 2006 <sup>7</sup> RCT	BPAP ST	VPAP STII (ResMed UK Ltd; Abingdon, United Kingdom)	IPAP, EPAP, rate	NR	-Mean 9.3 hours/day (good bulbar) -Mean 3.8 hours/day (poor bulbar) -Mean IPAP 15 cmH2O -mean EPAP 4 cmH2O
Pinto, 1995 <sup>53</sup> Observational	BPAP NOS	NR	IPAP, EPAP	NR	NR
Vitacca, 2017 <sup>68</sup> Observational	HMV/BPAP mix using the following modes: ST, AVAPS,	NR	NR	≥4 hours/day and ≥120hours/month	-IPAP: 15.33 ± 3.62 cm H2O

Author, Year, Study Design	Device/mode	Model and Manufacturer	Device Characteristics	Prescribed Usage (frequency and duration)	Actual Usage
	Bi-level, volume cycled, pressure controlled ventilation				-EPAP: 5.34 ± 1.77 cm H <sub>2</sub> O -Tidal volume: 7.06 ± 1.47 ml/kg -Rate: 12.67 ± 1.46 breaths/minute
Sancho, 2017 <sup>59</sup> Observational	HMV (volume assist control ventilation)	Vivo 50; Breas Medical (Molndal, Sweden)  Trilogy 100; Philips Respironics (Madrid, Spain)	Tidal volume, PEEP, rate	≥4 hours/day	No/mild bulbar: -Tidal volume: 790.09 ± 154.41 ml -Rate: 14.5 ± 1.14 breaths/minute  Moderate/severe bulbar : -Tidal volume: 717.14 ± 124.67 ml -Rate: 14.80 ± 1.01 breaths/minute
Bertella, 2017 <sup>3</sup> RCT	BPAP volume assured pressure support ventilation	Trend II ST 30; Hoffrichter (Schwerin, Germany)  BiPAP Synchrony II, Philips Respironics (Murrysville, PA, USA)	IPAP, EPAP, rate, target minute ventilation	≥4 hours /day	Inpatient: 6.97 ± 1.05 hours/day  Outpatient: 7.68 ± 0.67 hours/day
Aboussouan, 1997, <sup>1</sup> Observational	HMV/BPAP mix	HMV: PLV-100; Life Care Products (Lafayette, Colorado, USA)  BPAP BiPAP; Respironics Inc. (Murrysville,	NR	NR	NR

Author, Year, Study Design	Device/mode	Model and Manufacturer	Device Characteristics	Prescribed Usage (frequency and duration)	Actual Usage
		Pennsylvania, USA)			
Farrero 2005, <sup>26</sup> Observational	HMV/BPAP mix	HMV: PLV-100; Life Care Products or PV 501; BREAS Medical (Gothenburg, Sweden)  BPAP: BiPAP; Respironics or VPAP ST II; Sullivan	NR	NR	NR
Tsolaki, 2011 <sup>66</sup> Observational	BPAP ST	VPAP III ST (ResMed; Sydney, Australia)	IPAP, EPAP, rate	≥ 4 hours daily	-IPAP: 14.1 ± 2.1 cm H <sub>2</sub> O -EPAP: 5.2 ± 0.4 cm H <sub>2</sub> O

Note: ± denotes standard deviation. Equipment parameters not reported: mask type, supplemental oxygen, heat and moisture exchanger

BPAP: Bilevel Positive Airway Pressure, cm: Centimeter, EPAP: Expiratory positive airway pressure, FDA: Food and Drug Administration, H<sub>2</sub>O: Hydrogen dioxide, HMV: Home Mechanical Ventilation, IPAP: Inspiratory positive airway pressure, NMD: Neuromuscular Disease, NOS: Not otherwise specified, NR: Not Reported, PEEP: positive end expiratory pressure, ST: Spontaneous/timed, USA: United States of America, VPAP: Variable positive airway pressure

**Table F.21. Obesity Hypoventilation Syndrome – Equipment parameters**

Author, Year, Study Design	Device/mode	Model and Manufacturer	Device Characteristics	Prescribed Usage (frequency and duration)	Actual Usage
Howard, 2016 <sup>36</sup> RCT	BPAP ST	NR	IPAP, EPAP, rate	NR	-5.3 (2.63) hours/night -IPAP: 19.3 ± 2.8 cm H2O -EPAP: 11.9 ± 2.3 cmH2O -Rate: 15.0 ± 2.7 breaths/minute
	CPAP	NR	CPAP	NR	-5.0(2.4) hours/night -CPAP: 15.2 ± 2.8 cm H2O
Salturk, 2015 <sup>57</sup> Observational	BPAP ST	NR	IPAP, EPAP, rate	NR	-6.4 ± 2.4 hours/day -IPAP: 23 ± 3 cm H2O -EPAP: 5.8 ± 0.8 cmH2O
Masa, 2000 <sup>39</sup> Observational	HMV (volume cycled or pressure cycled)	Monal DCC (Taema; Paris, France).  If could not tolerate Monal DCC, then patients were switched to a Onyx Plus (Mallinckrodt SEFAM; Nancy, France).	NR	NR	-7.2 ± 0.8 hours/day
Castillejo, 2014 <sup>11</sup> Observational	BPAP ST	Harmony BiPAP (Respironics; Louisville, USA)	IPAP, EPAP, rate	NR	-5.7 ± 1.3 hours/night
Masa, 2015 <sup>40, 41</sup> RCT	Mixed: HMV and BPAP mix (all with bilevel pressure with assured volume)	Breas Vivo 40 (General Electric; England)  BiPAP AVAPS (Phylips-Respironics; Netherlands)  Trilogy 100 (Philips-Respironics; Netherlands)  VS Ultra (ResMed; Australia)  Monal T50 (Air Liquide; France)  Puritan Bennett 560 (Puritan Bennett; USA)	IPAP, EPAP, rate, target minute ventilation	NR	-IPAP: at Initiation 20 ± 3.3 cm H2O; at 2 months 20 ± 3 cm H2O -EPAP: at Initiation 7.7 ± 1.8 cm H2O; at 2 months 7.8 ± 1.8 cm H2O -Rate: at Initiation 14 ± 3 breaths/minute ; at 2 months 14 ± 3.1 breaths/minute
	CPAP	NR	CPAP	NR	-CPAP: at Initiation 11 ± 2.5 cm H2O; at 2 months 11 ± 2.6 cm H2O
Borel, 2011 <sup>6</sup> RCT	BPAP ST	GoodKnight-425ST (Covidien)	IPAP, EPAP, rate	NR	-5.6 ± 2.2 hours/night -IPAP: 18 ± 3 cm H2O

Author, Year, Study Design	Device/mode	Model and Manufacturer	Device Characteristics	Prescribed Usage (frequency and duration)	Actual Usage
					-EPAP: 11 ± 2 cm H2O -Rate: 13 ± 2 breaths/minute
Murphy, 2012 <sup>45</sup> RCT	BPAP volume assured pressure support ventilation	BiPAP synchrony (Respironics Inc.; Murrysville, USA)	IPAP, EPAP, rate, target minute ventilation	NR	-EPAP: 9 ± 1 cm H2O -Tidal volume: 657 ± 96 ml -Rate: 14 ± 1 breaths/minute
	BPAP ST	BiPAP synchrony (Respironics Inc.; Murrysville, USA)	IPAP, EPAP, rate	NR	-IPAP: 25 ± 3 cm H2O -EPAP: 10 ± 2 cm H2O -Rate: 14 ± 1 breaths/minute
Piper, 2008 <sup>55</sup> RCT	BPAP S	NR	IPAP, EPAP	NR	-IPAP: 16 ± 2 cm H2O -EPAP: 10 ± 2 cm H2O
	CPAP	NR	CPAP	NR	NR
Blankenburg, 2017 <sup>5</sup> Observational	HMV (pressure controlled ventilation or pressure support ventilation)	VS III; ResMed (Saime SA, France)	Inspiratory pressure, PEEP, inspiratory time, rate, inspiratory flow trigger	12 hours/day	-5.2 ± 3.2 hours/day -Inspiratory pressure 22 ± 3.9 cm H2O -PEEP: 5.3 ± 2.7 cm H2O -Rate: 15.3 ± 2.9 breaths/minute
Masa 2016 <sup>42</sup> RCT	BPAP volume assured pressure support ventilation	NR	IPAP, EPAP, rate, target minute ventilation	NR	NR
Perez de Llano 2005 <sup>52</sup> Observational	HMV/BPAP mix	HMV: Home 2; Airox (Pau, France)  BPAP: DP-90; Taema (Paris, France)  PV-102; Breas (Gothenburg, Sweden)	HMV: volume cycled NOS	NR	NR
Priou, 2010 <sup>56</sup> , Observational	BPAP	NR	NR	NR	NR
Tsolaki, 2011 <sup>66</sup> Observational	BPAP ST	VPAP III ST (ResMed; Sydney, Australia)	IPAP, EPAP, rate	≥ 4 hours daily	-IPAP: 16.3 ± 2.4 cm H2O -EPAP: 6.1 ± 1.0 cm H2O

Note: ± denotes standard deviation.

BPAP: Bilevel Positive Airway Pressure, cm: Centimeter, CPAP: Continuous Positive Airway Pressure, EPAP: Expiratory positive airway pressure, FDA: Food and Drug Administration, H2O: Hydrogen dioxide, HMV: Home Mechanical Ventilation, IPAP: Inspiratory Positive Airway Pressure, NR: Not reported, OHS: Obesity hypoventilation syndrome, PEEP: positive end expiratory pressure, S: spontaneous mode, ST: Spontaneous/timed mode, USA: United States of America.

**Table F.22. Other Respiratory Diseases – Equipment parameters**

Author, Year, Study Design	Device/mode	Model and Manufacturer	Device Characteristics	Prescribed Usage (frequency and duration)	Actual Usage
Salturk, 2015 <sup>57</sup> Observational	BPAP ST	NR	IPAP, EPAP, rate	NR	-5.8 ± 1.4 hours/day -IPAP: 21 ± 5 cm H2O -EPAP: 5.5 ± 0.7 cmH2O
Benhamou, 1997 <sup>2</sup> Observational	HMV (volume assist control ventilation)	Monnal D (Taema; Antony, France)  Eole 3 (Saime; Savigny-Le-Temple, France)	tidal volume, PEEP, rate	NR	NR

Note: ± denotes standard deviation.

BPAP: Bilevel Positive Airway Pressure, cm: Centimeter, EPAP: Expiratory positive airway pressure, FDA: Food and Drug Administration, H2O: Hydrogen dioxide, HMV: Home Mechanical Ventilation, IPAP: Inspiratory Positive Airway Pressure, NR: Not reported, PEEP: positive end expiratory pressure, ST: Spontaneous/timed mode

**Table F.23. Mixed Diseases – Equipment parameters**

Author, Year, Study Design	Diseases	Device/mode	Model and Manufacturer	Device Characteristics	Prescribed Usage (frequency and duration)	Actual Usage
Hazenberg, 2014 <sup>33</sup> RCT	NMD, TRD	HMV (pressure controlled ventilation) with change to HMV (volume assist control ventilation) if not tolerated	Elisee 150 (ResMed; Paris, France) (FDA approved)	HMV (pressure controlled ventilation): inspiratory pressure, PEEP, rate, inspiratory time  HMV (volume assist control ventilation): tidal volume, PEEP, rate	≥ 6 hours/night	-IPAP: 10 cm H2O (pressure mode) -EPAP: 4 cm H2O (pressure mode) -Tidal volume: 8-10 ml/kg (pressure mode)
Crespo, 2009 <sup>18</sup> Observational	COPD, TRD, NMD, OHS, Other	HMV (pressure or volume controlled NOS)	NR	NR	NR	<u>Age ≥ 75 years old</u> -IPAP: 14-20 cm H2O

Author, Year, Study Design	Diseases	Device/mode	Model and Manufacturer	Device Characteristics	Prescribed Usage (frequency and duration)	Actual Usage
						-EPAP: 3-8 cmH2O -Tidal volume: 500-800 ml -Rate: 16- 22 breaths/minute <u>Age 65-74 years old</u> -IPAP: 14-24 cm H2O -EPAP: 14-20 cmH2O -Tidal volume: 400-800 ml -Rate: 14- 22 breaths/minute <u>Age &lt;65 years old</u> -IPAP: 14-30 cm H2O -EPAP: 3-8 cmH2O -Tidal volume: 400-1000 ml -Rate: 12- 24 breaths/minute
Munoz, 2005 <sup>44</sup> Observational	NMD, TRD	HMV (volume assist control ventilation)	NR	tidal volume, PEEP, rate	NR	-Tidal volume: 9.5 ± 0.7 ml/kg -Rate: 16.8 ± 2.7 breaths/minute
		HMV (volume control)	NR	tidal volume, PEEP, rate	NR	-Tidal volume: 8.61 ± 1.6 ml/kg -Rate: 16.7 ± 2.7 breaths/minute
Chiang, 2003 <sup>13</sup> RCT	COPD, Other	BPAP NOS	NR	IPAP, EPAP	NR	-IPAP: 11.8 ± 0.6 cm H2O -EPAP: 4.5 ± 0.4 cm H2O
Windisch, 2006 <sup>69</sup> Observational	HMV (pressure controlled ventilation)	PV401(Breas Medical AB; Moelnlycke, Sweden)	inspiratory pressure, PEEP, inspiratory time, rate	NR	-IPAP: 23.2 ± 2.8 mbar -Rate: 20.5 ± 1.9 breaths/minute -Inspiratory time	

Author, Year, Study Design	Diseases	Device/mode	Model and Manufacturer	Device Characteristics	Prescribed Usage (frequency and duration)	Actual Usage
					1.2 ± 0.1 seconds	

Note: ± denotes standard deviation

cm: Centimeter, COPD: Chronic obstructive pulmonary diseases, EPAP: Expiratory positive airway pressure, FDA: Food and Drug Administration, H<sub>2</sub>O: Hydrogen dioxide, IPAP: Inspiratory Positive Airway Pressure, ml: milliliter, kg: kilogram, NMD: Neuromuscular Disease, NR: Not reported, OHS: Obesity hypoventilation syndrome, PEEP: positive end expiratory pressure, TRD: Thoracic Restrictive Disease



KQ4. What respiratory services, other than the technical support of the use of the prescribed equipment, are being provided to the patients in the home?

**Table F.24. COPD – Respiratory services**

Author, Year, Study Design	Device/Mode	Respiratory Services delivered in the home (including by whom and how frequently)
Murphy, 2017 <sup>46</sup> RCT	BPAP ST	-Smoking cessation NOS -COPD education NOS
Oscroft, 2014 <sup>50</sup> RCT	BPAP iVAPS versus BPAP ST	-24 hour hotline NOS
Bhatt, 2013 <sup>4</sup> RCT	BPAP NOS	-Daily phone call by respiratory therapist during first week -One home visit by respiratory therapist during first week
Duiverman, 2011 <sup>22, 23</sup> RCT	BPAP ST	-Supervision by specialized nurse NOS
Oscroft, 2010 <sup>48</sup> Observational	BPAP ST started after AECOPD versus BPAP ST started in stable patient without exacerbation	-24 hour hotline NOS
Crespo, 2009 <sup>18</sup> Observational	HMV (pressure or volume NOS)	-Emergency phone number
Cheung, 2010 <sup>12</sup> RCT	BPAP ST versus CPAP	-Nurse hotline NOS
McEvoy, 2009 <sup>43</sup> RCT	BPAP S	-Telephone calls answered by nurses as needed
Casanova, 2000 <sup>10</sup> RCT	BPAP S	-“Close contact was maintained” for first 3 weeks
Garrod, 2000 <sup>30</sup> RCT	BPAP S	-Phone call every 2 weeks to encourage use
Clini, 1996 <sup>14</sup> Observational	BPAP ST	-Home care program (initial evaluation of physical, occupational, and dietary needs; monthly physician visits; monthly education about treatments and correct medication use and coping strategies; periodic phone calls).
Köhnlein, 2014 <sup>37</sup> RCT	BPAP ST	-24 hour hotline staffed by health-care providers and specialized nurses
Gay, 1996 <sup>28</sup> RCT	BPAP ST	-Regular phone calls to ensure compliance.
Durao, 2018 <sup>25</sup> RCT	HMV/BPAP mix	-Smoking cessation NOS
Tsolaki, 2011 <sup>66</sup> , Observational	BPAP ST	-Full technical support when required by “technically skilled personnel”

Medical services not reported: oxygen use, disease optimization [such as inhalers, vaccination, etc.], scheduled rehospitalizations, regular office visits with physician, pulmonary rehabilitation

AECOPD: acute exacerbation of chronic obstructive pulmonary disorder, BPAP: Bilevel Positive Airway Pressure, COPD: chronic obstructive pulmonary disease, CPAP: Continuous Positive Airway Pressure, HMV: Home Mechanical Ventilation, iVAPS: intelligent volume assured pressure support, NOS: Not otherwise Specified, PSV: pressure support ventilation, RCT: randomized controlled trail, S: spontaneous mode, ST: spontaneous/timed breath mode, VPAP: variable positive airway pressure

**Table F.25. Thoracic Restrictive Disorders – Respiratory services**

Author, Year, Study Design	Device/Mode	Respiratory Services delivered in the home (including by whom and how frequently)
Doménech-Clar, 2003 <sup>20</sup> Observational	BPAP NOS	-Telephone helpline (24 hours)
Tsolaki, 2011 <sup>66</sup> Observational	BPAP ST	-Full technical support when required by “technically skilled personnel”

Medical services not reported: oxygen use, disease optimization [such as inhalers, vaccination, etc.], scheduled rehospitalizations, regular office visits with physician, pulmonary rehabilitation

BPAP: Bilevel Positive Airway Pressure, NOS: Not otherwise Specified

**Table F.26. Neuromuscular Disease – Respiratory services**

Author, Year, Study Design	Device/Mode	Respiratory Services delivered in the home (including by whom and how frequently)	Outcome
Sanjuan-López, 2014 <sup>60</sup> Observational	HMV (PSV or ST)	-Telephone calls NOS -Home visit from the equipment supply company nurse -Mechanical cough assistance (Cough Assist, insufflator-exsufflator, MI-E, Emerson) was provided and caregiver training was provided if expectoration problems with a cough peak flow lower < 270 l/minute despite assisted cough physiotherapy.	
Pinto, 2010 <sup>54</sup> Observational	BPAP ST + weekly telemonitoring versus BPAP ST without weekly telemonitoring	-Telephone helpline	The BPAP ST + Weekly telemonitoring group had significantly lower number of office visits (IRR: 0.34, 95% CI: 0.29 to 0.38); ER visits (IRR: 0.19; 95% CI: 0.10 to 0.37); hospital admission (IRR: 0.17; 95% CI: 0.07 to 0.41). There was no significant difference on mortality (OR: 1.00; 95% CI: 0.24 to 4.18) or median survival time (from BPAP adoption to death) (865 days vs. 334 days, p=0.13).
Doménech-Clar, 2003 <sup>20</sup> Observational	BPAP NOS	-Telephone helpline (24 hours)	

<b>Author, Year, Study Design</b>	<b>Device/Mode</b>	<b>Respiratory Services delivered in the home (including by whom and how frequently)</b>	<b>Outcome</b>
Nauffal, 2002 <sup>47</sup> Observational	BPAP NOS	-Telephone helpline (24 hours)	
Gonzalez-Bermejo, 2013 <sup>32</sup> Observational	BPAP ST	-Instruction on assisted cough techniques including mechanical insufflation-exufflation by a respiratory physiotherapist	
Sancho, 2014 <sup>58</sup> Observational	HMV (volume cycled) versus BPAP ST	-Guideline based multidisciplinary care, management of cough impairment when necessary, nutritional support, and medical treatment with riluzole.	
Coco, 2006 <sup>17</sup> Observational	BPAP ST	-Suction devices for secretion clearance -All patients were also taught assisted cough techniques by an experienced respiratory physiotherapist, including mechanical insufflators-exsufflators.	
Bourke, 2006 <sup>7</sup> RCT	BPAP ST	-Multidisciplinary clinical team review, education about assisted cough techniques, posture, bed raisers, adjustable beds, palliative care, hospice as needed.	
Sancho, 2017 <sup>59</sup> Observational	HMV (volume assist control ventilation)	-Guideline based multidisciplinary care, management of cough impairment when necessary, nutritional support, and medical treatment with riluzole.	
Farrero, 2005 <sup>26</sup> RCT	HMV/BPAP mix	-Salviary aspirator if ineffective cough -Training of caregivers using assisted cough maneuvers and hyperinflation with a compressible ventilator bag or volume ventilator	
Tsolaki, 2011 <sup>66</sup> Observational	BPAP ST	-Full technical support when required by "technically skilled personnel"	

Medical services not reported: oxygen use, disease optimization [such as inhalers, vaccination, etc.], scheduled rehospitalizations, regular office visits with physician, pulmonary rehabilitation

BPAP: Bilevel Positive Airway Pressure, CI: confidence interval, ER: emergency room, HMV: Home Mechanical Ventilation, IRR: incidence rate ratio, NOS: Not otherwise Specified, OR: odds ratio, PSV: pressure support ventilation, RCT: randomized controlled trail, S: spontaneous mode, ST: spontaneous/timed breath mode, VPAP: variable positive airway pressure

**Table F.27. Obesity Hypoventilation Syndrome – Respiratory services**

Author, Year, Study Design	Device/Mode	Respiratory Services delivered in the home (including by whom and how frequently)
Masa, 2015 <sup>40, 41</sup> RCT	Mixed: HMV and BPAP mix (all with bilevel pressure with assured volume)	Lifestyle counseling: 1,000-calorie diet, correct sleep hygiene and habits (avoiding the supine decubitus position; maintaining regular sleep habits and exercise, not consuming sedatives, stimulants, or alcohol; not smoking tobacco; and avoiding heavy meals within 4 hours before bedtime).
Borel, 2011 <sup>6</sup> RCT	BPAP ST	Lifestyle counseling: 1 hour education session, patients were informed about the general health risks associated with obstructive sleep apnea and obesity (i.e., information about harmful lifestyle factors, such as smoking, reduced physical activity, and alcohol drinking). A specialized nurse provided dietary and lifestyle counseling, with the emphasis placed on diet, exercise, and modification of lifestyle in general, specifically focusing on eating behavior. The patients were advised to reduce fat by increasing their intake of fruits and vegetables and by limiting fatty meat, sweets, pastries, and desserts. The subjects were recommended to increase their overall level of daily physical activity.
Tsolaki, 2011 <sup>66</sup> Observational	BPAP ST	-Full technical support when required by “technically skilled personnel”

Medical services not reported: oxygen use, disease optimization [such as inhalers, vaccination, etc.], scheduled rehospitalizations, regular office visits with physician, pulmonary rehabilitation

BPAP: Bilevel Positive Airway Pressure, HMV: Home Mechanical Ventilation, OHS: obesity hypoventilation syndrome, RCT: randomized controlled trail, ST: spontaneous/timed breath mode

**Table F.28. Mixed Diseases – Respiratory services**

Author, Year, Study Design	Diseases	Device/Mode	Respiratory Services delivered in the home (including by whom and how frequently)
Munoz, 2005 <sup>44</sup> Observational	NMD/TRD	HMV volume assist/control mode versus HMV volume control mode	-Telephone helpline
Chiang, 2003 <sup>13</sup> RCT	COPD, other	BPAP NOS	-Telephone interviews by respiratory therapist every 2 weeks to assess compliance and ventilator usage.

Medical services not reported: oxygen use, disease optimization [such as inhalers, vaccination, etc.], scheduled rehospitalizations, regular office visits with physician, pulmonary rehabilitation

BPAP: Bilevel Positive Airway Pressure, COPD; chronic obstructive pulmonary disease HMV: Home Mechanical Ventilation, NMD: neuromuscular disease, NOS: not otherwise specified, RCT: randomized controlled trail, ST: spontaneous/timed breath mode, TRD: Thoracic Restrictive Disorder