

## Appendix D. Characteristics of Included Studies

**Table D.1. Characteristics of included studies**

Author, Year	Study Country, Study Design, Study Period	Risk of Bias	Inclusion / Exclusion Criteria	Intervention and comparisons (Groups)	Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.	Patient Characteristics	Disease
Aboussouan, 1997 <sup>1</sup>	Observational in United States, 03/1993 to 02/1996	Moderate ROB	Inclusion: ALS via el Escorial criteria; dyspnea on exertion or PaCO <sub>2</sub> ≥ 45 mmHg or orthopnea or FVC < 60% predicted.	HMV/BPAP mix (tolerant)	HMV PLV-100; Life Care Products (Lafayette, Colorado, USA) (FDA approved 510(k) clearance)	18 Patients aged 61.5 ± 11.9, 22.2% female	NMD
				HMV/BPAP mix (intolerant)	BPAP BiPAP; Respironics Inc. (Murrysville, Pennsylvania, USA) (FDA approved 510(k) clearance)	21 Patients aged 61.8 ± 15.2, 33.3% female	
Benhamou, 1997 <sup>2</sup>	Observational comparative case-control study in France	High ROB	Inclusion: Treated by home non-invasive mechanical ventilation & LTOT for severe chronic respiratory failure from diffuse bronchiectasis.	HMV (volume assist control ventilation)	HMV Monnal D; Taema (Antony, France) (Not FDA approved)	14 Patients aged 64±10	Other
				Oxygen alone	No PAP	14 Patients aged 66±9	
Bertella, 2017 <sup>3</sup>	RCT in Italy, 03/2011-03/2014	Low ROB	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.	BPAP volume assured pressure support ventilation inpatient initiation	BPAP Trend II ST 30; Hoffrichter (Schwerin, Germany) (Not FDA approved)	25 patients aged 65.92±10.18, 32% female	NMD
				BPAP volume assured pressure support ventilation outpatient initiation	BiPAP Synchrony II; Philips Respironics (Murrysville, PA, USA) (FDA approved 510(k) clearance)	25 patients aged 61.26±8.64, 44% female	

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Bhatt, 2013 <sup>4</sup>	RCT in USA	High ROB	<p>Inclusion: Stable COPD with 10 pack year smoking history, low clinical probability of OSA</p> <p>Exclusion: Congestive heart failure, OSA, chronic respiratory conditions other than COPD, age&lt;35 years, diseases limiting life expectancy &lt;2 years, active malignancies in previous 2 years, process precluding a nasal mask.</p>	BPAP NOS	BPAP BiPAP Synchrony; Respironics Inc. (FDA approved 510(k) clearance)	15 Patients, 47% female	COPD
				No BPAP	No PAP	12 Patients aged 68 (IQR 65-78), 0% female	
Blankenburg, 2017 <sup>5</sup>	Observational, Prospective in Germany, 01/01/2011 to 12/31/2011	Moderate ROB	<p>Inclusion: COPD (GOLD criteria NOS), PaCO<sub>2</sub>&gt;7.0kPa, pH&gt;7.35, stable disease (no exacerbation in 2 weeks prior)</p> <p>Exclusion: decompensated heart failure or "other conditions" that cause chronic respiratory failure, systemic corticosteroids</p>	HMV (pressure controlled ventilation or pressure support ventilation)	HMV VS III; ResMed (Saime SA, France) (FDA approved 510(k) clearance)	51 patients aged 66.9 (SE 1.3), 37% female	COPD
						<p>Inclusion: OHS (BMI &gt;30kg/m<sup>2</sup>, chronic daytime hypercapnia PaCO<sub>2</sub>&gt;6.7kPa, pH&gt;7.35), symptoms of hypercapnia NOS)</p> <p>Exclusion: decompensated heart failure or "other conditions" that cause chronic respiratory failure</p>	
Borel, 2011 <sup>6</sup>	RCT in Switzerland	High ROB	<p>Inclusion: Age 20-75 years, BMI &gt;30</p> <p>Exclusion: Declined or presented any significant</p>	BPAP ST	BPAP GoodKnight-425ST; Covidien (FDA approved 510(k) clearance)	19 Patients aged 58±11, 56% female	OHS

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			airway obstruction, scoliosis, cardiac failure, progressive NMD.	Lifestyle counseling	No PAP	18 Patients aged 54±6, 59% female	
Bourke, 2006 <sup>7</sup>	RCT in United Kingdom, 03/2000 to 12/2003	High ROB	Inclusion:  Exclusion: Current or previous NIPPV use, significant comorbidities, age>75 years, inability to complete quality of life assessment.	BPAP ST (full cohort)	BPAP VPAP STII; ResMed United Kingdom Ltd (Abingdon, United Kingdom) (FDA approved 510(k) clearance)	22 Patients aged 63.7±10.3, 36% female	NMD
				No BPAP ST (full cohort)	No PAP	19 Patients aged 63±8.1, 47% female	
				BPAP ST (good bulbar patients)	BPAP good bulbar	11 Patients	
				No BPAP ST (good bulbar patients)	No PAP good bulbar	9 Patients	
				BPAP ST (poor bulbar patients)	BPAP poor bulbar	11 Patients	
				No BPAP ST (poor bulbar patients)	No PAP poor bulbar	10 Patients	
Budweiser, 2007 <sup>8</sup>	Observational Prospective in Germany, 01/2002 to 12/2005	Low ROB	Inclusion: Less than 80 years old, severe COPD (GOLD IV), FEV1/FVC <70%, FEV1 <50% predicted, PaCO <sub>2</sub> > 50mmHg after therapy/treatment for exacerbation  Exclusion: Malignancy diagnosis within prior 5 years, intubation or tracheostomy prior to NIPPV	BPAP (pressure controlled ventilation)	BPAP Twin Air; Airox Inc. (Pau, France) (Not FDA approved)  Smart Air; Airox Inc. (Pau, France) (Not FDA approved)  BiPAP Synchrony; Respironics Inc. (Murrysville, USA) (FDA approved 510(k) clearance)	99 Patients aged 34.2±8.4, 36.4% female	COPD
				No BPAP	No PAP	41 Patients aged 66.6±8.6, 31.7% female	

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Buyse, 2003 <sup>9</sup>	Observational Retrospective in Belgium, 09/1990 to 03/2001	Moderate ROB	Inclusion: Consecutive kyphoscoliosis & respiratory insufficiency patients started on LTOT and/or NIPPV at center.	HMV (volume or pressure cycled ventilator NOS) + oxygen	HMV Eole 3; Saime (Savigny-Le-Temple, France) (Not FDA approved)  O'nyx; Nellcor Puritan Bennet (Villers-les-Nancy, France) (FDA approved 510(k) clearance)	18 Patients aged 61±7, 77.8% female	TRD
				Oxygen alone	No PAP	15 Patients aged 62±7, 66.66% female	
Casanova, 2000 <sup>10</sup>	RCT in Spain, 1995 to 1997	High ROB	Inclusion: Age 45-75 years, smoking history 20 pack years, clinically stable  Exclusion: Refusal to stop smoking, OSA, >10 apnea-hypopnea episodes per hour, other etiologies of chronic airway obstruction, significant comorbidities.	BPAP S + standard care	BPAP DP-90; Taema (Paris, France) (Not FDA approved)	20 Patients aged 64±5, 0% female	COPD
				Standard care	No PAP	24 Patients aged 68±4, 4% female	
Castillejo, 2014 <sup>11</sup>	Observational Prospective in Spain, 1998 to 2010	High ROB	Inclusion: OHS & BMI >30  Exclusion: Obstructive disease with FEV1/FVC ratio <70%, NMD with respiratory involvement, respiratory disease other than OHS.	BPAP ST in OHS without OSA	BPAP Harmony BiPAP; Respironics (Louisville, USA) (FDA approved 510(k) clearance)	50 Patients aged 64.62±9.8, 82% female	OHS
				BPAP ST in OHS with OSA		33 Patients aged 64.47±8.2, 57.6% female	
Cheung, 2010 <sup>12</sup>	RCT in China, 01/2007 to 03/2009	High ROB	Inclusion: Severe exacerbation with persistent respiratory acidosis (despite treatment with bronchodilators, corticosteroids, antibiotics), required NIPPV treatment  Exclusion: Active smokers, RF	CPAP	CPAP BiPAP Synchrony; Respironics Inc. (Murrysville, USA) (FDA approved 510(k) clearance)	24 Patients aged 71±7.7, 8.3% female	COPD

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			from non-COPD cause, evidence of pneumonia, transmissible infections, requiring long-term systemic steroids, comorbidity giving life expectancy <1 year, significant OSA, already on home NIPPV, inability to comply with study protocol.	BPAP ST	BPAP BiPAP Synchrony; Respironics Inc. (Murrysville, USA) (FDA approved 510(k) clearance)	23 Patients aged 69.5±7.8, 8.7% female	
Chiang, 2003 <sup>13</sup>	RCT in Taiwan, 06/2001 to 11/2002	Moderate ROB	Inclusion: Diagnosed with COPD and asthma and bronchiectasis, repeat admission due to lung deterioration despite treatment, well-motivated, sleepy during day or headache upon waking in morning  Exclusion: Uncooperative, poor motivation, unable to tolerate nocturnal nasal positive pressure ventilation, OSA, unable to perform 6-minute walk distance test due to other disease.	BPAP NOS	BPAP BiPAP; Respironics (Murrysville, USA) (FDA approved 510(k) clearance)	13 Patients aged 62.5±11.5, 23.1% female	Mixed( COPD, Other)
				no BPAP NOS	No PAP	14 Patients aged 65.5±10, 35.7% female	
Clini, 1996 <sup>14</sup>	Observational Prospective in Italy, 12/1991 to 09/1992	High ROB	Inclusion: Severe COPD, ≥1 admission due to severe exacerbation in prior 18 months  Exclusion: Suspicion of sleep apnea, comorbidities making patients unsuitable for long-term trials	BPAP ST + home care + oxygen	BPAP BiPAP; Respironics (Murrysville, USA) (FDA approved 510(k) clearance)	17 Patients aged 62±5, 29.4% female	COPD
				Home care + oxygen	No PAP	17 Patients aged 67±7, 47% female	
				Oxygen	No PAP	29 Patients aged 62±8, 34.5% female	

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Clini, 1998 <sup>15</sup>	Observational Prospective in Italy, 12/1991 to 12/1994	Low ROB	Inclusion: Clinically stable, $\geq 1$ ICU admission due to severe exacerbation within 2 years prior, care-giver at home, geographical allocation allowing access to the hospital  Exclusion: other organ failure, cancer, inability to cooperate to long-term trials, suspicion of sleep apnea.	BPAP ST + oxygen	BPAP BiPAP; Respironics (Murrysville, USA) (FDA approved 510(k) clearance)	28 Patients aged 66 $\pm$ 6, 21.4% female	COPD
				Oxygen	No PAP	21 Patients aged 66 $\pm$ 8, 33% female	
Clini, 2002 <sup>16</sup>	RCT in Italy/France, 06/1996 to 01/2000	High ROB	Inclusion: Age $\leq 75$ years, LTOT $\geq 6$ months, dyspnea score (assessed by Medical Research Council) $\geq 2$ , FEV1 $< 1.5$ liters, FEV1/FVC $< 60\%$ , TLC $\geq 90\%$ predicted, PaCO2 $> 6.6$ kPa, PaO2 $< 7.8$ kPa breathing room at rest  Exclusion: 15% increase FEV1 after salbutamol, pH $\leq 7.34$ , active smokers, history of OSA (defined by apnea-hypopnea index $> 10$ episodes per hour), therapy with systemic steroids, concomitant chronic systemic diseases (HF, diabetes, infections, neoplasm, etc.), other chronic respiratory diseases (fibrothorax, bronchiectasis, cystic fibrosis), home care program other than LTOT	BPAP ST + LTOT	BPAP BiPAP ST 30; Respironics (Murrysville, USA) (FDA approved 510(k) clearance)	39 Patients aged 64 $\pm$ 7, 18% female	COPD
				LTOT	No PAP	47 Patients aged 66 $\pm$ 14, 21.3% female	
Coco, 2006 <sup>17</sup>	Observational Prospective in Italy, 10/1999 to 07/2003	Low ROB	Inclusion: Definite/probable ALS  Exclusion: Primary lateral	BPAP ST (use $\geq 4$ hours/day)	BPAP BiPAP; Respironics (Vitalaire, Italy) (FDA approved)	44 Patients aged 62.3 $\pm$ 11.4, 31.8% female	NMD

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			sclerosis, diagnosis other than ALS during followup.	BPAP ST < 4 hours/day)	510(k) clearance)	27 Patients aged 61.1±9.9, 40.7% female	
Crespo, 2009 <sup>18</sup>	Observational Retrospective in Spain, 1998 to 2001	High ROB	Inclusion: Stable disease initiating scheduled HMV with nasal mask  Exclusion: Invasive ventilation by tracheostomy, NIPPV with face mask/mouthpiece, HMV with nasal mask started during acute phase of disease.	HMV (pressure or volume NOS) in age ≥ 75 years old	HMV NOS	10 Patients aged 76.9±2.1, 30% female	Mixed (COPD, TRD, NMD, OHS, Other)
				HMV (pressure or volume NOS) in 65-74 years old		40 Patients aged 69.5±3.2, 45% female	
				HMV (pressure or volume NOS) in <65 years old		41 Patients aged 52.7±12.0, 54% female	
De Backer, 2011 <sup>19</sup>	RCT in Belgium	Moderate ROB	Inclusion: Age 18-80 years, COPD stage III/IV, exacerbation hospitalization, persisting hypercapnia, stopped smoking, no home NIPPV before admission  Exclusion: Invasive ventilation, asthmatic, restrictive lung disease, malignancy, heart failure, OSA.	BPAP NOS	BPAP BiPAP synchrony; Respironics (Murrysville, USA) (FDA approved 510(k) clearance)	10 Patients aged 65±7	COPD
				Standard care		No PAP	
Domenéch-Clar, 2003 <sup>20</sup>	Observational Prospective in Spain, 01/1997 to 11/2001	High ROB	Inclusion: Hospitalized 48-72 hours, moderate to severe restrictive respiratory disorder from TWD or NMD, clinically stable.	BPAP NOS in thoracic wall diseases	BPAP DP-90; Taema (Paris, France) (FDA approved 510(k) clearance)	27 Patients aged 55.6, 40.7% female	TRD
				BPAP NOS in neuromuscular diseases		18 Patients aged 42.5, 50% female	NMD
Dreher, 2010 <sup>21</sup>	RCT in Germany	High ROB	Inclusion: CHRF due to COPD stage IV  Exclusion: Acute RF, invasive ventilation via tracheostomy, weaned from invasive ventilation, intubated during prior 3 months, other	HMV (pressure assist/control) (time period 1)	HMV Breas Vivo 40; Breas Medical AB (Molnlycke, Sweden) (FDA approved 510(k) clearance)	9 Patients	COPD
				HMV (PSV ST) (time period 1)		Smart Air; Airox (Pau)	

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			ventilatory support prior to study.	HMV (PSV ST) (time period 2) HMV (pressure assist/control) (time period 2)	Cedex, France) (Not FDA approved)		
Duiverman, 2011 <sup>22, 23</sup>	RCT in Netherlands	Moderate ROB	Inclusion: COPD stage III/IV, age 40-76 years, clinically stable, chronic hypercapnic RF  Exclusion: cardiac/neuromuscular disease limiting exercise tolerance, exposure to pulmonary rehab program (previous 18 months), previous exposure to chronic NIPPV ever, apnea-hypopnea index $\geq 10$ h.	BPAP ST + pulmonary rehabilitation  Pulmonary rehabilitation alone	BPAP BiPAP Synchrony; Respironics Inc. (FDA approved 510(k) clearance)  No PAP	24 Patients aged 63 $\pm$ 10, 33.3% female  32 Patients aged 61 $\pm$ 8, 46.9% female	COPD
Duiverman, 2017 <sup>24</sup>	RCT in Netherlands	High ROB	Inclusion: COPD (GOLD III or IV), $\geq 2$ AECOPD with acute hypercapnic respiratory failure (pH $<$ 7.35) per year, daytime Inclusion: PaCO <sub>2</sub> $\geq 6.7$ kPa (50 mmHg) or nocturnal PaCO <sub>2</sub> $\geq 7.3$ kPa (55 mmHg) or nighttime rise in PtCO <sub>2</sub> $\geq 1.3$ kPa (10 mmHg), stable (no AECOPD in prior 4 weeks, pH $>$ 7.35).  Exclusion: TRD, NMD	HMV/BPAP mix (pressure controlled ventilation) (high intensity)  HMV/BPAP mix (pressure support ventilation) (low intensity)	HMV Vivo 50; Breas Medical (Molndal, Sweden) (FDA approved 510(k) clearance)  BPAP Stellar 100; Resmed (Martinsried, Germany) (FDA approved 510(k) clearance)	Crossover – 11 patients aged 68.7 $\pm$ 8.5, 54% female	COPD
Durao, 2018 <sup>25</sup>	Observational Retrospective in Portugal, 08/1/2011 to 07/31/2014	Low ROB	Inclusion: COPD NOS  Exclusion: No clinical assessment in prior 6 months, OSA with a history of noncompliance with CPAP	HMV/BPAP mix started in AECOPD	BPAP VPAP ST S9; Resmed (FDA approved 510(k) clearance)  VPAP ST STA;	62 patients aged 64.6 $\pm$ 10.4, 12.9% female	COPD



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				HMV/BPAP mix started in stable disease	Resmed (FDA approved 510(k) clearance)  BIPAP PR1; Philips Respironics (FDA approved 510(k) clearance)  BiPAP A30; Philips Respironics (FDA approved 510(k) clearance)  BiPAP A40; Philips Respironics (FDA approved 510(k) clearance)  <u>HMV</u> Trilogy 100; Philips Respironics (FDA approved 510(k))	47 patients aged 66.9±8.4, 17% female	
Farrero, 2005 <sup>26</sup>	Observational in Spain, 1988 to 12/2002	Moderate ROB	Inclusion: ALS NOS	HMV/BPAP mix in pre protocol group  HMV/BPAP mix in post protocol group	<u>HMV</u> PLV-100; Life Care Products (FDA approved 510(k) clearance)	11 Patients aged 53 ± 11  48 Patients aged 62 ± 10	NMD

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					PV 501; BREAS Medical (Gothenburg, Sweden) (FDA approved 510(k) clearance)  BPAP BiPAP; Respironics (FDA approved 510(k) clearance)  VPAP ST II; Sullivan (FDA approved 510(k) clearance)		
Funk, 2010 <sup>27</sup>	RCT in Austria, 04/01/2003 to 02/28/2007	Moderate ROB	Inclusion: COPD requiring invasive/non-invasive mechanical ventilation due to acute RF, clinically stable, hypercapnic  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.	BPAP NOS	BPAP - Not reported ("various types of patient-triggered bi-level positive pressure ventilators were used")	13 Patients aged 62±6, 46% female	COPD
				Standard care	No PAP	13 Patients aged 65±6, 38% female	
Gad, 2014 <sup>28</sup>	Observational Prospective in Egypt, 10/2012 to 04/2014	Moderate ROB	Inclusion: Severe COPD stage III/IV, FEV1/FVC <70%, clinically stable  Exclusion: invasive mechanical ventilation, OSA, cardiac disease limiting exercise tolerance, NMDs, orthopedic impairment of shoulder girdle	BPAP ST + exercise program	BPAP	15 Patients aged 65.70±10, 40% female	COPD
				Exercise program	No PAP	15 Patients aged 66.41±9, 26.7% female	

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Galli, 2014 <sup>29</sup>	Observational Retrospective in USA, 01/2011 to 12/2011	High ROB	Inclusion: Primary/secondary discharge diagnosis of AECOPD, hypercapnic RF during hospitalization  Exclusion: discharged to hospice, no documented hypercapnia, not receiving NIPPV during hospitalization.	BPAP NOS post hospital admission	BPAP	78 Patients aged 61.6±10.2, 57.7% female	COPD
				No BPAP post hospital admission	No PAP	88 Patients aged 64.9±10.8, 67% female	
Garrod, 2000 <sup>30</sup>	RCT in England	High ROB	Inclusion: Severe COPD, all patients had limited exercise tolerance due to dyspnea and no previous exposure to NIPPV  Exclusion: unstable angina, intermittent claudication, and other mobility-limiting conditions.	BPAP S + pulmonary rehabilitation	BPAP BiPAP ST 30; Respironics (Murrysville, USA) (FDA approved 510(k) clearance)	23 Patients aged 63	COPD
				Pulmonary rehabilitation	No PAP	22 Patients aged 67	
Gay, 1996 <sup>31</sup>	RCT in USA, 1989 to 1992	High ROB	Inclusion: Age<80 years, BMI≤30, FEV1 <40%  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.	BPAP ST	BPAP BiPAP; Respironics (Murrysville, USA) (FDA approved 510(k) clearance)	7 Patients aged 71.0±4.5, 28.6% female	COPD
				Sham BPAP ST (CPAP at lowest setting)	No Device	6 Patients aged 66.5±9.1, 16.6% female	
Gonzalez-Bermejo, 2013 <sup>32</sup>	Observational Retrospective in France, 01/01/2003 to 12/31/2007	High ROB	Inclusion: 4h/night minimal adherence  Exclusion: Use of other ventilator types, without integrated SpO2 monitoring.	BPAP ST "correctly ventilated patients"	BPAP VPAP-III or VPAP-IV Resmed (Sydney, Australia) (FDA approved 510(k) clearance)	40 Patients aged 63±12, 32.5% female	NMD
				BPAP ST "insufficiently ventilated patients"		42 Patients aged 64±10, 17% female	

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Hazenberg, 2014 <sup>33</sup>	RCT in Netherlands, 10/2008 to 10/2012	Moderate ROB	<p>Inclusion: Chronic RF from NMD or thoracic cage disorder, orthopnea from diaphragm paralysis &amp; daytime normocapnia also included</p> <p>Exclusion: Strictly COPD patients, not mask naive, acute RF, age &lt; 18 years, invasive ventilation, nursing home residing.</p> <p>(77 total patients in both groups, 3 patients on volume control, 74 patients on pressure control)</p>	HMV started at home pressure controlled ventilation with change to volume assist control ventilation if not tolerated	HMV Elisee 150; ResMed (Paris, France) (Not FDA approved)	38 Patients aged 59.9±12.6, 47.4% female	Mixed (NMD, TRD)
				HMV started in the hospital pressure controlled ventilation with change to volume assist control ventilation if not tolerated		39 Patients aged 56.9±13.9, 35.9% female	
Heinemann, 2011 <sup>34</sup>	Observational Retrospective in Germany, 01/2002 to 02/2008	High ROB	<p>Inclusion: COPD, prolonged weaning from invasive mechanical ventilation</p> <p>Exclusion: Intubated from cardiogenic edema or cardiopulmonary resuscitation</p>	BPAP (pressure controlled ventilation)	BPAP NOS	39 Patients aged 64.6±10.8, 30.1% female	COPD
				No BPAP	No PAP	43 Patients aged 72.8±8.6, 25.6% female	
Hitzl, 2009 <sup>35</sup>	Observational Prospective in Germany	Low ROB	<p>Inclusion: HMV initiated ≥3 months prior to study, undergone bioelectrical impedance analysis measurement, regularly readmitted for routine followup, all had CHRf</p> <p>Exclusion: OHS, progressive NMD, tracheostomy.</p>	HMV (pressure controlled ventilation) in COPD	HMV NOS	93 Patients aged 65.5±8, 30.1% female	COPD
				HMV (pressure controlled ventilation) in restrictive thoracic disease		38 Patients aged 64.9±11.3, 57.9% female	TRD

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Howard, 2016 <sup>36</sup>	RCT in Australia, 11/01/2011 to 12/31/2013	Moderate ROB	Inclusion: Primary OHS diagnosis  Exclusion: Other conditions contributing to hypoventilation.	BPAP ST	BPAP Harmony; Philips Respironics (USA) (FDA approved 510(k) clearance)  VPAP III STa; ResMed (Bella Vista, Australia) (FDA approved 510(k) clearance)	29 Patients aged 53.2±10.7, 51.7% female	OHS
				CPAP	CPAP Harmony; Philips Respironics (USA) (FDA approved 510(k) clearance)  VPAP III STa; ResMed (Bella Vista, Australia) (FDA approved 510(k) clearance)	31 Patients aged 52.9±10, 41.9% female	
Köhnlein, 2014 <sup>37</sup>	RCT in Germany and Austria, 10/29/2004 to 07/31/2011	High ROB	Inclusion: Clinically stable, hypercapnic stage IV COPD, no acute exacerbation  Exclusion: Thorax/lung abnormalities other than COPD, BMI≥35, other conditions resulting in hypercapnia, previously initiated NIPPV, malignant comorbidities, severe HF, unstable angina, severe arrhythmias.	BPAP ST + standard care	BPAP Models not reported, but all were BPAP machines from these manufacturers: ResMed (Martinsried, Germany), Weinmann (Hamburg, Germany, or Tyco Healthcare (Neubrug, Germany)	102 Patients aged 62.2±8.6, 36% female	COPD
				Standard care	No PAP	93 Patients aged 64.4±8.0, 40% female	
Marquez-Martin, 2014 <sup>38</sup>	RCT in Spain, 05/2007 to 09/2011	Moderate ROB	Inclusion: Adults with COPD, clinically stable, chronic RF with hypoxemia.	BPAP ST	BPAP BiPAP; Respironics (Murrysville, USA) (FDA approved 510(k) clearance)	15 Patients aged 69 (64-73)	COPD

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				Exercise program	No PAP	14 Patients aged 69 (64-73)	
				BPAP ST + exercise program	BPAP + no PAP	14 Patients aged 69 (64-73)	
Masa, 2000 <sup>39</sup>	Observational Prospective in Spain	Moderate ROB	Inclusion: OHS or kyphoscoliosis Exclusion: Apnea-hypopnea index >20 events/h.	HMV (volume cycled or pressure cycled) in OHS	HMV Monal DCC (Taema; Paris, France). (Not FDA approved)	22 Patients aged 61±14, 81.8% female	OHS
				HMV (volume cycled or pressure cycled) in kyphoscoliosis	Onyx Plus (Mallinckrodt SEFAM; Nancy, France). (Not FDA approved)	14 Patients aged 43±20, 50% female	TRD

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Masa, 2015 <sup>40, 41</sup>	RCT in Spain, 05/2009 to 03/2013	Moderate ROB	<p>Inclusion: OHS, no relevant COPD, severe OSA, absence of narcolepsy or restless leg syndrome, correctly executed 30 minute CPAP/NIPPV treatment test</p> <p>Exclusion: Psychophysical inability to complete questionnaires, severe chronic debilitating illness, severe chronic nasal obstruction.</p>	Mixed: HMV and BPAP mix (all with bilevel pressure with assured volume)	<p>HMV/BPAP Breas Vivo 40 (General Electric; England) (FDA approved 510(k) clearance)</p> <p>BiPAP AVAPS (Phylips-Respironics; Netherlands) (FDA approved 510(k) clearance)</p> <p>Trilogy 100 (Philips-Respironics; Netherlands) (FDA approved 510(k) clearance)</p> <p>VS Ultra (ResMed; Australia) (FDA approved 510(k) clearance)</p> <p>Monal T50 (Air Liquide; France) (Not FDA approved)</p> <p>Puritan Bennett 560 (Puritan Bennett; USA) (FDA approved 510(k) clearance)</p>	71 Patients aged 64±11, 65% female	OHS
				CPAP + lifestyle modifications	CPAP NOS	80 Patients aged 57±13, 47% female	
				Lifestyle modifications	No PAP	70 Patients aged 60±13, 56% female	

Author, Year	Study Country, Study Design, Study Period	Risk of Bias	Inclusion / Exclusion Criteria	Intervention and comparisons (Groups)	Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.	Patient Characteristics	Disease
Masa, 2016 <sup>42</sup>	RCT in Spain, 05/2009 to 03/2013	High ROB	<p>Inclusion: OHS (BMI <math>\geq</math> 30 kg/m<sup>2</sup>, no COPD, no NMD, no TRD, no narcooepsy, no restless leg syndrome), stable hypercapnic respiratory failure (daytime awake PaCO<sub>2</sub> <math>\geq</math> 45 mmHg, pH <math>\geq</math> 7.35 and no clinical worsening in prior 2 months), ability to use NIPPV in 30 minute trial period</p> <p>Exclusion: Psychophysical inability to complete questionnaires, severe chronic debilitating illness, severe chronic nasal obstruction, lack of informed consent.</p>	BPAP volume assured pressure support ventilation	BPAP volume assured pressure support ventilation	40 Patients aged 67 [IQR12], 75% female	OHS
				No PAP	No PAP	46 Patients aged 69 [IQR 15], 83% female	
McEvoy, 2009 <sup>43</sup>	RCT in Australia, 06/30/1998 to 05/15/2004	Moderate ROB	<p>Inclusion: Age &lt;80 years, severe COPD secondary to smoking, stable hypercapnic ventilatory failure, on LTOT <math>\geq</math>3 months, not currently smoking</p> <p>Exclusion: significant comorbidities (malignancies, left ventricular heart failure, unstable angina) likely affecting 2 year survival, severe psychiatric disorder impairing ability to comply to NIPPV, BMI &gt;40, evidence of sleep apnea.</p>	BPAP S + Oxygen	BPAP VPAP S mode; ResMed (Sydney, Australia) (FDA approved 510(k) clearance)	72 Patients aged 67.2 (IQR 65.3 to 69.1), 31% female	COPD
				Oxygen alone	No PAP	72 Patients aged 68.8 (IQR 67.1 to 70.5), 39% female	
Munoz, 2005 <sup>44</sup>	Observational Retrospective in Spain, 1997 to 2001	Moderate ROB	<p>Inclusion: Treated with non-invasive home volumetric ventilator, followup <math>\geq</math>1 year</p> <p>Exclusion: BPAP users.</p>	HMV volume assist control ventilation	HMV volume assist/control mode	45 Patients aged 65.1 $\pm$ 12.9, 48.9% female	Mixed (NMD, TRD)
				HMV volume control	HMV volume control mode	65 Patients aged 60.3 years $\pm$ 14.7 years, 46.2% female	



Author, Year	Study Country, Study Design, Study Period	Risk of Bias	Inclusion / Exclusion Criteria	Intervention and comparisons (Groups)	Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.	Patient Characteristics	Disease
Murphy, 2012 <sup>45</sup>	RCT in United Kingdom	Moderate ROB	Inclusion: BMI>40, absence of other identifiable hypoventilation cause  Exclusion: Inability to provide written consent.	BPAP AVAPS	BPAP BiPAP synchrony; Respironics (Murrysville, USA) (FDA approved 510(k) clearance)	25 Patients aged 53±9, 52% female	OHS
				BPAP ST		25 Patients aged 56±11, 56% female	
Murphy, 2017 <sup>46</sup>	RCT in United Kingdom, 2010 to 2015	High ROB	Inclusion: Persistent hypercapnia and hypoxemia, >30% sleep time <90% oxygen saturation, arterial pH >7.30 breathing room air  Exclusion: BMI >35, OSA, other RF causes.	BPAP ST + Home oxygen	BPAP Harmony 2; Philips Respironics (FDA approved 510(k) clearance)  VPAP III STa; ResMed (FDA approved 510(k) clearance)	57 Patients aged 66.4±10.2, 51% female	COPD
				Home oxygen		No PAP	
Nauffal, 2002 <sup>47</sup>	Observational Prospective in Spain, 01/1997 to 03/2000	Moderate ROB	Inclusion: Chronic hypoventilation due to kyphoscoliosis or NMD, moderate to severe restrictive ventilatory pattern, clinically stable.	BPAP NOS in kyphoscoliosis	BPAP DP-90; Taema (Paris, France) (Not FDA approved)	35 Patients aged 55.9, 40% female	TRD
				BPAP NOS in neuromuscular diseases		27 Patients aged 42.5, 48% female	NMD
Oscroft, 2010 <sup>48</sup>	Observational Retrospective in United Kingdom, 01/2000 to 12/2003	Moderate ROB	Inclusion: COPD diagnosis, smoking history >20 pack years, ventilatory failure with a daytime PaCO <sub>2</sub> > 7 kPa with a pH > 7.35 or nocturnal transcutaneous PaCO <sub>2</sub> > 9	BPAP ST started after AECOPD	BPAP NIPPY I, 2 or 3; B & D Electromedical (Stratford, United Kingdom) (Not FDA approved)	31 Patients aged 66±6, 49% female	COPD
						16 Patients aged	

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			kPa, hospital admission immediately prior to referral with clinical diagnosis of exacerbation of COPD  Exclusion: Age>80 years, other respiratory disease, BMI>35, significant OSA, tracheostomy, impaired left ventricular function.	BPAP ST started in stable patient without exacerbation		63±7	
Oscroft, 2010 <sup>49</sup>	RCT in United Kingdom, 07/01/2005 to 09/30/2006	Moderate ROB	Inclusion: COPD, FEV1<50%, 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)  Exclusion: >80 years old, other respiratory disease (interstitial lung disease, asthma, bronchiectasis, neuromuscular or restrictive chest wall disorders, left ventricular ejection fraction <40%)	NIPPV continue  NIPPV discontinue (no PAP)	BPAP NIPPY 2; B and D Electromedical (Stratford, United Kingdom) (Not FDA approved)  No PAP	5 Patients aged 69.2±7.4  5 Patients aged 58.6±6.3	COPD
Oscroft, 2014 <sup>50</sup>	RCT in United Kingdom, 09/2007 to 12/2011	High ROB	Inclusion: COPD diagnosis, smoking history >20 pack years, ventilatory failure with a daytime PaCO2 > 7 kPa with a pH > 7.35 or nocturnal transcutaneous PaCO2 > 9 kPa	BPAP IVAPS	BPAP Intelligent volume assured pressure support (iVAPS); ResMed (Bella Vista, Australia (FDA approved 510(k) clearance)	20 Patients aged 67.6±7.9, 55% female	COPD

Author, Year	Study Country, Study Design, Study Period	Risk of Bias	Inclusion / Exclusion Criteria	Intervention and comparisons (Groups)	Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.	Patient Characteristics	Disease
			Exclusion: Age>80 years, other respiratory disease, BMI>40, significant OSA.	BPAP ST	BPAP NIPPY 3; B and D Electromedical (Stratford, United Kingdom) (Not FDA approved)	20 Patients aged 67.4±8.2, 50% female	
Paone, 2014 <sup>51</sup>	Observational Prospective in Italy, 3/2007 and 1/2010	Low ROB	Inclusion: Acute RF needing NIPPV, clinical stability with symptoms of nocturnal Hypoventilation, FEV1 < 50% predicted, <20% improvement in FEV1 following bronchodilator and a ratio FEV1/FVC < 0.70  Exclusion: Significant comorbidities affecting survival (cancer, left ventricular heart failure, unstable angina), psychiatric disorders potentially affecting ability to undergo NIPPV, other chronic respiratory disease, history of OSA, BMI>40, systemic steroids therapy.	BPAP ST (PSV ST) + Home oxygen	BPAP Synchrony; Philips Respironics (Andover MA, USA) (FDA approved 510(k) clearance)  Neftis; Linde (Munich Germany) (Not FDA approved)	48 Patients, 56.2% female	COPD
				Home oxygen	No PAP	45 Patients aged 72 (IQR 66-78), 48.9% female	
Perez de Llano, 2005 <sup>52</sup>	Observational in Spain, 03/1995 to 12/2002	Low ROB	Inclusion: OHS, BMI > 30, PaCO2 ≥ 50 mmHg, FEV1/FVc < 70%, absence of other respiratory disorder such as kyphoscoliosis or diaphragmatic paralysis  Exclusion: <12 months followup	HMV/BPAP mix	HMV Home 2; Airox (Pau, France) (Not FDA approved)  BPAP DP-90; Taema (Paris, France) (Not FDA approved)  PV-102; Breas (Gothenburg, Sweden) (FDA approved 510(k) clearance)	54 patients aged 56 ± 13, 33.3% female	OHS

Author, Year	Study Country, Study Design, Study Period	Risk of Bias	Inclusion / Exclusion Criteria	Intervention and comparisons (Groups)	Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.	Patient Characteristics	Disease
				No PAP		15 patients aged 62 years, 66.7% female	
Pinto, 1995 <sup>53</sup>	Observational Prospective in Portugal	High ROB	Inclusion: Consecutive ALS patients with bulbar features  Exclusion: tracheotomised, refusal of attempts to prolong survival.	BPAP NOS	BPAP	10 Patients aged 60.66, 45% female between both groups	NMD
				No BPAP NOS	No PAP	10 Patients aged 57.22, 45% female between both groups	
Pinto, 2010 <sup>54</sup>	Observational Prospective in Portugal, 01/2003 to 09/2006	High ROB	Inclusion: No signs/symptoms of respiratory insufficiency, age 18-75 years  Exclusion: Gastrostomy, cognitive impairment, other significant disorders.	BPAP ST + weekly telemonitoring + standard care	BPAP Goodknight 425ST bi-level device; Tyco Healthcare Group LP (California, USA) (FDA approved 510(k) clearance)	20 Patients aged 62±12.90, 31.6% female	NMD
				BPAP ST + standard care		20 Patients aged 60±10, 30% female	
Piper, 2008 <sup>55</sup>	RCT in Australia	Low ROB	Inclusion: BMI≥30, stable awake compensated RF, absence of significant respiratory, NMD, or other disorder that could account for hypercapnia, no psychiatric illness capable of affecting participation, not currently treated with positive pressure therapy  Exclusion: Oxygen saturation below 80% continuously, acute rise in tcCO <sub>2</sub> during episodes of REM sleep, increase in afternoon to morning PaCO <sub>2</sub> ≥10mmHg.	BPAP S	BPAP	18 Patients aged 47±13, 50% female	OHS
				CPAP	CPAP	18 Patients aged 52±17, 22.2% female	

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Priou, 2010 <sup>56</sup>	Observational in France, 01/1995 to 12/2006	Moderate ROB	Inclusions: BMI $\geq$ 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%]).	BPAP started in stable hypercapnia	NR	92 patients aged 59.7 $\pm$ 12.9, 41.3% female	OHS
				BPAP started in acute exacerbation		38 patients aged 60.7 $\pm$ 16.3, 47.4% female	
Salturk, 2015 <sup>57</sup>	Observational Retrospective in Turkey, 01/2011 to 01/2012	Low ROB	Inclusion: Received NIPPV in ICU/home at least 4 hour/day, attending 1 month and 1 year followup  Exclusion: Disabled or unwilling to walk, clinical airway infection, current exacerbations, unstable cardiac arrhythmia.	BPAP ST COPD	BPAP COPD	37 Patients aged 65 $\pm$ 10, 8.1% female	COPD
				BPAP ST OHS	BPAP OHS	34 Patients aged 65 $\pm$ 8, 50% female	OHS
				BPAP ST Kyphoscoliosis	BPAP	20 Patients aged 46 $\pm$ 10, 45% female	TRD
				BPAP ST Diffuse Parenchymal Lung Disease	BPAP	14 Patients aged 62 $\pm$ 12, 21.4% female	Other
Sancho, 2014 <sup>58</sup>	Observational Retrospective in Spain/France, 03/2003 to 12/2007	Low ROB	Inclusion: Indication for NIPPV from presence of hypoventilation symptoms  Exclusion: Presence of previous pulmonary/airway disease, rapidly progressing disease with survival expectancy <1 month, severe frontotemporal dementia, NIPPV tolerance <4 consecutive hour/night.	HMV (volume assist control ventilation)	HMV PV 501; Breas Medical (Molndal, Sweden) (FDA approved 510(k) clearance)  Legendair; Airox (Pau, France) (Not FDA approved)	62 Patients aged 62.21 $\pm$ 8.81, 54.8% female	NMD
				BPAP ST	BPAP VPAP-III or VPAP-IV plus automatic ventilatory signal analysis (Reslink); Resmed (Sydney, Australia) (FDA approved 510(k) clearance)	82 Patients aged 63.80 $\pm$ 110.65, 24.4% female	

Author, Year	Study Country, Study Design, Study Period	Risk of Bias	Inclusion / Exclusion Criteria	Intervention and comparisons (Groups)	Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.	Patient Characteristics	Disease
Sancho, 2017 <sup>59</sup>	Observational Prospective in Spain, 01/1/2013 to 12/31/2015	High ROB	Inclusion: ALS (Escorial criteria), hospital admission  Exclusion: lung disease, <1 year life expectancy, NIV use <4 consecutive hours/night, slow disease progression (>3 yrs), severe frontotemporal dementia	HMV (volume assist control ventilation) in no/mild bulbar	HMV Vivo 50; Breas Medical (Molndal, Sweden) (FDA approved 510(k) clearance)	105 patients aged 64.05±9.11, 53% female	NMD
				HMV (volume assist control ventilation) in moderate/severe bulbar	Trilogy 100; Philips Respironics (Madrid, Spain) (FDA approved 510(k) clearance)	15 patients aged 64.05±9.11, 53% female	
				No device in no/mild bulbar	No PAP	14 patients aged 66.05±10.27, 70% female	
				No device in moderate/severe bulbar	No PAP	6 patients aged 66.05±10.27, 70% female	
Sanjuan-López, 2014 <sup>60</sup>	Observational Retrospective in Spain, 01/01/2000 to 12/31/2010	High ROB	Inclusion: Definitive ALS diagnosis by neurologist  Exclusion: Neuromuscular processes other than ALS, treatment in social welfare palliative center.	HMV (PSV or BPAP ST) started after outpatient pulmonary evaluation	HMV VS ultra and VS III; ResMed (FDA approved 510(k) clearance)	26 Patients aged 67.3±10.8, 50% female	NMD
				HMV (PSV or BPAP ST) started in an emergency situation without prior outpatient pulmonary evaluation		11 Patients aged 67.3±10.8, 50% female	

Author, Year	Study Country, Study Design, Study Period	Risk of Bias	Inclusion / Exclusion Criteria	Intervention and comparisons (Groups)	Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.	Patient Characteristics	Disease
Schonhofer, 2001 <sup>61</sup>	Observational Prospective in Germany	High ROB	<p>Inclusion: Chronic respiratory failure from thoracic disease &amp; hypercapnic, clinically stable, no significant difference in blood gas analysis parameters</p> <p>Exclusion: Rapidly progressive NMD, OHS, COPD, acute RF, severe acidosis</p>	Mixed: HMV (volume assist control ventilation) with change to BPAP ST if not tolerated	<p><u>HMV</u> Drager EV 800 (Drager; Lubeck, Germany) (Not FDA approved)</p> <p>PLV 100 (Respironics; Murrysville, USA) (FDA approved 510(k) clearance)</p> <p><u>BPAP</u> BP-T (Respironics Inc.; Murrysville, USA) (FDA approved 510(k) clearance)</p>	10 Patients aged 53.5±8.2, 50% female	TRD
				Standard care without HMV	No HMV	10 Patients aged 52.2±9.5, 50% female	
Sin, 2007 <sup>62</sup>	RCT in Canada,	Moderate ROB	<p>Inclusion: Diagnosis of COPD, age≥40 years, &gt;10 pack year smoking history</p> <p>Exclusion: Comorbidities making survival &lt;6 months unlikely, clinical history of left ventricular heart failure, apnea-hypopnea index &gt;20</p>	BPAP NOS + standard care	<u>BPAP</u> VPAP II, ResMed (Sydney, Australia) (FDA approved 510(k) clearance)	11 Patients aged 64.1±10.6, 64% female	COPD
				Sham BPAP (CPAP 4)	<u>Sham Device</u> S7Elite; ResMed (Sydney, Australia) (FDA approved 510(k) clearance)	10 Patients aged 66.6±9.7, 40% female	
Sivori, 2007 <sup>63</sup>	Observational Prospective in Argentina,	Moderate ROB	Inclusion: Diagnosis of ALS.	BPAP NOS + riluzole	BPAP + riluzole	18 Patients aged 53±15.46, 44.4% female	NMD

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	12/1999 to 12/2004			BPAP NOS	BPAP	11 Patients aged 56.4±15.5, 36.4% female	
				no BPAP, no riluzole	No PAP	42 Patients aged 52.3±11.4, 31% female	
				Riluzole	Riluzole	26 Patients aged 57.4±12.6, 38.5% female	
Struik, 2014 <sup>64</sup>	RCT in the Netherlands, 12/01/2007 to 07/01/2012	Moderate ROB	Inclusion: COPD (GOLD III/IV), >48 hours independence from ventilator support for acute RF, hypercapnia (PaCO <sub>2</sub> >6.0 kPa) daytime at rest	BPAP ST	BPAP BiPAP Synchrony; Respironics (Murrysville, USA) (FDA approved 510(k) clearance)	101 Patients aged 63.92±8.6, 59% female	COPD
				Standard care	No PAP	100 Patients aged 63.5±7.9, 58% female	
Tsolaki, 2008 <sup>65</sup>	Observational Prospective in Greece, 09/2005 to 12/2006	High ROB	Inclusion: Age ≤75 years, smoking history >20 pack years  Exclusion: Significant comorbidities (OSA, OHS, RF from disease other than COPD), important concomitant chronic systemic disorders, poor ventilator compliance, apnea-hypopnea index ≥10 episodes/hr.	BPAP ST	BPAP VPAP III ST; ResMed (Sydney, Australia) (FDA approved 510(k) clearance)	24 Patients aged 65.2±8.9, 29.2% female	COPD
				Standard care	No PAP	22 Patients aged 68.9±5.6, 36.4% female	
Tsolaki, 2011 <sup>66</sup>	Observational in Greece, dates not reported	Low ROB	Inclusion: symptoms of nocturnal hypoventilation (fatigue, dyspnea, morning headaches) combined with daytime hypercapnia (PaCO <sub>2</sub> ≥45mmHg for TRD and NMD, PaCO <sub>2</sub> ≥50mmHg for COPD).	BPAP ST in COPD	BPAP VPAP III ST; ResMed (Sydney, Australia) (FDA approved 510(k) clearance)	35 patients aged 67.1 ± 9.0, 20.0% female	COPD
				BPAP ST in TRD		17 patients aged 65.8 ± 7.8, 35.3% female	TRD



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			For OHS: BMI > 30, PaCO <sub>2</sub> >45mmHg, PaO <sub>2</sub> <70mmHg) in the absence of other diseases, persistent hypoventilation despite overnight trial of CPAP.  Exclusion: Apnea-hypopnea index >10/h (except patients with OHS), acute respiratory failure (pH < 7.35 and symptoms such as increasing cough, purulent sputum, need for antibiotics) or patients with an exacerbation during 4 weeks preceding, respiratory support other than NIV, poor compliance with NIV (i.e. mean use < 4 h/day) at the first follow-up visit	BPAP ST in NMD  BPAP ST in OHS		11 patients aged 62.8 ± 11.0, 63.6% female  28 patients aged 63.0 ± 9.9, 35.7% female	NMD  OHS
Vasquez, 2017 <sup>67</sup>	Observational Retrospective in USA, 01/1/2009 to 10/31/2014	Moderate ROB	Inclusion: At least 2 COPD claims, age ≥40 years, continuous enrollment 12 month prior & 6 months after claim	BPAP NOS CPAP NOS HMV NOS	BPAP CPAP HMV	9,156 Patients, 35.8% female 39,385 Patients, 45.1% female 315 Patients, 48.9% female	COPD
Vitacca, 2017 <sup>68</sup>	Observational Retrospective in Italy, 2008-2013	Moderate ROB	Inclusion: ALS NOS admitted to hospital, NIPPV use  Exclusion: dementia confirmed by Mini-Mental State Examination score <20, refusal of NIPPV	HMV/BPAP mix started in FVC ≥ 80% (early) HMV/BPAP mix started in FVC <80% (late)	HMV/BPAP mix	65 patients aged 62.62±11.34, 30.77% female 129 patients aged 64.66±11.33, 48.06% female	NMD
Windisch, 2006 <sup>69</sup>	Observational in Germany	Moderate ROB	Inclusion: Stable disease hospitalized for establishing NIPPV, matched controls	HMV (pressure controlled ventilation)	HMV PV401; Breas Medical AB (Moelnlycke, Sweden)	6 Patients aged 55.2±10.0, 16.7% female	COPD

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			Exclusion: Acute RF, signs of respiratory infection, intubated or tracheotomised previously in life, established on other ventilatory support prior to admission		(FDA approved 510(k) clearance)	6 Patients aged 61.7±11.3, 33% female	Mixed (TRD +OHS)
Zhou, 2017 <sup>70</sup>	RCT in China, 10/01/2015 to 05/31/2016	High ROB	Inclusion: Clinically stable, stage III/IV flow limitation & chronic hypercapnic, age > 40 years  Exclusion: Abnormalities of lung/thorax other than COPD, previously treated on NIPPV, OSA, severe HF, severe arrhythmias, unstable angina, malignant comorbidities, COPD with OSA overlap syndrome, impairments that could affect ability for followup.	BPAP ST	BPAP Flexo ST 30 NIV; Curative Co. (SuZhou, China) (Not FDA approved)	57 Patients aged 66.91±7.1, 36.8% female	COPD
				Standard care	No PAP	58 Patients aged 68.47±6.57, 39.7% female	

Note: ± denotes standard deviation.

AECOPD: acute exacerbation of chronic obstructive pulmonary disease, ALS: amyotrophic lateral sclerosis, AVAPS: average volume assured pressure support, BMI: Body Mass Index, BPAP: Bilevel Positive Airway Pressure, CHRF: chronic hypercapnic respiratory failure, COPD: chronic obstructive pulmonary disease, CPAP: Continuous Positive Airway Pressure, FDA: Food and Drug Administration, FEV1: Forced expiratory volume in one second, FVC: Forced vital capacity, HF: heart failure, HMV: Home Mechanical Ventilation, ICU: Intensive Care Unit, IQR: Interquartile range, kPa: kilopascal, LTOT: Long term oxygen, 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, PAP: positive airway pressure, pH: potential of hydrogen, PSV: Pressure support ventilation, RCT: randomized controlled trial, REM: rapid eye movement, ROB: risk of bias, RF: Respiratory Failure, S: spontaneous mode, SE: standard error, SpO<sub>2</sub>: Blood oxygen saturation level, ST: spontaneous/timed breath mode, tcCO<sub>2</sub>/PtCO<sub>2</sub>: transcutaneous carbon dioxide, TRD: Thoracic Restrictive Disorder, TWD: Thoracic Wall Diseases, USA: United States of America