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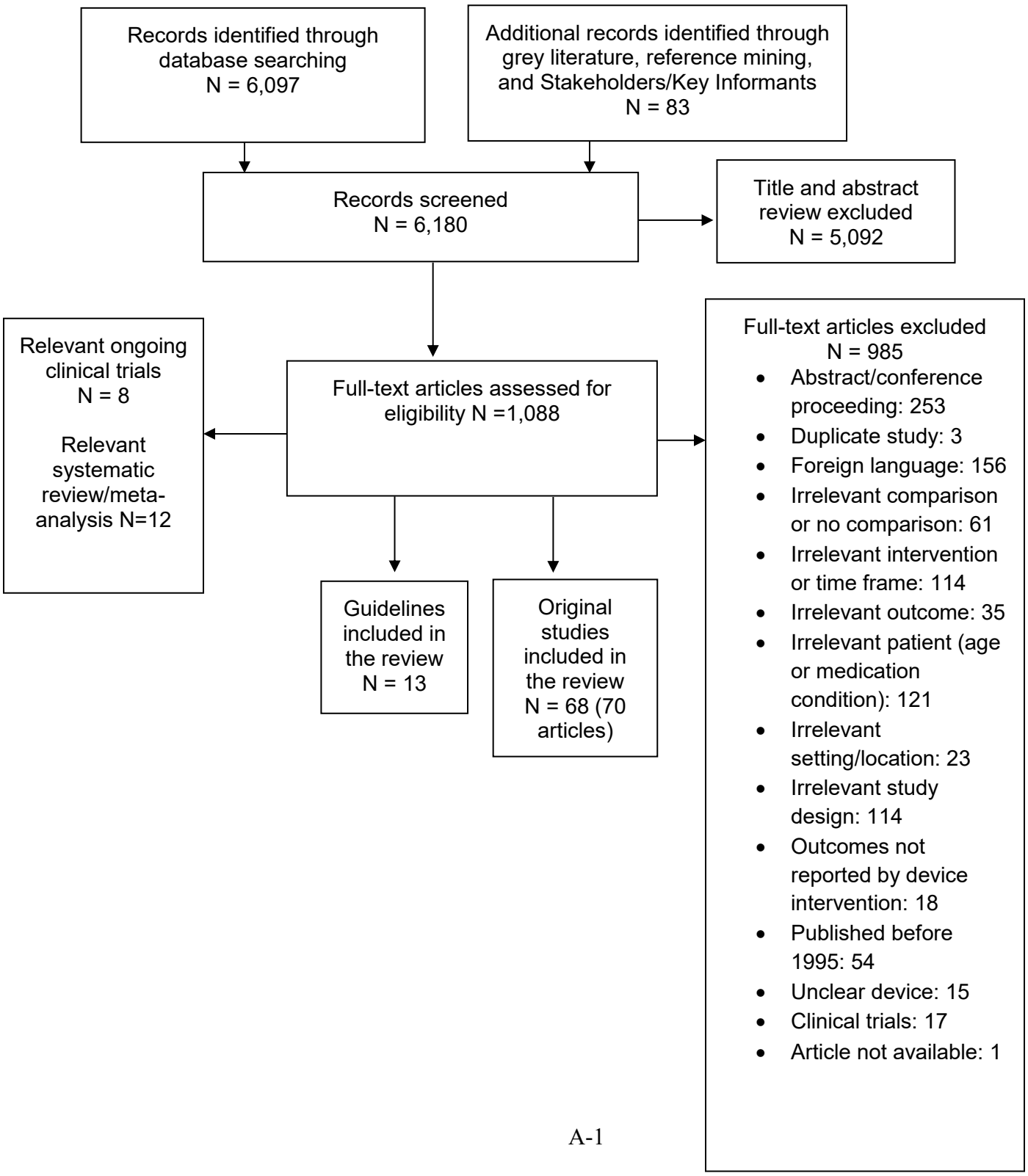
Appendix A. Flow Chart

Figure A.1. Flow chart



Appendix B. Search Strategy

Search Strategy 1

Ovid

Database(s): Embase 1988 to 2018 Week 26, EBM Reviews - Cochrane Central Register of Controlled Trials May 2018, EBM Reviews - Cochrane Database of Systematic Reviews 2005 to June 20, 2018, Ovid MEDLINE(R) Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Search Strategy:

#	Searches
1	*noninvasive ventilation/ or exp *positive-pressure respiration/
2	(BiPAP or BPAP or CPAP or "noninvasive positive pressure ventilation" or "non-invasive positive pressure ventilation" or "noninvasive ventilation" or "non-invasive ventilation" or NPPV or "Positive Airway Pressure*" or "positive end-expiratory pressure*").ti.
3	1 or 2
4	*Amyotrophic Lateral Sclerosis/
5	*Bronchiectasis/
6	*Cystic Fibrosis/
7	*Hypercapnia/
8	*Hypoventilation/
9	*Idiopathic Pulmonary Fibrosis/
10	*Lung Diseases, Interstitial/
11	*Pulmonary Fibrosis/
12	*Idiopathic Pulmonary Fibrosis/
13	*Kyphosis/
14	*Obesity/
15	*Respiratory Insufficiency/
16	*Scoliosis/
17	*Spinal Cord Injuries/
18	*Obesity Hypoventilation Syndrome/
19	*respiratory failure/
20	*Lung Diseases, Obstructive/
21	*Pulmonary Disease, Chronic Obstructive/
22	*Neuromuscular Diseases/
23	*Motor Neuron Disease/
24	*Muscular Atrophy, Spinal/
25	*Muscular Diseases/
26	*Muscular Disorders, Atrophic/
27	*Myopathies, Structural, Congenital/
28	*Myositis/
29	*Myotonic Disorders/
30	("Amyotrophic lateral sclerosis" or "Atrophic Muscular Disorder*" or Bronchiectasis or "Chronic Obstructive Pulmonary Disease*" or "congenital structual myopath*" or "Cystic Fibrosis" or hypercapnia or hypoventilation or "Interstitial Lung Disease*" or kyphosis or "Motor Neuron Disease*" or "Muscular Disease*" or Myositis or "Myotonic Disorder*" or

"Neuromuscular Disease*" or Obesity or "obstructive lung disease*" or "Pulmonary fibrosis" or "respiratory failure" or "respiratory insufficiency" or scoliosis or "Spinal Cord Injur*" or "Spinal Muscular Atrophy" or "structural congenital myopath*").ti.
31 or/4-30
32 3 and 31
33 limit 32 to ("all adult (19 plus years)" or "young adult (19 to 24 years)" or "adult (19 to 44 years)" or "young adult and adult (19-24 and 19-44)" or "middle age (45 to 64 years)" or "middle aged (45 plus years)" or "all aged (65 and over)" or "aged (80 and over)") [Limit not valid in Embase,CCTR,CDSR; records were retained]
34 limit 33 to (adult <18 to 64 years> or aged <65+ years>) [Limit not valid in CCTR,CDSR,Ovid MEDLINE(R),Ovid MEDLINE(R) Daily Update,Ovid MEDLINE(R) In-Process,Ovid MEDLINE(R) Publisher; records were retained]
35 limit 32 to ("all infant (birth to 23 months)" or "all child (0 to 18 years)" or "newborn infant (birth to 1 month)" or "infant (1 to 23 months)" or "preschool child (2 to 5 years)" or "child (6 to 12 years)" or "adolescent (13 to 18 years)") [Limit not valid in Embase,CCTR,CDSR; records were retained]
36 limit 35 to (embryo or infant or child or preschool child <1 to 6 years> or school child <7 to 12 years> or adolescent <13 to 17 years>) [Limit not valid in CCTR,CDSR,Ovid MEDLINE(R),Ovid MEDLINE(R) Daily Update,Ovid MEDLINE(R) In-Process,Ovid MEDLINE(R) Publisher; records were retained]
37 36 not 34
38 32 not 37
39 exp Guideline/ or exp Practice Guideline/
40 exp meta analysis/
41 exp Meta-Analysis as Topic/
42 exp "systematic review"/
43 exp controlled study/
44 exp Randomized Controlled Trial/
45 exp triple blind procedure/
46 exp Double-Blind Method/
47 exp Single-Blind Method/
48 exp latin square design/
49 exp comparative study/
50 exp Cohort Studies/
51 exp longitudinal study/
52 exp retrospective study/
53 exp prospective study/
54 exp population research/
55 exp observational study/
56 clinical study/
57 exp Evaluation Studies/
58 exp quantitative study/
59 exp validation studies/
60 exp quasi experimental study/
61 exp field study/
62 in vivo study/

63 exp panel study/
64 exp prevention study/
65 exp replication study/
66 exp Feasibility Studies/
67 exp trend study/
68 exp correlational study/
69 exp case-control studies/
70 exp confidence interval/
71 exp regression analysis/
72 exp proportional hazards model/
73 ((evidence adj based) or (meta adj analys*) or (systematic* adj3 review*) or "consensus development" or guideline* or "position statement*" or (control* adj3 study) or (control* adj3 trial) or (randomized adj3 study) or (randomized adj3 trial) or (randomised adj3 study) or (randomised adj3 trial) or "pragmatic clinical trial" or (random* adj1 allocat*) or (doubl* adj blind*) or (doubl* adj mask*) or (singl* adj blind*) or (singl* adj mask*) or (tripl* adj blind*) or (tripl* adj mask*) or (trebl* adj blind*) or (trebl* adj mask*) or "latin square" or placebo* or nocebo* or multivariate or "comparative study" or "comparative survey" or "comparative analysis" or (intervention* adj2 study) or (intervention* adj2 trial) or crossover or "cross-over" or cohort* or "longitudinal study" or "longitudinal survey" or "longitudinal analysis" or "longitudinal evaluation" or longitudinal* or ((retrospective or "ex post facto") adj3 (study or survey or analysis or design)) or retrospectiv* or "prospective study" or "prospective survey" or "prospective analysis" or prospectiv* or (population adj3 (stud* or survey* or analys* or research)) or "concurrent study" or "concurrent survey" or "concurrent analysis" or "incidence study" or "incidence survey" or "incidence analysis" or (("follow-up" or followup) adj (stud* or survey or analysis)) or ((observation or observational) adj (study or survey or analysis)) or "clinical study" or "clinical trial" or "evaluation study" or "evaluation survey" or "evaluation analysis" or "quantitative study" or "quantitative analys*" or "numerical study" or "validation study" or "validation survey" or "validation analysis" or "quasi experimental study" or "quasi experimental analysis" or "quasiexperimental study" or "quasiexperimental analysis" or "field study" or "field survey" or "field analysis" or "in vivo study" or "in vivo analysis" or "panel study" or "panel survey" or "panel analysis" or ((prevention or preventive) adj3 (trial or study or analysis or survey)) or "replication study" or "replication analysis " or "replication trial" or "feasibility study" or "feasibility analysis" or "trend study" or "trend survey" or "trend analysis" or ((correlation* adj2 study) or (correlation* adj2 analys*)) or "case control study" or "case base study" or "case referrent study" or "case referent study" or "case referent study" or "case compeer study" or "case comparison study" or "matched case control" or "multicenter study" or "multi-center study" or "odds ratio" or "confidence interval" or (hazard* adj (model* or analys* or regression or ratio or ratios)) or "Cox model" or "Cox multivariate analyses" or "Cox multivariate analysis" or "Cox regression" or "Cox survival analyses" or "Cox survival analysis" or "Cox survival model" or "change analysis" or ((study or trial or random* or control*) and compar*)).mp,pt.
74 or/39-73
75 38 and 74
76 limit 75 to (editorial or erratum or letter or note or addresses or autobiography or bibliography or biography or blogs or comment or dictionary or directory or interactive tutorial or interview or lectures or legal cases or legislation or news or newspaper article or overall or

patient education handout or periodical index or portraits or published erratum or video-audio media or webcasts) [Limit not valid in Embase,CCTR,CDSR,Ovid MEDLINE(R),Ovid MEDLINE(R) Daily Update,Ovid MEDLINE(R) In-Process,Ovid MEDLINE(R) Publisher; records were retained]

77 from 76 keep 130-138

78 (75 not 76) or 77

79 limit 78 to yr="1995 -Current"

80 remove duplicates from 79

Scopus

1 TITLE(BiPAP or BPAP or CPAP or "noninvasive positive pressure ventilation" or "non-invasive positive pressure ventilation" or "noninvasive ventilation" or "non-invasive ventilation" or NPPV or "Positive Airway Pressure*" or "positive end-expiratory pressure*")

2 TITLE("Amyotrophic lateral sclerosis" or "Atrophic Muscular Disorder*" or Bronchiectasis or "Chronic Obstructive Pulmonary Disease*" or "congenital structual myopath*" or "Cystic Fibrosis" or hypercapnia or hypoventilation or "Interstitial Lung Disease*" or kyphosis or "Motor Neuron Disease*" or "Muscular Disease*" or Myositis or "Myotonic Disorder*" or "Neuromuscular Disease*" or Obesity or "obstructive lung disease*" or "Pulmonary fibrosis" or "respiratory failure" or "respiratory insufficiency" or scoliosis or "Spinal Cord Injur*" or "Spinal Muscular Atrophy" or "structural congenital myopath*")

3 TITLE-ABS-KEY((evidence W/1 based) or (meta W/1 analys*) or (systematic* W/3 review*) or "consensus development" or guideline* or "position statement*" or (control* W/3 study) or (control* W/3 trial) or (randomized W/3 study) or (randomized W/3 trial) or (randomised W/3 study) or (randomised W/3 trial) or "pragmatic clinical trial" or (random* W/1 allocat*) or (doubl* W/1 blind*) or (doubl* W/1 mask*) or (singl* W/1 blind*) or (singl* W/1 mask*) or (tripl* W/1 blind*) or (tripl* W/1 mask*) or (trebl* W/1 blind*) or (trebl* W/1 mask*) or "latin square" or placebo* or nocebo* or multivariate or "comparative study" or "comparative survey" or "comparative analysis" or (intervention* W/2 study) or (intervention* W/2 trial) or crossover or "cross-over" or cohort* or "longitudinal study" or "longitudinal survey" or "longitudinal analysis" or "longitudinal evaluation" or longitudinal* or ((retrospective or "ex post facto") W/3 (study or survey or analysis or design)) or retrospectiv* or "prospective study" or "prospective survey" or "prospective analysis" or prospectiv* or (population W/3 (stud* or survey* or analys* or research)) or "concurrent study" or "concurrent survey" or "concurrent analysis" or "incidence study" or "incidence survey" or "incidence analysis" or (("follow-up" or followup) W/1 (stud* or survey or analysis)) or ((observation or observational) W/1 (study or survey or analysis)) or "clinical study" or "clinical trial" or "evaluation study" or "evaluation survey" or "evaluation analysis" or "quantitative study" or "quantitative analys*" or "numerical study" or "validation study" or "validation survey" or "validation analysis" or "quasi experimental study" or "quasi experimental analysis" or "quasiexperimental study" or "quasiexperimental analysis" or "field study" or "field survey" or "field analysis" or "in vivo study" or "in vivo analysis" or "panel study" or "panel survey" or "panel analysis" or ((prevention or preventive) W/3 (trial or study or analysis or survey)) or "replication study" or "replication analysis " or "replication trial" or "feasibility study" or "feasibility analysis" or "trend study" or "trend survey" or "trend analysis" or ((correlation* W/2 study) or (correlation* W/2 analys*)) or "case control study" or "case base study" or "case referrent study" or "case referent study" or "case referent study" or "case compeer study" or "case comparison study" or "matched case control" or "multicenter

study" or "multi-center study" or "odds ratio" or "confidence interval" or (hazard* W/1 (model* or analys* or regression or ratio or ratios)) or "Cox model" or "Cox multivariate analyses" or "Cox multivariate analysis" or "Cox regression" or "Cox survival analyses" or "Cox survival analysis" or "Cox survival model" or "change analysis" or ((study or trial or random* or control*) and compar*))

4 PUBYEAR AFT 1994

5 1 and 2 and 3 and 4

6 TITLE-ABS-KEY(newborn* or neonat* or infant* or toddler* or child* or adolescent* or paediatric* or pediatric* or girl or girls or boy or boys or teen or teens or teenager* or preschooler* or "pre-schooler*" or preteen or preteens or "pre-teen" or "pre-teens" or youth or youths) AND NOT TITLE-ABS-KEY(adult or adults or "middle age" or "middle aged" OR elderly OR geriatric* OR "old people" OR "old person*" OR "older people" OR "older person*" OR "very old")

7 5 and not 6

8 DOCTYPE(le) OR DOCTYPE(ed) OR DOCTYPE(bk) OR DOCTYPE(er) OR DOCTYPE(no) OR DOCTYPE(sh)

9 7 and not 8

10 PMID(0*) OR PMID(1*) OR PMID(2*) OR PMID(3*) OR PMID(4*) OR PMID(5*) OR PMID(6*) OR PMID(7*) OR PMID(8*) OR PMID(9*)

11 9 and not 10

National Guidelines Clearinghouse

("Amyotrophic lateral sclerosis" or "Atrophic Muscular Disorder*" or Bronchiectasis or "Chronic Obstructive Pulmonary Disease*" or "congenital structual myopath*" or "Cystic Fibrosis" or hypercapnia or hypoventilation or "Interstitial Lung Disease*" or kyphosis or "Motor Neuron Disease*" or "Muscular Disease*" or Myositis or "Myotonic Disorder*" or "Neuromuscular Disease*" or Obesity or "obstructive lung disease*" or "Pulmonary fibrosis" or "respiratory failure" or "respiratory insufficiency" or scoliosis or "Spinal Cord Injur*" or "Spinal Muscular Atrophy" or "structural congenital myopath*") AND (BiPAP OR BPAP OR CPAP OR "noninvasive positive pressure ventilation" OR "non-invasive positive pressure ventilation" OR "noninvasive ventilation" OR "non-invasive ventilation" OR NPPV OR "Positive Airway Pressure*" OR "positive end-expiratory pressure*")
Limited to Adults

ClinicalTrials.gov

All limited to Adults

("Amyotrophic lateral sclerosis" or "Atrophic Muscular Disorder*" or Bronchiectasis or "Chronic Obstructive Pulmonary Disease*" or "congenital structual myopath*" or "Cystic Fibrosis" or hypercapnia or hypoventilation or "Interstitial Lung Disease*") AND (BiPAP or BPAP or CPAP or "noninvasive positive pressure ventilation" or "non-invasive positive pressure ventilation" or NPPV or "Positive Airway Pressure*" or "positive end-expiratory pressure*") (kyphosis or "Motor Neuron Disease*" or "Muscular Disease*" or Myositis or "Myotonic Disorder*" or "Neuromuscular Disease*" or Obesity or "obstructive lung disease*" or "Pulmonary fibrosis" or "respiratory failure" or "respiratory insufficiency") AND (BiPAP or BPAP or CPAP or "noninvasive positive pressure ventilation" or "non-invasive positive pressure ventilation" or NPPV or "Positive Airway Pressure*" or "positive end-expiratory pressure*")

(scoliosis or "Spinal Cord Injur*" or "Spinal Muscular Atrophy" or "structural congenital myopath*") AND (BiPAP or BPAP or CPAP or "noninvasive positive pressure ventilation" or "non-invasive positive pressure ventilation" or NPPV or "Positive Airway Pressure*" or "positive end-expiratory pressure*")

Search Strategy 2

Ovid

Database(s): Embase 1988 to 2018 Week 26, EBM Reviews - Cochrane Central Register of Controlled Trials May 2018, EBM Reviews - Cochrane Database of Systematic Reviews 2005 to June 20, 2018, Ovid MEDLINE(R) Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Search Strategy:

#	Searches
1	exp Home Care Services/
2	(((domestic or home or domiciliary) adj3 (residence or residences or setting or settings or care or nurs* or help or service* or treatment* or therap* or "respiratory care" or "respiratory treatment*" or "respiratory therap*" or "respiratory service*" or "respiratory assist*" or ventilat*)) or "assisted living" or homecare).ti,ab,hw,kw.
3	"nursing home*".ti,ab,hw,kw.
4	(1 or 2) not 3
5	exp Respiration, Artificial/
6	(((facial or face or nasal) adj3 mask*) or ((respiration* or respiratory or breathing) adj3 (assist* or controlled or mechanical)) or "artificial respiration*" or BiPAP or CPAP or "Fluidic Breathing Assister" or HMV or IPPB or IPPV or NIAV or NIV or NPPV or "Oxygen Regulator*" or PAP or PAV or "Portable Oxygen" or "Positive Airway Pressure*" or "positive end-expiratory pressure*" or "positive pressure*" or respirator or respirators or "Respiratory insufficiency" or Tracheostom* or ventilation or ventilator*).ti,ab,hw,kw.
7	5 or 6
8	4 and 7
9	limit 8 to ("all adult (19 plus years)" or "young adult (19 to 24 years)" or "adult (19 to 44 years)" or "young adult and adult (19-24 and 19-44)" or "middle age (45 to 64 years)" or "middle aged (45 plus years)" or "all aged (65 and over)" or "aged (80 and over)") [Limit not valid in Embase,CCTR,CDSR; records were retained]
10	limit 9 to (adult <18 to 64 years> or aged <65+ years>) [Limit not valid in CCTR,CDSR,Ovid MEDLINE(R),Ovid MEDLINE(R) Daily Update,Ovid MEDLINE(R) In-Process,Ovid MEDLINE(R) Publisher; records were retained]
11	limit 8 to ("all infant (birth to 23 months)" or "all child (0 to 18 years)" or "newborn infant (birth to 1 month)" or "infant (1 to 23 months)" or "preschool child (2 to 5 years)" or "child (6 to 12 years)" or "adolescent (13 to 18 years)") [Limit not valid in Embase,CCTR,CDSR; records were retained]
12	limit 11 to (embryo or infant or child or preschool child <1 to 6 years> or school child <7 to 12 years> or adolescent <13 to 17 years>) [Limit not valid in CCTR,CDSR,Ovid MEDLINE(R),Ovid MEDLINE(R) Daily Update,Ovid MEDLINE(R) In-Process,Ovid MEDLINE(R) Publisher; records were retained]
13	12 not 10
14	8 not 13
15	exp Guideline/ or exp Practice Guideline/

16 exp meta analysis/
17 exp Meta-Analysis as Topic/
18 exp "systematic review"/
19 exp controlled study/
20 exp Randomized Controlled Trial/
21 exp triple blind procedure/
22 exp Double-Blind Method/
23 exp Single-Blind Method/
24 exp latin square design/
25 exp comparative study/
26 exp Cohort Studies/
27 exp longitudinal study/
28 exp retrospective study/
29 exp prospective study/
30 exp population research/
31 exp observational study/
32 clinical study/
33 exp Evaluation Studies/
34 exp quantitative study/
35 exp validation studies/
36 exp quasi experimental study/
37 exp field study/
38 in vivo study/
39 exp panel study/
40 exp prevention study/
41 exp replication study/
42 exp Feasibility Studies/
43 exp trend study/
44 exp correlational study/
45 exp case-control studies/
46 exp confidence interval/
47 exp regression analysis/
48 exp proportional hazards model/
49 ((evidence adj based) or (meta adj analys*) or (systematic* adj3 review*) or "consensus development" or guideline* or "position statement*" or (control* adj3 study) or (control* adj3 trial) or (randomized adj3 study) or (randomized adj3 trial) or (randomised adj3 study) or (randomised adj3 trial) or "pragmatic clinical trial" or (random* adj1 allocat*) or (doubl* adj blind*) or (doubl* adj mask*) or (singl* adj blind*) or (singl* adj mask*) or (tripl* adj blind*) or (tripl* adj mask*) or (trebl* adj blind*) or (trebl* adj mask*) or "latin square" or

placebo* or nocebo* or multivariate or "comparative study" or "comparative survey" or "comparative analysis" or (intervention* adj2 study) or (intervention* adj2 trial) or crossover or "cross-over" or cohort* or "longitudinal study" or "longitudinal survey" or "longitudinal analysis" or "longitudinal evaluation" or longitudinal* or ((retrospective or "ex post facto") adj3 (study or survey or analysis or design)) or retrospectiv* or "prospective study" or "prospective survey" or "prospective analysis" or prospectiv* or (population adj3 (stud* or survey* or analys* or research)) or "concurrent study" or "concurrent survey" or "concurrent analysis" or "incidence study" or "incidence survey" or "incidence analysis" or (("follow-up" or followup) adj (stud* or survey or analysis)) or ((observation or observational) adj (study or survey or analysis)) or "clinical study" or "clinical trial" or "evaluation study" or "evaluation survey" or "evaluation analysis" or "quantitative study" or "quantitative analys*" or "numerical study" or "validation study" or "validation survey" or "validation analysis" or "quasi experimental study" or "quasi experimental analysis" or "quasiexperimental study" or "quasiexperimental analysis" or "field study" or "field survey" or "field analysis" or "in vivo study" or "in vivo analysis" or "panel study" or "panel survey" or "panel analysis" or ((prevention or preventive) adj3 (trial or study or analysis or survey)) or "replication study" or "replication analysis " or "replication trial" or "feasibility study" or "feasibility analysis" or "trend study" or "trend survey" or "trend analysis" or ((correlation* adj2 study) or (correlation* adj2 analys*)) or "case control study" or "case base study" or "case referrent study" or "case referent study" or "case referent study" or "case compeer study" or "case comparison study" or "matched case control" or "multicenter study" or "multi-center study" or "odds ratio" or "confidence interval" or (hazard* adj (model* or analys* or regression or ratio or ratios)) or "Cox model" or "Cox multivariate analyses" or "Cox multivariate analysis" or "Cox regression" or "Cox survival analyses" or "Cox survival analysis" or "Cox survival model" or "change analysis" or ((study or trial or random* or control*) and compar*)).mp,pt.

50 or/15-49

51 14 and 50

limit 51 to (editorial or erratum or letter or note or addresses or autobiography or bibliography or biography or blogs or comment or dictionary or directory or interactive tutorial or interview or lectures or legal cases or legislation or news or newspaper article or overall or

52 patient education handout or periodical index or portraits or published erratum or video-audio media or webcasts) [Limit not valid in Embase,CCTR,CDSR,Ovid MEDLINE(R),Ovid MEDLINE(R) Daily Update,Ovid MEDLINE(R) In-Process,Ovid MEDLINE(R) Publisher; records were retained]

53 from 52 keep 45-48

54 (51 not 52) or 53

55 remove duplicates from 54

Scopus

1 TITLE-ABS-KEY(((domestic or home or domiciliary) W/3 (residence or residences or setting or settings or care or nurs* or help or service* or treatment* or therap* or "respiratory care" or "respiratory treatment*" or "respiratory therap*" or "respiratory service*" or "respiratory assist*" or ventilat*)) OR "assisted living" OR homecare or HMV)

2 TITLE-ABS-KEY(((facial or face or nasal) W/3 mask*) OR ((respiration* or respiratory or breathing) W/3 (assist* or controlled or mechanical)) OR "artificial respiration*" OR BiPAP OR CPAP OR "Fluidic Breathing Assister" OR HMV OR IPPB OR IPPV OR NIAV OR NIV OR NPPV OR "Oxygen Regulator*" OR PAP OR PAV OR "Portable Oxygen" OR "Positive Airway Pressure*" OR "positive end-expiratory pressure*" OR "positive pressure*" OR respirator OR respirators OR "Respiratory insufficiency" OR Tracheostom* OR ventilation OR ventilator*)

3 TITLE-ABS-KEY((evidence W/1 based) or (meta W/1 analys*) or (systematic* W/3 review*) or "consensus development" or guideline* or "position statement*" or (control* W/3 study) or (control* W/3 trial) or (randomized W/3 study) or (randomized W/3 trial) or (randomised W/3 study) or (randomised W/3 trial) or "pragmatic clinical trial" or (random* W/1 allocat*) or (doubl* W/1 blind*) or (doubl* W/1 mask*) or (singl* W/1 blind*) or (singl* W/1 mask*) or (tripl* W/1 blind*) or (tripl* W/1 mask*) or (trebl* W/1 blind*) or (trebl* W/1 mask*) or "latin square" or placebo* or nocebo* or multivariate or "comparative study" or "comparative survey" or "comparative analysis" or (intervention* W/2 study) or (intervention* W/2 trial) or crossover or "cross-over" or cohort* or "longitudinal study" or "longitudinal survey" or "longitudinal analysis" or "longitudinal evaluation" or longitudinal* or ((retrospective or "ex post facto") W/3 (study or survey or analysis or design)) or retrospectiv* or "prospective study" or "prospective survey" or "prospective analysis" or prospectiv* or (population W/3 (stud* or survey* or analys* or research)) or "concurrent study" or "concurrent survey" or "concurrent analysis" or "incidence study" or "incidence survey" or "incidence analysis" or (("follow-up" or followup) W/1 (stud* or survey or analysis)) or ((observation or observational) W/1 (study or survey or analysis)) or "clinical study" or "clinical trial" or "evaluation study" or "evaluation survey" or "evaluation analysis" or "quantitative study" or "quantitative analys*" or "numerical study" or "validation study" or "validation survey" or "validation analysis" or "quasi experimental study" or "quasi experimental analysis" or "quasiexperimental study" or "quasiexperimental analysis" or "field study" or "field survey" or "field analysis" or "in vivo study" or "in vivo analysis" or "panel study" or "panel survey" or "panel analysis" or ((prevention or preventive) W/3 (trial or study or analysis or survey)) or "replication study" or "replication analysis " or "replication trial" or "feasibility study" or "feasibility analysis" or "trend study" or "trend survey" or "trend analysis" or ((correlation* W/2 study) or (correlation* W/2 analys*)) or "case control study" or "case base study" or "case referrent study" or "case referent study" or "case referent study" or "case compeer study" or "case comparison study" or "matched case control" or "multicenter study" or "multi-center study" or "odds ratio" or "confidence interval" or (hazard* W/1 (model* or analys* or regression or ratio or ratios)) or "Cox model" or "Cox multivariate analyses" or "Cox multivariate analysis" or "Cox regression" or "Cox survival analyses" or "Cox survival analysis" or "Cox survival model" or "change analysis" or ((study or trial or random* or control*) and compar*))

4 1 and 2 and 3

5 TITLE-ABS-KEY(newborn* or neonat* or infant* or toddler* or child* or adolescent* or paediatric* or pediatric* or girl or girls or boy or boys or teen or teens or teenager* or preschooler* or "pre-schooler*" or preteen or preteens or "pre-teen" or "pre-teens" or youth or youths) AND NOT TITLE-ABS-KEY(adult or adults or "middle age" or "middle aged" OR elderly OR

geriatric* OR "old people" OR "old person*" OR "older people" OR "older person*" OR "very old")

6 4 and not 5

7 DOCTYPE(le) OR DOCTYPE(ed) OR DOCTYPE(bk) OR DOCTYPE(er) OR DOCTYPE(no) OR DOCTYPE(sh)

8 6 and not 7

9 PMID(0*) OR PMID(1*) OR PMID(2*) OR PMID(3*) OR PMID(4*) OR PMID(5*) OR PMID(6*) OR PMID(7*) OR PMID(8*) OR PMID(9*)

10 8 and not 9

National Guidelines Clearinghouse

((home OR domestic OR domiciliary or homecare or "assisted living" or HMV) AND (BiPAP OR CPAP OR "face mask*" OR "facial mask*" OR "Fluidic Breathing Assister" OR HMV OR IPPB OR IPPV OR "nasal mask*" OR NIAV OR NIV OR NPPV OR "Oxygen Regulator*" OR PAP OR PAV OR "Portable Oxygen" OR "Positive Airway Pressure*" OR "positive end-expiratory pressure*" OR "positive pressure*" OR respirat* OR Tracheostom* OR ventilat*)) NOT "nursing home*"

ClinicalTrials.gov

All limited to Adults

((domestic OR home OR domiciliary OR homecare OR "assisted living") NOT "nursing home") AND ((facial OR face OR nasal) AND mask*)

((domestic OR home OR domiciliary OR homecare OR "assisted living") NOT "nursing home") AND ((respiration* OR respiratory OR breathing) AND (assist* OR controlled OR mechanical))

((domestic OR home OR domiciliary OR homecare OR "assisted living") NOT "nursing home") AND ("artificial respiration*" OR BiPAP OR CPAP OR "Fluidic Breathing Assister" OR HMV OR IPPB)

((domestic OR home OR domiciliary OR homecare OR "assisted living") NOT "nursing home") AND (IPPV OR NIAV OR NIV OR NPPV OR "Oxygen Regulator*" OR PAP OR PAV OR "Portable Oxygen")

((domestic OR home OR domiciliary OR homecare OR "assisted living") NOT "nursing home") AND ("Positive Airway Pressure*" OR "positive end-expiratory pressure*" OR "positive pressure*" OR respirator)

((domestic OR home OR domiciliary OR homecare OR "assisted living") NOT "nursing home") AND (respirators OR "Respiratory insufficiency" OR Tracheostom* OR ventilation OR ventilator*)

Appendix C. Excluded Studies

1. [Guidelines for home mechanical ventilation. Swiss Association for the control of Tuberculosis and Lung Diseases (ASTP). Swiss Society of Pneumology (SSP)]. Schweizerische Medizinische Wochenschrift Journal Suisse de Medecine. 1996 Dec 28;126(51-52):2245-50. PMID: 9011937. [Foreign language study].

2. [Guidelines for indications and implementation of intermittent self-ventilation. German Society of Pneumology]. Pneumologie. 1994 May;48 Suppl 1:331-3. PMID: 8084877. [Foreign language study].

3. [Guidelines for mechanical home ventilation. SVTL (Swiss Society against Tuberculosis and Lung Diseases). SGP (Swiss Society for Pneumology)]. Schweizerische Medizinische Wochenschrift Journal Suisse de Medecine. 1996 Dec 14;126(50):2191-6. PMID: 9005530. [Foreign language study].

4. [Searching the literature for non-invasive positive pressure ventilation for neuromuscular diseases]. Revue des Maladies Respiratoires. 2006 Nov;23(5 Pt 4):14S1-S3. PMID: 17151546. [Foreign language study].

5. A trial comparing artificial noses and heat exchanges during assisted ventilation by tracheotomy in the home. [French]. Revue des Maladies Respiratoires. 1993;10(5):437-44. PMID: 23344441. [Published before 1995].

6. AARC (American Association for Respiratory Care) clinical practice guideline. Long-term invasive mechanical ventilation in the home.[Reprint in Respir Care. 2007 Aug;52(8):1056-62; PMID: 17715560]. Respiratory Care. 1995 Dec;40(12):1313-20. PMID: 10153257. [Irrelevant intervention (or time frame)].

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14. Adler D, Perrig S, Takahashi H, et al. Polysomnography in stable COPD under non-invasive ventilation to reduce patient-ventilator asynchrony and morning breathlessness. Sleep & Breathing. 2012 Dec;16(4):1081-90. PMID: 22051930. [Irrelevant comparison or no comparison].

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Launching of Nocturnal Non Invasive. 2010 October. PMID: NCT01225614. [Irrelevant patient (age, medical condition)].

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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[1]	Observational in United States, 03/1993 to 02/1996	Moderate ROB	Inclusion: ALS via el Escorial criteria; dyspnea on exertion or PaCO2 ≥ 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) BPAP BiPAP; Respironics Inc. (Murrysville, Pennsylvania, USA) (FDA approved 510(k) clearance)	18 Patients aged 61.5 ± 11.9, 22.2% female	NMD
				HMV/BPAP mix (intolerant)		21 Patients aged 61.8 ± 15.2, 33.3% female	
Benhamou, 1997[2]	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[3]	RCT in Italy, 03/2011-03/2014	Low ROB	Inclusion: ALS (definite via El Esocrial 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) BiPAP Synchrony II; Philips Respironics (Murrysville, PA, USA) (FDA approved 510(k) clearance)	25 patients aged 65.92±10.18, 32% female	NMD
				BPAP volume assured pressure support ventilation outpatient initiation		25 patients aged 61.26±8.64, 44% 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
Bhatt, 2013[4]	RCT in USA	High ROB	Inclusion: Stable COPD with 10 pack year smoking history, 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.	BPAP NOS	<u>BPAP</u> 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[5]	Observational, Prospective in Germany, 01/01/2011 to 12/31/2011	Moderate ROB	Inclusion: COPD (GOLD criteria NOS), PaCO2>7.0kPa, pH>7.35, stable disease (no exacerbation in 2 weeks prior) Exclusion: decompensated heart failure or "other conditions" that cause chronic respiratory failure, systemic corticosteroids	HMV (pressure controlled ventilation or pressure support ventilation)	<u>HMV</u> VS III; ResMed (Saime SA, France) (FDA approved 510(k) clearance)	51 patients aged 66.9 (SE 1.3), 37% female	COPD
			Inclusion: OHS (BMI >30kg/m2, chronic daytime hypercapnia PaCO2>6.7kPa, pH>7.35), symptoms of hypercapnia NOS) Exclusion: decompensated heart failure or "other conditions" that cause chronic respiratory failure			34 patients aged 65.4 (SE 1.8), 50% female	OHS
Borel, 2011[6]	RCT in Switzerland	High ROB	Inclusion: Age 20-75 years, BMI >30 Exclusion: Declined or presented any significant	BPAP ST	<u>BPAP</u> GoodKnight-425ST; Covidien (FDA approved 510(k) clearance)	19 Patients aged 58±11, 56% 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
			airway obstruction, scoliosis, cardiac failure, progressive NMD.	Lifestyle counseling	No PAP	18 Patients aged 54±6, 59% female	
Bourke, 2006[7]	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[8]	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, PaCO2 > 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	

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
Buyse, 2003[9]	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[10]	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[11]	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[12]	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[13]	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[14]	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	

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
Clini, 1998[15]	Observational Prospective in Italy, 12/1991 to 12/1994	Low ROB	Inclusion: Clinically stable, ≥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±6, 21.4% female	COPD
				Oxygen	No PAP	21 Patients aged 66±8, 33% female	
Clini, 2002[16]	RCT in Italy/France, 06/1996 to 01/2000	High ROB	Inclusion: Age ≤75 years, LTOT ≥ 6 months, dyspnea score (assessed by Medical Research Council) ≥2, FEV1 <1.5 liters, FEV1/FVC <60%, TLC ≥90% predicted, PaCO2 >6.6 kPa, PaO2 <7.8 kPa breathing room at rest Exclusion: 15% increase FEV1 after salbutamol, pH ≤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±7, 18% female	COPD
				LTOT	No PAP	47 Patients aged 66±14, 21.3% female	
Coco, 2006[17]	Observational Prospective in Italy, 10/1999 to 07/2003	Low ROB	Inclusion: Definite/probable ALS Exclusion: Primary lateral	BPAP ST (use ≥ 4 hours/day)	BPAP BiPAP; Respironics (Vitalaire, Italy) (FDA approved	44 Patients aged 62.3±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[18]	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[19]	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	5 Patients aged 66±6	
Domenéch-Clar, 2003[20]	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[21]	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	8 Patients	

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			ventilatory support prior to study.	HMV (PSV ST) (time period 2)	Cedex, France) (Not FDA approved)		
				HMV (pressure assist/control) (time period 2)			
Duiverman, 2011[22] [23]	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 ≥10h.	BPAP ST + pulmonary rehabilitation	<u>BPAP</u> BiPAP Synchrony; Respironics Inc. (FDA approved 510(k) clearance)	24 Patients aged 63±10, 33.3% female	COPD
				Pulmonary rehabilitation alone	No PAP	32 Patients aged 61±8, 46.9% female	
Duiverman, 2017[24]	RCT in Netherlands	High ROB	Inclusion: COPD (GOLD III or IV), ≥ 2 AECOPD with acute hypercapnic respiratory failure (pH<7.35) per year, daytime Inclusion: PaCO2 ≥6.7 kPa (50 mmHg) or nocturnal PaCO2 ≥7.3 kPa (55 mmHg) or nighttime rise in PtCO2 ≥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)	<u>HMV</u> Vivo 50; Breas Medical (Molndal, Sweden) (FDA approved 510(k) clearance) <u>BPAP</u> Stellar 100; Resmed (Martinsried, Germany) (FDA approved 510(k) clearance)	Crossover – 11 patients aged 68.7±8.5, 54% female	COPD
				HMV/BPAP mix (pressure support ventilation) (low intensity)			
Durao, 2018[25]	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	<u>BPAP</u> VPAP ST S9; Resmed (FDA approved 510(k) clearance) VPAP ST STA;	62 patients aged 64.6±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[26]	Observational in Spain, 1988 to 12/2002	Moderate ROB	Inclusion: ALS NOS	HMV/BPAP mix in pre protocol group	<u>HMV</u> PLV-100; Life Care Products (FDA approved 510(k) clearance)	11 Patients aged 53 ± 11	NMD
				HMV/BPAP mix in post protocol group		48 Patients aged 62 ± 10	

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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[27]	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[28]	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[29]	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[30]	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[31]	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[32]	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[33]	RCT in Netherlands, 10/2008 to 10/2012	Moderate ROB	Inclusion: Chronic RF from NMD or thoracic cage disorder, orthopnea from diaphragm paralysis & daytime normocapnia also included Exclusion: Strictly COPD patients, not mask naive, acute RF, age < 18 years, invasive ventilation, nursing home residing. (77 total patients in both groups, 3 patients on volume control, 74 patients on pressure control)	HMV started at home pressure controlled ventilation with change to volume assist control ventilation if not tolerated	<u>HMV</u> 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[34]	Observational Retrospective in Germany, 01/2002 to 02/2008	High ROB	Inclusion: COPD, prolonged weaning from invasive mechanical ventilation Exclusion: Intubated from cardiogenic edema or cardiopulmonary resuscitation	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[35]	Observational Prospective in Germany	Low ROB	Inclusion: HMV initiated ≥3 months prior to study, undergone bioelectrical impedance analysis measurement, regularly readmitted for routine followup, all had CHRF Exclusion: OHS, progressive NMD, tracheostomy.	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[36]	RCT in Australia, 11/01/2011 to 12/31/2013	Moderate ROB	Inclusion: Primary OHS diagnosis Exclusion: Other conditions contributing to hypoventilation.	BPAP ST	<u>BPAP</u> 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	<u>CPAP</u> 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[37]	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	<u>BPAP</u> 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[38]	RCT in Spain, 05/2007 to 09/2011	Moderate ROB	Inclusion: Adults with COPD, clinically stable, chronic RF with hypoxemia.	BPAP ST	<u>BPAP</u> 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[39]	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	<u>HMV</u> Monal DCC (Taema; Paris, France). (Not FDA approved) Onyx Plus (Mallinckrodt SEFAM; Nancy, France). (Not FDA approved)	22 Patients aged 61±14, 81.8% female	OHS
				HMV (volume cycled or pressure cycled) in kyphoscoliosis		14 Patients aged 43±20, 50% female	TRD

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Masa, 2015[40, 41]	RCT in Spain, 05/2009 to 03/2013	Moderate ROB	Inclusion: OHS, no relevant COPD, severe OSA, absence of narcolepsy or restless leg syndrome, correctly executed 30 minute CPAP/NIPPV treatment test Exclusion: Psychophysical inability to complete questionnaires, severe chronic debilitating illness, severe chronic nasal obstruction.	Mixed: HMV and BPAP mix (all with bilevel pressure with assured volume)	HMV/BPAP Breas Vivo 40 (General Electric; England) (FDA approved 510(k) clearance) BiPAP AVAPS (Phylips-Respironics; Netherlands) (FDA approved 510(k) clearance) Trilogy 100 (Philips-Respironics; Netherlands) (FDA approved 510(k) clearance) VS Ultra (ResMed; Australia) (FDA approved 510(k) clearance) Monal T50 (Air Liquide; France) (Not FDA approved) Puritian Bennett 560 (Puritan Bennett; USA) (FDA approved 510(k) clearance)	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	

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Masa, 2016[42]	RCT in Spain, 05/2009 to 03/2013	High ROB	Inclusion: OHS (BMI ≥ 30 kg/m2, no COPD, no NMD, no TRD, no narcooepsy, no restless leg syndrome), stable hypercapnic respiratory failure (daytime awake PaCO2 ≥ 45 mmHg, pH ≥ 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 consent.	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[43]	RCT in Australia, 06/30/1998 to 05/15/2004	Moderate ROB	Inclusion: Age<80 years, severe COPD secondary to smoking, stable hypercapnic ventilatory failure, on LTOT ≥3 months, not currently smoking 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>40, evidence of sleep apnea.	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[44]	Observational Retrospective in Spain, 1997 to 2001	Moderate ROB	Inclusion: Treated with non-invasive home volumetric ventilator, followup ≥1 year Exclusion: BPAP users.	HMV volume assist control ventilation	HMV volume assist/control mode	45 Patients aged 65.1±12.9, 48.9% female	Mixed (NMD, TRD)
				HMV volume control	HMV volume control mode	65 Patients aged 60.3 years±14.7 years, 46.2% female	

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Murphy, 2012[45]	RCT in United Kingdom	Moderate ROB	Inclusion: BMI>40, absence of other identifiable hypoventilation cause Exclusion: Inability to provide written consent.	BPAP AVAPS	<u>BPAP</u> 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[46]	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	<u>BPAP</u> 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	59 Patients aged 67.1±9.0, 54% female	
Nauffal, 2002[47]	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	<u>BPAP</u> 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[48]	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 PaCO2 > 7 kPa with a pH > 7.35 or nocturnal transcutaneous PaCO2 > 9	BPAP ST started after AECOPD	<u>BPAP</u> 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	

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
			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[49]	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	BPAP NIPPY 2; B and D Electromedical (Stratford, United Kingdom) (Not FDA approved)	5 Patients aged 69.2±7.4	COPD
				NIPPV discontinue (no PAP)	No PAP	5 Patients aged 58.6±6.3	
Oscroft, 2014[50]	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[51]	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[52]	Observational in Spain, 03/1995 to 12/2002	Low ROB	Inclusion: OHS, BMI > 30, $PaCO_2 \geq 50$ 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[53]	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[54]	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	<u>BPAP</u> 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[55]	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 tcCO2 during episodes of REM sleep, increase in afternoon to morning PaCO2 ≥10mmHg.	BPAP S	BPAP	18 Patients aged 47±13, 50% female	OHS
				CPAP	CPAP	18 Patients aged 52±17, 22.2% 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
Priou, 2010[56]	Observational in France, 01/1995 to 12/2006	Moderate ROB	Inclusions: BMI ≥ 30 kg/m 2 and daytime hypercapnia (PaCO2> 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 1 to vital capacity ratio , 70%]).	BPAP started in stable hypercapnia	NR	92 patients aged 59.7 ± 12.9, 41.3% female	OHS
				BPAP started in acute exacerbation		38 patients aged 60.7 ± 16.3, 47.4% female	
Salturk, 2015[57]	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±10, 8.1% female	COPD
				BPAP ST OHS	BPAP OHS	34 Patients aged 65±8, 50% female	OHS
				BPAP ST Kyphoscoliosis	BPAP	20 Patients aged 46±10, 45% female	TRD
				BPAP ST Diffuse Parenchymal Lung Disease	BPAP	14 Patients aged 62±12, 21.4% female	Other
Sancho, 2014[58]	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±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±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[59]	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) Trilogy 100; Philips Respironics (Madrid, Spain) (FDA approved 510(k) clearance)	105 patients aged 64.05±9.11, 53% female	NMD
				HMV (volume assist control ventilation) in moderate/severe bulbar		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[60]	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[61]	Observational Prospective in Germany	High ROB	Inclusion: Chronic respiratory failure from thoracic disease & hypercapnic, clinically stable, no significant difference in blood gas analysis parameters Exclusion: Rapidly progressive NMD, OHS, COPD, acute RF, severe acidosis	Mixed: HMV (volume assist control ventilation) with change to BPAP ST if not tolerated	HMV Drager EV 800 (Drager; Lubeck, Germany) (Not FDA approved) PLV 100 (Respironics; Murrysville, USA) (FDA approved 510(k) clearance) BPAP BP-T (Respironics Inc.; Murrysville, USA) (FDA approved 510(k) clearance)	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[62]	RCT in Canada,	Moderate ROB	Inclusion: Diagnosis of COPD, age≥40 years, >10 pack year smoking history Exclusion: Comorbidities making survival <6 months unlikely, clinical history of left ventricular heart failure, apnea-hypopnea index >20	BPAP NOS + standard care	BPAP VPAP II, ResMed (Sydney, Australia) (FDA approved 510(k) clearance)	11 Patients aged 64.1±10.6, 64% female	COPD
				Sham BPAP (CPAP 4)	Sham Device S7Elite; ResMed (Sydney, Australia) (FDA approved 510(k) clearance)	10 Patients aged 66.6±9.7, 40% female	
Sivori, 2007[63]	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

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
	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[64]	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 (PaCO2 >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[65]	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	
				Standard care	No PAP	22 Patients aged 68.9±5.6, 36.4% female	COPD
Tsolaki, 2011[66]	Observational in Greece, dates not reported	Low ROB	Inclusion: symptoms of nocturnal hypoventilation (fatigue, dyspnea, morning headaches) combined with daytime hypercapnia (PaCO2 ≥45mmHg for TRD and NMD, PaCO2 ≥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

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
			For OHS: BMI> 30, PaCO2 >45mmHg, PaO2 <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		11 patients aged 62.8 ± 11.0, 63.6% female	NMD
				BPAP ST in OHS		28 patients aged 63.0 ± 9.9, 35.7% female	OHS
Vasquez, 2017[67]	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	BPAP	9,156 Patients, 35.8% female	COPD
				CPAP NOS	CPAP	39,385 Patients, 45.1% female	
				HMV NOS	HMV	315 Patients, 48.9% female	
Vitacca, 2017[68]	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	65 patients aged 62.62±11.34, 30.77% female	NMD
				HMV/BPAP mix started in FVC <80% (late)		129 patients aged 64.66±11.33, 48.06% female	
Windisch, 2006[69]	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

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: 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[70]	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, PaCO2: 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, SpO2: Blood oxygen saturation level, ST: spontaneous/timed breath mode, tcCO2/PtCO2: transcutaneous carbon dioxide, TRD: Thoracic Restrictive Disorder, TWD: Thoracic Wall Diseases, USA: United States of America

Appendix E. Risk of Bias

Table E.1. Risk of bias for RCTs (Cochrane ROB tool) for included studies

Author, Year	Sequence Generation	Allocation Concealment	Blinding of Participants, Personnel	Blinding of Outcome Assessors	Incomplete Outcome Data	Selective Outcome Reporting	Other Sources of Bias	Overall RoB
Bertella, 2017[3]	Low ROB	Low ROB	High ROB	Low ROB	Low ROB	Low ROB	Low ROB	Low ROB
Bhatt, 2013[4]	Low ROB	Low ROB	High ROB	Unclear	Low ROB	Low ROB	High ROB	High ROB
Borel, 2011[6]	Low ROB	Low ROB	High ROB	Low ROB	Low ROB	Unclear	High ROB	High ROB
Bourke, 2006[7]	Low ROB	Unclear	High ROB	High ROB	Low ROB	Unclear	Low ROB	High ROB
Casanova, 2000[10]	Low ROB	Unclear	High ROB	Low ROB	Unclear	Unclear	Unclear	High ROB
Cheung, 2010[12]	Low ROB	Low ROB	High ROB	Unclear	High ROB	Low ROB	Low ROB	High ROB
Chiang, 2003[13]	Low ROB	Unclear	High ROB	Unclear	Low ROB	Unclear	Low ROB	Moderate ROB
Clini, 2002[16]	Unclear	Low ROB	High ROB	Low ROB	High ROB	Low ROB	High ROB	High ROB
De Backer, 2011[19]	Unclear	Unclear	Unclear	Unclear	Low ROB	Unclear	Low ROB	Moderate ROB
Dreher, 2010[21]	Unclear	Unclear	High ROB	High ROB	Low ROB	Unclear	High ROB	High ROB
Duiverman, 2008, 2011[22, 23]	Low ROB	Unclear	High ROB	Unclear	Low ROB	Low ROB	Low ROB	Moderate ROB
Duiverman, 2017[24]	Unclear	Unclear	High ROB	High ROB	High ROB	Low ROB	High ROB	High ROB
Funk, 2010[27]	Low ROB	Unclear	High ROB	High ROB	Low ROB	Low ROB	Low ROB	Moderate ROB
Garrod, 2000[30]	Unclear	Low ROB	High ROB	Unclear	Low ROB	Unclear	Unclear	High ROB
Gay, 1996[31]	Unclear	Unclear	High ROB	High ROB	Low ROB	Unclear	Unclear	High ROB
Hazenberg, 2014[33]	Low ROB	Unclear	High ROB	High ROB	Low ROB	Low ROB	Low ROB	Moderate ROB
Howard, 2016[36]	Low ROB	Low ROB	Low ROB	Low ROB	Low ROB	Low ROB	High ROB	Moderate ROB
Köhnlein, 2014[37]	Low ROB	Low ROB	High ROB	Low ROB	Low ROB	Low ROB	High ROB	High ROB
Marquez-Martin, 2014[38]	Low ROB	Unclear	High ROB	Unclear	Low ROB	Unclear	Low ROB	Moderate ROB
Masa, 2015[40, 41]	Low ROB	Unclear	High ROB	Unclear	Low ROB	Unclear	Low ROB	Moderate ROB
Masa, 2016[42]	Unclear	Unclear	High ROB	Unclear	Low ROB	High ROB	Low ROB	High ROB
McEvoy, 2009[43]	Low ROB	Low ROB	High ROB	Unclear	Low ROB	Low ROB	Low ROB	Moderate ROB
Murphy, 2012[45]	Unclear	Low ROB	High ROB	Unclear	Low ROB	Low ROB	Low ROB	Moderate ROB
Murphy, 2017[46]	Low ROB	Low ROB	High ROB	Low ROB	Low ROB	Low ROB	High ROB	High ROB
Oscroft, 2010[49]	Low ROB	Unclear	High ROB	Unclear	Low ROB	Low ROB	Low ROB	Moderate ROB
Oscroft, 2014[50]	Low ROB	Low ROB	High ROB	High ROB	Low ROB	Unclear	High ROB	High ROB
Piper, 2008[55]	Low ROB	Low ROB	Unclear	Unclear	Low ROB	Low ROB	Low ROB	Low ROB
Sin, 2007[62]	Low ROB	Unclear	Low ROB	Low ROB	Unclear	Unclear	Unclear	Moderate ROB
Struik, 2014[64]	Low ROB	Unclear	High ROB	Unclear	Low ROB	Low ROB	High ROB	Moderate ROB
Zhou, 2017[70]	Low ROB	Low ROB	High ROB	Low ROB	Low ROB	Unclear	High ROB	High ROB

ROB: Risk of Bias

Table E.2. Risk of bias for observational studies (Newcastle-Ottawa Quality Assessment Scale) for included studies

Author, Year	Representativeness of the Study Population	Ascertainment of Exposure	Assessment of Outcome	Adequate of Followup	Conflict of Interest	Overall RoB
Aboussouan, 1997[1]	Low ROB	Low ROB	Unclear	Low ROB	Unclear	Moderate ROB
Benhamou, 1997[2]	High ROB	Low ROB	Low ROB	Low ROB	Unclear	High ROB
Blankenburg, 2017[5]	Low ROB	Low ROB	Low ROB	High ROB	Low ROB	Moderate ROB
Budweiser, 2007[8]	Low ROB	Low ROB	Low ROB	Low ROB	Low ROB	Low ROB
Buyse, 2003[9]	Low ROB	Low ROB	Low ROB	Low ROB	Unclear	Moderate ROB
Castillejo, 2014[11]	High ROB	Low ROB	Low ROB	Low ROB	Unclear	High ROB
Clini, 1996[14]	High ROB	Low ROB	Low ROB	Low ROB	Unclear	High ROB
Clini, 1998[15]	High ROB	Low ROB	Low ROB	Low ROB	Low ROB	Low ROB
Coco, 2006[17]	Low ROB	Low ROB	Low ROB	Low ROB	Unclear	Low ROB
Crespo, 2009[18]	High ROB	High ROB	High ROB	Low ROB	Unclear	High ROB
Doménech-Clar, 2003[20]	Low ROB	Low ROB	Low ROB	High ROB	Unclear	High ROB
Durao, 2018[25]	Low ROB	Low ROB	Low ROB	Unclear	Low ROB	Low ROB
Farrero, 2005[26]	Low ROB	Low ROB	Unclear	Low ROB	Unclear	Moderate ROB
Gad, 2014[28]	Low ROB	Low ROB	High ROB	Low ROB	Low ROB	Moderate ROB
Galli, 2014[29]	Low ROB	Low ROB	Low ROB	Low ROB	High ROB	Low ROB
Gonzalez-Bermejo, 2013[32]	High ROB	High ROB	High ROB	Low ROB	Unclear	High ROB
Heinemann, 2011[34]	Low ROB	Low ROB	High ROB	Low ROB	Unclear	High ROB
Hitzl, 2009[35]	Low ROB	Low ROB	Low ROB	Low ROB	Low ROB	Low ROB
Masa, 2000 [39]	High ROB	Low ROB	Low ROB	Low ROB	Low ROB	Moderate ROB
Munoz, 2005[44]	Low ROB	Low ROB	Low ROB	Low ROB	Unclear	Moderate ROB
Nauffal, 2002[47]	Low ROB	Low ROB	Low ROB	Low ROB	Unclear	Moderate ROB
Oscroft, 2010[48]	High ROB	Low ROB	Low ROB	Low ROB	Unclear	Moderate ROB
Paone, 2014[51]	Low ROB	Low ROB	Low ROB	High ROB	Low ROB	Low ROB
Perez de Llano, 2005[52]	Low ROB	Low ROB	Low ROB	Low ROB	Unclear	Low ROB
Pinto, 1995[53]	High ROB	Low ROB	Low ROB	Low ROB	Unclear	High ROB
Pinto, 2010[54]	Low ROB	Low ROB	Low ROB	Low ROB	High ROB	High ROB
Priou, 2010[56]	Low ROB	Low ROB	Unclear	Low ROB	Low ROB	Moderate ROB
Salturk, 2015[57]	Low ROB	Low ROB	Low ROB	Low ROB	Low ROB	Low ROB
Sancho, 2014[58]	Low ROB	Low ROB	Low ROB	Low ROB	Low ROB	Low ROB
Sancho, 2017[59]	High ROB	Low ROB	Unclear	Unclear	Low ROB	High ROB
Sanjuan-López, 2014[60]	Low ROB	High ROB	Low ROB	Low ROB	Low ROB	High ROB
Schonhofer, 2001[61]	High ROB	Low ROB	Unclear	Unclear	Unclear	High ROB
Sivori, 2007[63]	Low ROB	Low ROB	High ROB	Unclear	Low ROB	Moderate ROB
Tsolaki, 2008[65]	High ROB	Low ROB	High ROB	Low ROB	Unclear	High ROB
Tsolaki, 2011[66]	Low ROB	Low ROB	Low ROB	Low ROB	Unclear	Low ROB
Vasquez, 2017[67]	Low ROB	Low ROB	Unclear	Unclear	Unclear	Moderate ROB
Vitacca, 2017[68]	Low ROB	Unclear	Unclear	Low ROB	Low ROB	Moderate ROB
Windisch,2006[69]	Low ROB	Low ROB	Low ROB	Unclear	Unclear	Moderate ROB

ROB: Risk of Bias

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[46] RCT	BPAP ST	-COPD (FEV1 < 50%) -NIPPV during hospital admission	-PaCO2 >53 mmHg -PaO2 <55 mmHg or PaO2 < 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[50] RCT	BPAP IVAPS versus BPAP ST	-COPD (FEV1 < 50%) -Mixed stable disease or following AECOPD	-PaCO2 >7 kPa (53 mmHg) -pH >7.35 or PtcCO2 >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 cmH2O while awake. The device then attempted to reproduce target minute ventilation overnight by automatically adjusting the inspiratory pressures in the range 7-25 cmH2O. (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 PtcCO2. EPAP set at 5cmH20. (Titration took on average 5.2 [SD 2.8] days)
Paone, 2014[51] Observational	BPAP ST	-COPD (FEV1 < 50%) -NIPPV during hospital admission	-PaCO2 > 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 cmH2O. 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 [29] Observational	BPAP NOS	-COPD (ICD-9) -NIPPV during hospital admission	-PaCO2 > 45 mmHg	
Bhatt, 2013[4] RCT	BPAP NOS	-COPD (FEV1 NOS) -Stable (no AECOPD in prior 4 weeks)	-PaCO2 <52 mmHg	IPAP set at 15 cmH2O. EPAP set at 5 cmH2O. Initiation performed in home by respiratory therapist over 1 week.
Duiverman[22, 23], 2011 RCT	BPAP ST	-COPD (FEV1<50%) -Stable (no AECOPD in prior 4 weeks)	-PaCO2 >6.0 kPa (45 mmHg) -pH >7.35 (daytime, room air)	Maximum tolerated IPAP to target PaCO2<6.0 kPa and PaO2 > 8.0 kPa.
Oscroft, 2010[48] Observational	BPAP ST started in AECOPD	-COPD (FEV1 <50%) -NIPPV during hospital admission for AECOPD	-PaCO2 >7.5 kPa (56 mmHg) -pH 7.35-7.45 (daytime) or -PaCO2 >6.5 kPa (49 mmHg) -pH 7.35-7.45 + PtcCO2 >9 kPa (68 mmHg) (daytime)	
	BPAP ST started in stable COPD	-COPD (FEV1 <50%) -Stable (no current AECOPD)	-PaCO2 >7.5 kPa (56 mmHg) -pH 7.35-7.45 (daytime) or -PaCO2 >6.5 kPa (49 mmHg) -pH 7.35-7.45 + PtcCO2 >9 kPa (68 mmHg) (daytime)	
Cheung, 2010[12] RCT	CPAP versus BPAP ST	-NIPPV during hospital admission for AECOPD	-PaCO2 > 6 kPa (45 mmHg) -pH <7.35	CPAP: CPAP set at 5 cmH2O BPAP ST: Maximum tolerated IPAP (range 10 to 20 cmH2O) to target tidal volume 7-10 mL/kg. EPAP set at 5 cmH2O. 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[10] RCT	BPAP S	-COPD (FEV1 <45%) -Stable (no AECOPD in prior 3 months)		Maximum tolerated IPAP (≥8 cmH2O above EPAP) to target 20% decrease in respiratory rate and visible decrease in accessory muscle use and dyspnea. EPAP set at 4 cmH2O. (Titrated in hospital for 1 week).
Garrod, 2000[30] 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[15] Observational	BPAP ST	-COPD (FEV1<50%) -Stable (no AECOPD in prior 4 weeks) -LTOT ≥12 months -≥1 ICU admission due to AECOPD in prior 2 years	-PaCO2 >6 kPa (45 mmHg) -pH >7.35 -PaO2 <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[14] Observational	BPAP ST	-COPD (FEV1 30-49%) -LTOT ≥18 months -≥1 hospital admission due to AECOPD in prior 18 months	-PaCO2 >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[70] RCT	BPAP ST	-COPD (FEV1<50%) -Stable (no AECOPD in prior 4 weeks)	-Hypercapnia (daytime, rest) NOS	Maximum tolerated IPAP (≥ 10 cmH2O). EPAP set at 4 cmH2O. Backup rate set at 16 breaths/min.
Marquez-Martin, 2014[38] RCT	BPAP ST	-COPD (FEV1<50%) -Stable (no AECOPD in prior 3 months)	-PaCO2 > 45 mmHg -PaO2 < 60 mmHg	Maximum tolerated IPAP (10-20 cmH2O) to target good clinical response and SaO2. EPAP set at 4 cmH2O. Backup rate set at 12 breaths/min.
Köhnlein, 2014[37] RCT	BPAP ST	-COPD (FEV1<30%) -Stable (no AECOPD in prior 4 weeks)	-PaCO2 ≥ 7 kPa (53 mmHg) -pH ≥ 7.35 (daytime, rest)	Targeted to reduce baseline PaCO2 by ≥ 20% or achieve PaCO2 <6.5 kPa (49 mmHg).
De Backer, 2011[19] RCT	BPAP NOS	-COPD (FEV1<50%) -AECOPD requiring hospitalization	-PaCO2 >45 mmHg on day 5-12 of hospitalization	Targeted SaO2 >90% during 90% of time and reduction in PaCO2 ≥ 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[21] RCT	HMV (pressure assist/control) versus HMV (PSV ST)	-COPD (Gold stage IV) -Stable (no current AECOPD).	-PaCO2 >45 mmHg (daytime) and PaCO2 >50 mmHg (nocturnal)	HMV (pressure assist/control): Maximum tolerated IPAP to target maximum reduction in PaCO2 (normocapnia if possible). EPAP set to avoid dynamic hyperinflation (3-6 cmH2O). 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[43] RCT	BPAP S	-COPD (FEV1<50% or <1.5L) -Stable disease -LTOT for ≥3 months	-PaCO2 >46 mmHg (at least twice in prior 6 months during stability)	Maximum tolerated IPAP-EPAP difference (≥5 cmH2O). EPAP set at 3 cmH2O 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[65] Observational	BPAP ST	-COPD (FEV1 <50%) -Stable (no AECOPD in prior 4 weeks)	-PaCO2>50 mmHg -PaO2<60 mmHg (room air)	IPAP and EPAP to target patient comfort, decreased accessory muscle use, lower respiratory rate, and decrease in PaCO2 >5% after 1 hour. (Titration in hospital).
Windisch, 2006[69] 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 PaCO2
Gay, 1996[31] RCT	BPAP ST versus sham CPAP lowest setting	-COPD (FEV1 < 40%) -Stable disease	-PaCO2 > 45 mmHg (daytime, rest)	IPAP set to 10 cmH2O. EPAP set to lowest possible. Backup rate to target patient comfort.
Gad, 2014[28] Observational	BPAP ST	-COPD (FEV1 < 50%) -Stable (no AECOPD in prior 4 weeks) PaCO2>50 mmHg	-PaCO2 >50 mmHg -pH > 7.35 (daytime)	Maximum tolerated IPAP (targeting 15-20 cmH2O). EPAP 3-6 cmH2O. (Titration occurred in hospital over 2-3 day period.)
Sin, 2007[62] RCT	BPAP NOS versus sham	-COPD (FEV1 NOS) -Stable disease		Maximum tolerated IPAP (maximum of 20 cmH2O). EPAP set at 4 cmH2O.

Author, Year, Study Design	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
	CPAP 4 cmH2O			
Heinemann, 2011[34] Observational	BPAP (pressure controlled ventilation)	-COPD (FEV1 NOS) -invasive mechanical ventilation for AECOPD, pneumonia, or postoperative respiratory failure -prolonged weaning from invasive mechanical ventilation	-PaCO2>52.5mmHg or -pH<7.35 (recurrent acidosis)	
Budweiser, 2007[8] Observational	BPAP (pressure controlled ventilation)	-COPD (FEV1 <50%) -Stable and unstable disease	-PaCO2>55mmHg -pH<7.35 (recurrent acidosis)	Maximum tolerated IPAP to achieve maximum reduction in PaCO2.
Clini, 2002[16] RCT	BPAP ST	-COPD (FEV1 NOS) -Stable (no AECOPD in prior 4 weeks)	-PaCO2 >6.6 kPa (50 mmHg) -pH>7.35 (daytime, room air)	Maximum tolerated IPAP with goal decrease in PaCO2 >5% after 1 hour; and nocturnal SaO2≥90% for 90% of time. (Titration in hospital).
Struik, 2014[64] RCT	BPAP ST	-COPD (FEV1 <50%) -NIPPV or invasive mechanical ventilation in hospital admission	-PaCO2 >6 kPa (45 mmHg)	Maximum tolerated IPAP to achieve normal PaCO2. 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.
Durao, 2018[25] Observational	HMV/BPAP mix	-COPD (NOS) -AECOPD		Maximum tolerated IPAP to achieve maximum reduction in PaCO2. Backup raespiratory 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 cmH2O)
	HMV/BPAP mix	-COPD (NOS) -Stable (no current AECOPD)		
Duiverman, 2017[24] 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	-PaCO2 ≥6.7 kPa (50 mmHg) (daytime) or -PaCO2 ≥7.3 kPa (55 mmHg) (nighttime) or -Nighttime rise in PtCO2 ≥1.3 kPa (10 mmHg)	Pressure controlled ventilation: Maximum tolerated IPAP to achieve maximum reduction in PaCO2. Backup rate set just above spontaneous breathing frequency. EPAP set at 4-6cm H2O. Pressure support ventilation: Maximum tolerated IPAP, with maximum IPAP of 18 cmH2O 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
				breaths/minute.
Blankenburg, 2017[5] Observational	HMV (pressure controlled ventilation or pressure support ventilation)	-COPD (FEV1 NOS) -Stable (no AECOPD in prior 2 weeks)	-PaCO2>7.0kPa (53mmHg) -pH>7.35	Goal of titration was normal PaCO2 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[66], Observational	BPAP ST	-COPD (FEV1 NOS) -Stable (no AECOPD in prior 4 weeks) -Symptoms of nocturnal hypoventilation (fatigue, dyspnea, morning headaches)	-PaCO2 ≥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 2 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[57] 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[35] Observational	HMV (pressure cycled assist control mode)	-Stable (no current AECOPD) -HMV initiated ≥3 months		
Funk, 2010[27] 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[67] cohort	BPAP NOS versus CPAP NOS versus HMV NOS	-COPD (ICD-9)		
Oscroft, 2010[49] 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[57] Observational	BPAP ST	Kyphoscoliosis NOS		IPAP titrated to achieve "desired tidal volume" (maximum 30 mbar)
Domenéech-Clar, 2003[20] 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[47] 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[39] 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[61] 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[66] Observational	BPAP ST	-TRD NOS -Stable (no acute exacerbation in prior 4 weeks) -Symptoms of nocturnal hypoventilation (fatigue, dyspnea, morning headaches)	-PaCO2 ≥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 2 values was considered as adequate ventilatory support.

BPAP: Bilevel Positive Airway Pressure, cmH2O: centimeters of water (pressure), EPAP: expiratory positive airway pressure, FEV1: 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, PaCO2: 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[35] Observational	HMV (pressure cycled assist control mode) in restrictive thoracic disease	-TRD NOS -HMV initiated ≥3 months		
Buyse, 2003[9] 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[60] 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[53] Observational	BPAP NOS	-ALS (El Escorial criteria) -bulbar features		
Doménech-Clar, 2003[20] Observational	BPAP NOS	-NMD 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 -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[47] Observational	BPAP NOS	-NMD 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)
Sancho, 2014[58] Observational	HMV (volume cycled) versus BPAP ST	-ALS NOS -symptoms (fatigue, dyspnea, orthopnea, morning headache)	-PaCO2 >45 mmHg or -FVC <50% or -MIP <60 cm H2O or -SaO2 < 88% for ≥ 5 consecutive minutes by nocturnal oximetry	Titration occurred in the hospital.

Author, Year, Study Design	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
Sivori, 2007[63] Observational	BPAP NOS	-ALS (El Escorial criteria) -symptomatic ventilatory impairment (dyspnea, morning headache, fatigue)	-PaCO2 >45 mmHg or -FVC <50% or -MIP <60 cm H2O or -nocturnal SaO2 < 88% for ≥ 5 consecutive minutes	IPAP adjusted to maintain SpO2 >92% (ranged 13-25 cmH2O). EPAP set from 5-9 cmH2O.
Coco, 2006[17] Observational	BPAP ST	-ALS (El Escorial criteria) -symptomatic ventilatory impairment (dyspnea, morning headache, fatigue)	-PaCO2 >45 mmHg or -FVC <50% or -MIP <60 cm H2O or -nocturnal SaO2 < 88% for ≥ 5 consecutive minutes	Maximum tolerated IPAP and EPAP to target patient comfort, leaks, normal PaO2, PaCO2, SpO2, and symptom relief. IPAP started at 8-12 cmH2Oand EPAP started at 3-4c cmH2).
Bourke, 2006[7] 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[68] Observational	HMV/BPAP mix started in FVC≥80% (early)	-ALS NOS -FVC≥80%		Pressures adjusted to patient comfort, normalization of PaCO2, 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[59] Observational	HMV (volume assist control ventilation)	-ALS (El Escorial criteria) -symptomatic ventilatory impairment (dyspnea, orthopnea, fatigue, morning headache, daytime hypersomnolence, decreased cognitive function)	-PaCO2 >45 mmHg and -FVC <50% and -nocturnal SaO2 < 90% for ≥ 5% of time	Ventilator adjusted to target PaCO2<45mmHg, nocturnal SaO2 < 90% for <5% of time, optimize comfort, prevent air leaks.
Bertella, 2017[3] RCT	BPAP volume assured pressure support ventilation	-ALS (definite via El Esocrial Criteria) -Stable disease (no respiratory infection in prior 3 months	-PaCo2>45mmHg, 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 PaCO2. Device titrated in patient versus outpatient according to randomization.
Tsolaki, 2011[66]	BPAP ST	-NMD NOS	-PaCO2 ≥45mmHg	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		-Stable (no acute exacerbation in prior 4 weeks) -Symptoms of nocturnal hypoventilation (fatigue, dyspnea, morning headaches)	-pH>7.35	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 2 values was considered as adequate ventilatory support.
Aboussouan, 1997[l] Observational	HMV/BPAP mix	-ALS via el Escorial criteria -dyspnea on exertion or or orthopnea or FVC < 60% predicted.	- PaCO2 ≥ 45 mmHg	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

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[26] Observational	HMV/BPAP mix	-ALS NOS -Symptoms (orthopnea) -FVC ≤50% predicted or a decrease in FVC of ≥ 500 mL on two consecutive visits	-Desaturations in nocturnal pulse oximetry (arterial oxygen saturation, <90% during 5 consecutive min) Or PaCO2 >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, cmH2O: 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, PaO2: partial pressure of arterial oxygen, PaCO2: partial pressure of arterial carbon dioxide, Pimax: maximal inspiratory mouth pressures, PSV: Pressure support ventilation, RCT: randomized controlled trial, SaO2: arterial blood oxygen saturation, SpO2: 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[54] Observational	BPAP ST + weekly telemonitoring versus BPAP ST without weekly telemonitoring	-ALS NOS -home BPAP use -FVC ≥75%	-PaCO2 ≤ 45 mmHg -PaO2 ≥80 mmHg	Increase in IPAP to achieve normal breathing patterns, daytime and nocturnal SaO2 > 95%. Backup rate set slightly lower than the patient's own respiratory frequency. (Titration occurred in hospital or outpatient clinic)
Gonzalez-Bermejo, 2013[32] 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 PaO2, PaCO2, and SpO2. EPAP ranged from 3-5 cmH2O.

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

Author, Year, Study Design	Device/mode	Patient characteristics to start or continue device	Laboratory characteristics to start or continue device	Device titration
Howard, 2016[36] 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[57] 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[39] 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[11] 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[40, 41] RCT	HMV/BPAP mix (all with bilevel pressure with assured volume) versus CPAP (fixed pressure)	-OHS (BMI ≥ 30; stable PaCO2 ≥ 45 mmHg; pH ≥ 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 ≥30) -Correctly executed 30min CPAP/NIPPV treatment trial test	-PaCO2 ≥ 45 mmHg -pH ≥ 7.35	HMV/BPAP mix: IPAP maximum tolerated to target reduction in PaCO2, normal SaO2, 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[6] RCT	BPAP ST	-OHS (BMI >30; daytime PaCO2 ≥ 45 mmHg, other causes of hypoventilation ruled out including airway obstruction, scoliosis, cardiac failure, progressive NMD)	-PaCO2 ≥ 45 mmHg (daytime)	(Titration occurred in hospital over 3-4 nights)
Murphy, 2012[45] RCT	BPAP (volume assured pressure support ventilation) versus BPAP ST	-OHS (BMI>40, daytime chronic PaCO2 >6 kPa, pH >7.35), absence of other identifiable hypoventilation cause, FEV1/FVC >70%, FVC <70% -Stable disease	-PaCO2 >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[55] RCT	BPAP S versus CPAP	-OHS (BMI≥30, PaCO2 ≥ 45 mmHg [awake, stable], absence of another cause for hypercapnia, FEV1/FVC ≥ 70%)	-PaCO2 ≥45 mmHg -pH ≥7.34 (daytime, stable) Excluded during CPAP titration study: -SaO2 <80% for 10 minutes in absence of apnea -TcCO2 during REM ≥10mmHg -increase in afternoon to morning PaCO2 ≥10mmHg in patients with awake PaCO2 >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[5]	HMV (pressure controlled ventilation or pressure support ventilation)	-OHS (BMI>30, PaCO2 >6.7kPa[50mmHg], symptoms of hypercapnia NOS), absence of another cause for hypercapnia	-PaCO2>7.0kPa (53mmHg) -pH>7.35	Goal of titration was normal PaCO2 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[66] 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	-PaCO2 >45mmHg -PaO2 <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 2 values was considered as adequate ventilatory support.
Masa, 2016[42] RCT	BPAP volume assured pressure support ventilation	-OHS (BMI ≥ 30 kg/m2, no COPD, no NMD, no TRD, no narcolepsy, no restless leg syndrome) -Stable disease	-PaCO2 ≥ 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 H2O and the inspiratory positive airway pressure (IPAP) was set between 18 and 22 cm H2O (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
				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 PaCO2 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
Perez de Llano, 2005[52], Observational	HMV/BPAP mix	-OHS, BMI > 30, , FEV1/FVc < 70%, absence of other respiratory disorder such as kyphoscoliosis or diaphragmatic paralysis	-PaCO2 ≥ 50 mmHg	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_2O, and the positive inspiratory pressure (PIP) was set at 10 cm H_2O. PIP was gradually adjusted upward as tolerated. Oxygen was administered, when needed, through the mask until the arterial oxygen saturation (Sao_2) 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 (*ie*, pH > 7.35), daytime NIPPV therapy was stopped. We employed daytime arterial blood gas measurements and overnight pulse oximetry to determine

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_2 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_2 during overnight oximetry was ≥ 88% and diurnal $Paco_2$ 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,[56] Observational	BPAP	-OHS (PaCO2> 45 mm Hg in the absence of any other cause of hypoventilation on the basis of clinical xamination, chest radiograph, and pulmonary function tests (eg, COPD [FEV 1 to vital capacity ratio , 70%]).	PaCO2> 45 mm Hg	Expiratory positive airway pressure (EPAP) and inspiratory positive airway pressure (IPAP) were adapted by 2 cm H_2O steps using repeated oximetry and arterial blood gases (ABG) to alleviate OSA-related desaturations, to improve mean nocturnal oxygen desaturation index ($Sa\ o_2$), and to achieve a maximal reduction in daytime $Pa\ co_2$. Supplemental oxygen was added to NPPV in patients with persistent nocturnal hypoxia (as defi ned arbitrarily by ≥ 20% of time with $Sa\ o_2$, < 90%) despite a delta between EPAP and IPAP of at least 10 cm H_2O 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, FEV1: 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, PaCO2: partial pressure of arterial carbon dioxide, S: spontaneous mode, SaO2: arterial blood oxygen saturation, ST: spontaneous/timed breath mode, TcCO2: 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[57] 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[2] Observational	Diffuse bronchiectasis	HMV (volume cycled)	-Diffuse bronchiectasis -Home HMV -LTOT		Target PaO2 > 9kPa (67 mmHg) without deterioration in PaCO2.

HMV: Home Mechanical Ventilation, kPa: kilopascal, LTOT: long term oxygen therapy, mmHg: millimeters of mercury (pressure), PaO2: partial pressure of arterial oxygen, PaCO2: 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[69] 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 PaCO2
Hazenberg, 2014[33]	NMD, TRD	HMV (pressure or volume	-NMD or thoracic cage disorder -Stable disease without acute respiratory failure	-PaCO2 >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 PaCO2 and PaO2. (Titration occurred at home)
		HMV (pressure or volume control) started in the hospital	-NMD or thoracic cage disorder -Stable disease without acute respiratory failure	-PaCO2 >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 PaCO2 and PaO2. (Titration occurred at the hospital)
Munoz, 2005[44] 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	-PaCO2 > 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[13] RCT	COPD, Other	BPAP NOS	-COPD or asthma or bronchiectasis -hospital readmission due to respiratory cause -Daytime sleepiness or morning headache	-PaCO2 > 50 mmHg (daytime rest) -SpO2 < 88% for more than 5 consecutive minutes while on usual oxygen during polysomnography	IPAP and EPAP and volumes set to target optimal daytime PaCO2

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, PaO2: partial pressure of arterial oxygen, PaCO2: partial pressure of arterial carbon dioxide, RCT: randomized controlled trail, SpO2: 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[18] 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[67] Observational	COPD	Inclusion: COPD (ICD-9); age ≥40 years	1) BPAP NOS 2) CPAP NOS 3) HMV 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).
Murphy, 2017[46], 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
		years); AECOPD requiring hospital admission and acute NIPPV; PaCO2 >53 mmHg; PaO2 <55 mmHg or PaO2 < 60 mmHg with polycythemia, pulmonary hypertension or cor pulmonale; >30% sleep time with SaO2 <90%; pH >7.30 room air Exclusion: intubated during AECOPD, current home NIPPV use, cognitive impairment, unstable psychiatric morbidity, undergoing renal replacement therapy, unstable coronary artery syndrome, age < 18 years, homeless, BMI >35, OSA.	2) Home oxygen		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.
Oscroft, 2014[50], RCT	COPD	Inclusion: COPD (FEV1 <50%, FEV1/FVC ratio <70%, TLC>80%, smoking history > 20 pack years); daytime PaCO2 >7 kPa and pH >7.35	1) BPAP volume assured pressure support ventilation	3 months	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).

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
		or PtcCO2 >9 kPa Exclusion: Age>80 years, other respiratory disease, BMI>40, significant OSA.	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[51], Observational	COPD	Inclusion: COPD (FEV1 <50%, FEV1/FVC ratio <70%, <20% improvement bronchodilator response); NIPPV during hospital stay; PaCO2 > 50 mmHg immediately after awakening from a night without NIPPV 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>40, systemic corticosteroids.	1) BPAP ST + Home oxygen 2) Home oxygen	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[29], Observational	COPD	Inclusion: AECOPD (ICD-9); PaCO2 > 45 mmHg; NIPPV during hospital stay Exclusion: discharged to hospice.	1) BPAP NOS 2) No BPAP	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).
Bhatt, 2013[4], 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 2) No BPAP	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.

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
Duiverman, 2011[22, 23], RCT	COPD	Inclusion: COPD (FEV1 <50%, FEV1/FVC < 70%, GOLD stage III/IV); age 40-76 years; no exacerbation in past 4 weeks; daytime PaCO2 >6.0 kPa Exclusion: cardiac/neuromuscular disease limiting exercise tolerance, pulmonary rehabilitation in past 18 months, prior NIPPV, apnea-hypopnea index ≥10h.	1) BPAP ST + Pulmonary rehabilitation 2) Pulmonary rehabilitation alone	Longest duration: 24 months	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.
Oscroft, 2010[48], Observational	COPD	Inclusion: COPD (FEV1 <50%, FEV1/FVC ratio <70%, smoking history >20 pack years); AECOPD requiring hospital admission; daytime PaCO2 >7.5 kPa with pH 7.35-7.45 or daytime PaCO2 >6.5 kPa with pH	1) BPAP ST started in AECOPD	28.6 months, 95% CI 10.9-46.8 months, Median 52.4 months	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).

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
		7.35-7.45 with PtcCO2 >9 kPa Exclusion: Age>80 years, other respiratory disease, BMI>35, significant OSA, tracheostomy, impaired left ventricular function.	2) BPAP ST started in stable COPD		
Cheung, 2010[12] RCT	COPD	Inclusion: AECOPD requiring hospital admission and NIPPV, pH <7.35, PaCO2 > 6 kPa Exclusion: Active smokers, RF from other cause, pneumonia, transmissible infections, long-term corticosteroid use, comorbidity giving life expectancy <1 year, significant OSA, already on home NIPPV	1) CPAP 2) BPAP ST	Longest duration: 12 months	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).

Author, Year, Study Design	Disease	Inclusion/Exclusion Criteria	Device And Settings Used (Group)	Duration Of Device Use In Home Setting	Conclusion
McEvoy, 2009[43] RCT	COPD	Inclusion: COPD (FEV1<50% or <1.5L, bronchodilator response <20%, FEV1/FVC ratio <60%); PaCO2 <46 mmHg at least twice in the prior 6 months during clinical stability; LTOT for ≥3 month; Age<80 years 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>40, evidence of sleep apnea.	1) BPAP S + Oxygen 2) Oxygen alone	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.
Casanova[10], 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[30] 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[15] Observational	COPD	Inclusion: COPD, prior smokers, LTOT ≥12 month; stable disease (no AECOPD in prior 4 weeks); stable PaCO2; pH>7.35; PaO2 < 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
		PaCO2 >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[14], Observational	COPD	Inclusion: COPD, LTOT ≥18 mo.; chronic PaCO2 >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 2) 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).
Zhou, 2017[70] 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[38] 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[37] RCT	COPD	Inclusion: COPD (GOLD IV); clinically stable (no AECOPD in prior 4 weeks); $PaCO2 \geq 7$	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
		kPa (51.9 mmHg); pH ≥ 7.35 (rest) Exclusion: Thorax/lung abnormalities other than COPD, BMI≥35, other conditions resulting in hypercapnia, previously initiated NPPV, malignant comorbidities, severe HF, unstable angina, severe arrhythmias.	2) Standard care		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.
De Backer, 2011[19] RCT	COPD	Inclusion: COPD (FEV1<50%, FEV1/FVC <70%), AECOPD requiring hospitalization, PaCO2 >45 mmHg, stopped smoking	1) BPAP NOS	At least 6 months	The 6-minute walk distance increased significantly in the BPAP group (232 ± 151 m to 282 ± 146 m, p = 0.01), while there was

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[27] RCT	COPD	Inclusion: COPD; AECOPD requiring NIPPV or invasive ventilation; chronic nocturnal NIPPV use at home for ≥ 6 months; clinically stable, PaCO2 > 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[21] RCT	COPD	Inclusion: COPD (Gold stage IV); daytime PaCO2 > 45 mmHg; nocturnal PaCO2 > 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) 2) HMV (pressure support ventilation)(time period 1) 2) Pulmonary rehabilitation alone	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.
Tsolaki, 2008[65] Observational	COPD	Inclusion: COPD (FEV1 <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
		years; Age≤75 years; PaO2 < 60 mmHg (room air); PaCO2 >50 mmHg (room air) 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.	2) Standard care		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). There was no significant difference on number of exacerbations, hospitalization due to exacerbations, endotracheal intubation, or mortality.
Chiang, 2003[13] 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
		headache; $PaCO_2$ > 50 mmHg (daytime rest); SpO_2 < 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.	2) Standard care		distance test group (101.2 meters vs. -33.8 meters, $p<0.05$), 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.
Gay, 1996[31] RCT	COPD	Inclusion: COPD (FEV1 < 40%); $PaCO_2$ >45 mmHg (daytime, rest); Age<80 years, BMI≤30	1) BPAP ST	3 months	No difference was found on 6-minute walk distance test, total sleep time, sleep efficiency,

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[28] 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[62] 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[34] Observational	COPD	Inclusion: COPD; prolonged weaning from invasive mechanical ventilation Exclusion: intubated due to cardiogenic edema or cardiopulmonary resuscitation	1) HMV pressure controlled ventilation 2) No device	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).
Budweiser, 2007[8] 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[16] 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≤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[64] 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 2 >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).
Durao, 2018[25] 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[24] RCT	COPD	Inclusion: COPD (GOLD III or IV), ≥ 2 AECOPD with acute hypercapnic respiratory failure (pH<7.35) per year, daytime Inclusion: PaCO2 ≥6.7 kPa (50 mmHg) or nocturnal PaCO2 ≥7.3 kPa (55 mmHg) or nighttime rise in PtCO2 ≥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[49]	COPD	COPD, FEV1<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[9] Observational	TRD	Inclusion: Kyphoscoliosis with respiratory insufficiency who started LTOT and/or NIPPV.	1) HMV (volume cycled or pressure cycled) + oxygen 2) Oxygen alone	10 months	Survival rate was significantly higher in patients treated with HMV plus long-term oxygen than patients with long-term oxygen alone ($p<0.05$)
Schonhofer, 2001[61] Observational	TRD	Inclusion: TRD (post-TB or scoliosis); $PaCO_2$ 45-55 mmHg; stable $PaCO_2$ 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 2) Standard care without HMV/BPAP device	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.

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, $PaCO_2$: 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[60] Observational	NMD	Inclusion: ALS; hospital admission; chronic RF by pulmonologist Exclusion: Neuromuscular processes other than ALS, treatment in social welfare palliative center.	1) HMV (pressure support ventilation mode or BPAP ST mode) started after outpatient pulmonary evaluation	23.3 months (95% CI, 16.7–28.8)	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<0.004).
			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[54] Observational	NMD	Inclusion: ALS; home BPAP use; FVC ≥75%; PaO2 ≥80 mmHg; PaCO2 ≤ 45 mmHg; age 18-75 years Exclusion: Gastrostomy, cognitive impairment, other significant disorders.	1) BPAP ST + Weekly telemonitoring + Standard care	36 months	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)
			2) BPAP ST + Standard care		
Gonzalez-Bermejo, 2013[32] Observational	NMD	Inclusion: ALS on home BPAP with 4 hour/night minimal adherence Exclusion: Use of other ventilator types, without integrated SpO2	1) BPAP ST "correctly ventilated patients"	12 months	The “correctly ventilated” patients had significantly lower mortality than those “insufficiently ventilated” patients (OR= 0.25; 95% CI: 0.10 to 0.64).
			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[58] Observational	NMD	Inclusion: ALS; symptoms (fatigue, dyspnea, orthopnea, morning headache) plus one of the following 1) PaCO2 >45 mmHg or 2) FVC <50% or 3) MIP <60 cm H2O or 4) SaO2 < 88% for ≥ 5 consecutive minutes by nocturnal oximetry Exclusion: Presence of previous pulmonary/airway disease, rapidly progressing disease w/ survival expectancy <1 month, severe frontotemporal dementia, NIPPV tolerance <4 consecutive hour/night.	1) HMV (volume assist control ventilation) 2) BPAP ST	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[63] Observational	NMD	Inclusion: ALS; symptomatic ventilatory impairment (dyspnea, morning headache, fatigue) plus 1) PaCo2 > 45 mmHg or 2) nocturnal oxygen saturation by pulse	1)BPAP, Riluzole 2) BPAP NOS 3) No BPAP, No Riluzole	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 ≤ 88% for 5 continuous minutes or 3) MIP < 60 cmH2O or 4) FVC < 50%.			
Coco, 2006[17] Observational	NMD	Inclusion: ALS; symptomatic ventilatory impairment (dyspnea, morning headache, hypersomnolence, fatigue) plus 1) PaCO2 ≥ 45 mmHg or 2) nocturnal oxygen saturation by pulse oximeter ≤ 88% for 5 continuous minutes or 3) MIP < 60 cmH2O or 4) FVC < 50% Exclusion: Primary lateral sclerosis, diagnosis other than ALS during followup.	1) BPAP ST (use ≥ 4 hours/day) 2) BPAP ST < 4 hours/day)	Longest Duration: 30 months	The group with ≥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
Bourke, 2006[7] 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[53] Observational	NMD	Inclusion: ALS; bulbar features Exclusion: Tracheotomised, refusal of attempts to prolong survival.	1) BPAP NOS 2) No BPAP NOS	Longest Duration: 42 months	With a 3-year followup, patients treated with BPAP were founded to have significantly higher overall survival than patients with palliative management (p=0.004).
Vitacca, 2017[68] 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) 2) HMV/BPAP mix started in FVC <80% (late)	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).
Sancho, 2018[59] 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) 2) No device	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:

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[3] RCT	NMD	Inclusion: ALS (definite via El Esocrial 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[1] Observational	NMD	Inclusion: ALS via el Escorial criteria; dyspnea on exertion or PaCO2 ≥ 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[26] 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, cmH2O: centimeters of water (pressure), CPAP: Continuous Positive Airway Pressure, BMI: Body Mass Index, FEV1: 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, PaO2: partial pressure of arterial oxygen, PaCO2: partial pressure of arterial carbon dioxide, Pimax: maximal inspiratory mouth pressures, PSV: Pressure support ventilation, RCT: randomized controlled trial, RF: Respiratory Failure, SaO2: arterial blood oxygen saturation, SF-36: Medical Outcomes Study Questionnaire Short Form, SpO2: 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[36] RCT	OHS	Inclusion: OHS (BMI >30, daytime PaCO2 >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[40, 41]RCT	OHS	Inclusion: OHS (BMI ≥ 30; stable PaCO2 ≥ 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 (FEV1/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[6], RCT	OHS	Inclusion: OHS (BMI >30; daytime PaCO2 ≥ 45 mmHg); age 20-75 years Exclusion: Declined or presented any significant airway obstruction, scoliosis, cardiac failure, progressive NMD.	1) BPAP ST 2) Lifestyle counseling	Longest Duration: 1 month	No significant difference were found on sleep quality measured by Epworth Sleepiness Scale (p=0.49).
Murphy, 2012[45], RCT	OHS	Inclusion: OHS (BMI>40, daytime chronic PaCO2 >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 2) BPAP ST	Longest Duration: 3 months 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).
Piper, 2008[55], RCT	OHS	Inclusion: OHS (BMI≥30, PaCO2 ≥ 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 ≥ 70%) Exclusion: psychiatric illness, current home NIPPV use, PtcCO2 during REM ≥10mmHg, increase in afternoon to morning PaCO2 ≥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, [42] RCT	OHS	Inclusion: OHS (BMI ≥ 30 kg/m2, no COPD, no NMD, no TRD, no narcooepsy, no restless leg syndrome), stable hypercapnic respiratory failure (daytime awake PaCO2 ≥ 45 mmHg, pH ≥ 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 signficaint 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[52] Observational	OHS	Inclusion: OHS, BMI > 30, PaCO2 ≥ 50 mmHg, FEV1/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[56] Observational	OHS	Inclusions: BMI ≥ 30 kg/m 2 and daytime hypercapnia (PaCO2> 45 mm Hg) in the absence of any other cause of hypoventilation on the basis of clinical xamination, chest radiograph, and pulmonary function tests (eg, COPD [FEV 1 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, FEV1: 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, PaCO2: 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, PtcCO2: 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[2], 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[33], RCT	NMD, TRD	Inclusion: NMD or thoracic cage disorder; PaCO2 >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[44], Observational	NMD, TRD	Inclusion: Hospital admission with CHRF secondary to NMD (ALS excluded) or kyphoscoliosis or post TB sequelae; PaCO2 > 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[13], RCT	COPD, other	Inclusion: COPD or asthma or bronchiectasis; hospital readmission due to respiratory cause; daytime sleepiness or morning headache; PaCO2 > 50 mmHg (daytime rest); SpO2 < 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 2) No 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.

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, PaCO2: partial pressure of arterial carbon dioxide, RCT: randomized controlled trial, RF: Respiratory Failure, SF-36: Medical Outcomes Study Questionnaire Short Form, SpO2: 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[67] Observational	BPAP NOS	NR	IPAP, EPAP	NR	NR
	CPAP NOS	NR	CPAP	NR	NR
	HMV NOS	NR	NR	NR	NR
Murphy, 2017[46] 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 H20 -EPAP: 4 (4-5) cmH20 -Rate: 14 (14-16) breaths/minute
Salturk, 2015[57] Observational	BPAP ST	NR	IPAP, EPAP, rate	NR	-6.7 ± 1.9 hours/day -IPAP: 24 ± 3 cm H20 -EPAP: 5.3 ± 0.7 cmH20
Oscroft, 2014[50] 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) cmH20 -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) cmH20 -EPAP: 5 (5-5) cmH20 -Rate: 15.0 (15-15) breaths/minute
Paone, 2014[51] 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[29] Observational	BPAP NOS	NR	IPAP, EPAP	NR	-IPAP: 22.1 ± 6.2 cm H2O -EPAP: 5.9 ± 1.8 cm H2O
Bhatt, 2013[4]	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 H2O
Duiverman, 2011[22, 23] RCT	BPAP ST	BiPAP Synchrony (Respironics Inc.; Murrysville, USA)	IPAP, EPAP, rate	NR	Followup #1: -IPAP: 23 ± 4 cm H2O -EPAP: 6 ± 2 cm H2O -Rate: 18(3) breaths/minute Followup #2: -7.7 (5.8-8.5) hours/day -IPAP: 20 ± 4 cm H2O -EPAP: 6 ± 2 cm H2O -Rate: 18 ± 3 breaths/minute
Oscroft, 2010[48] Observational	BPAP ST	NIPPY I, 2 or 3 (B & D Electromedical; Stratford, United Kingdom)	IPAP, EPAP, rate	NR	-IPAP: 26 ± 3 cm H2O -EPAP: 4 ± 1 cm H2O -Short inspiratory (0.8- 1 s) -Long expiratory time (2.5-3.5 s).
Cheung, 2010[12] 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 H2O -EPAP: 5 ± 0 cm H2O
Hitzl, 2009[35] Observational	HMV (pressure controlled ventilation)	NR	inspiratory pressure, PEEP, inspiratory time, rate	NR	-IPAP: 20.9 ± 4.0 cm H2O -EPAP: 4.2 ± 1.9 cm H2O -Rate: 19.1 ± 3.8 breaths/minute
McEvoy, 2009[43] 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 H2O -EPAP: 5.1 (4.8-5.3) cm H2O
Windisch, 2006[69] 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[10] 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[30] 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[15] 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[14] 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[38] 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[37] 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[19]	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[27] RCT	BPAP NOS	NR	IPAP, EPAP	NR	NR
Dreher, 2010[21] 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[65] 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[31] RCT	BPAP ST	BiPAP (Respironics Inc. ; Murrysville, USA)	IPAP, EPAP, rate	NR	-5.1 ± 3.8 hours/day
Gad, 2014[28] 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[62] RCT	BPAP NOS	VPAP II (ResMed; Sydney, Australia)	IPAP, EPAP	NR	NR
Heinemann, 2011[34] 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[8] 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[16] RCT	BPAP ST	BiPAP ST 30 (Respironics Inc.; Murrysville, USA)	IPAP, EPAP, rate	NR	-9 ± 2 hours/day -IPAP: 14 ± 3 cm H2O -EPAP: 2 ± 1 cm H2O
Struik, 2014[64] 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 H2O -EPAP: 4.8 ± 1.0 cm H2O -Rate: 15 ± 3 breaths/minute -Inspiratory time 1.1 ± 0.3 s
Durao, 2018[25] 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 H2O -Rate: 15.2 ± 1.4 breaths/minute
Duiverman, 2017[24] 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 H2O -EPAP: 5.4 ± 0.9 cm H2O -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 H2O -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[5] 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[49] 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[66] 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[57] Observational	BPAP ST	NR	IPAP, EPAP, rate	NR	-5.9 ± 1.8 hours/day -IPAP: 22 ± 5 cm H20 -EPAP: 5.3 ± 0.6 cmH20

Author, Year, Study Design	Device/mode	Model; Manufacturer (Location of manufacturer)	Device Characteristics	Prescribed Usage (frequency and duration)	Actual Usage
Hitzl, 2009[35] Observational	HMV (pressure controlled ventilation)	NR	inspiratory pressure, PEEP, inspiratory time, rate	NR	-IPAP: 20.9 ± 4.0 cm H2O -EPAP: 4.2 ± 1.9 cm H2O -Rate: 19.1 ± 3.8 breaths/minute
Doménech-Clar, 2003[20] Observational	BPAP NOS	DP-90 (Taema; Paris, France)	IPAP, EPAP	≥7 hours/night	-mean 6 hours/night
Buyse, 2003[9] 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[47] Observational	BPAP NOS	DP-90 (Taema; Paris, France)	IPAP, EPAP	NR	-mean 7 hours/night
Schonhofer, 2001[61] 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[39] 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[66] Observational	BPAP ST	VPAP III ST (ResMed; Sydney, Australia)	IPAP, EPAP, rate	≥ 4 hours daily	-IPAP: 14.7 ± 2.4 cm H2O -EPAP: 5.0 ± 1.1 cm H2O

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, 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[60] 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[54] 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[20] Observational	BPAP NOS	DP-90 (Taema; Paris, France)	IPAP, EPAP	≥7 hours/night	-Mean 6 hours/night
Nauffal, 2002[47] 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[32] 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[58] 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[63] Observational	BPAP NOS	NR	IPAP, EPAP	NR	NR
Coco, 2006[17] Observational	BPAP ST	BiPAP (Respironics Inc.; Vitalaire, Italy)	IPAP, EPAP, rate	Use ≥ 4 or < 4 hours/day	NR
Bourke, 2006[7] 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[53] Observational	BPAP NOS	NR	IPAP, EPAP	NR	NR
Vitacca, 2017[68] 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 H2O -Tidal volume: 7.06 ± 1.47 ml/kg -Rate: 12.67 ± 1.46 breaths/minute
Sancho, 2017[59] 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[3] 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, [1]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,[26] Observatrional	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[66] Observational	BPAP ST	VPAP III ST (ResMed; Sydney, Australia)	IPAP, EPAP, rate	≥ 4 hours daily	-IPAP: 14.1 ± 2.1 cm H2O -EPAP: 5.2 ± 0.4 cm H2O

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, H2O: 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[36] RCT	BPAP ST	NR	IPAP, EPAP, rate	NR	-5.3 (2.63) hours/night -IPAP: 19.3 ± 2.8 cm H20 -EPAP: 11.9 ± 2.3 cmH20 -Rate: 15.0 ± 2.7 breaths/minute
	CPAP	NR	CPAP	NR	-5.0(2.4) hours/night -CPAP: 15.2 ± 2.8 cm H20
Salturk, 2015[57] Observational	BPAP ST	NR	IPAP, EPAP, rate	NR	-6.4 ± 2.4 hours/day -IPAP: 23 ± 3 cm H20 -EPAP: 5.8 ± 0.8 cmH20
Masa, 2000[39] 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[11] Observational	BPAP ST	Harmony BiPAP (Respironics; Louisville, USA)	IPAP, EPAP, rate	NR	-5.7 ± 1.3 hours/night
Masa, 2015[40, 41] 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) Puritian 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[6] 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[45] 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[55] RCT	BPAP S	NR	IPAP, EPAP	NR	-IPAP: 16 ± 2 cm H2O -EPAP: 10 ± 2 cm H2O
	CPAP	NR	CPAP	NR	NR
Blankenburg, 2017[5] 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[42] RCT	BPAP volume assured pressure support ventilation	NR	IPAP, EPAP, rate, target minute ventilation	NR	NR
Perez de Llano 2005[52] 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[56], Observational	BPAP	NR	NR	NR	NR
Tsolaki, 2011[66] 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[57] Observational	BPAP ST	NR	IPAP, EPAP, rate	NR	-5.8 ± 1.4 hours/day -IPAP: 21 ± 5 cm H20 -EPAP: 5.5 ± 0.7 cmH20
Benhamou, 1997[2] 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[33] 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[18] Observational	COPD, TRD, NMD, OHS, Other	HMV (pressure or volume controlled NOS)	NR	NR	NR	Age ≥ 75 years old -IPAP: 14-20 cm H20

Author, Year, Study Design	Diseases	Device/mode	Model and Manufacturer	Device Characteristics	Prescribed Usage (frequency and duration)	Actual Usage
						-EPAP: 3-8 cmH20 -Tidal volume: 500-800 ml -Rate: 16- 22 breaths/minute <u>Age 65-74 years old</u> -IPAP: 14-24 cm H20 -EPAP: 14-20 cmH20 -Tidal volume: 400-800 ml -Rate: 14- 22 breaths/minute <u>Age <65 years old</u> -IPAP: 14-30 cm H20 -EPAP: 3-8 cmH20 -Tidal volume: 400-1000 ml -Rate: 12- 24 breaths/minute
Munoz, 2005[44] 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[13] 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[69] Observational	HMV (pressure controlled ventilation)	PV401(Breas Medical AB; MoeInlycke, 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, H2O: 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[46] RCT	BPAP ST	-Smoking cessation NOS -COPD education NOS
Oscroft, 2014[50] RCT	BPAP iVAPS versus BPAP ST	-24 hour hotline NOS
Bhatt, 2013[4] RCT	BPAP NOS	-Daily phone call by respiratory therapist during first week -One home visit by respiratory therapist during first week
Duiverman, 2011[22, 23] RCT	BPAP ST	-Supervision by specialized nurse NOS
Oscroft, 2010[48] Observational	BPAP ST started after AECOPD versus BPAP ST started in stable patient without exacerbation	-24 hour hotline NOS
Crespo, 2009[18] Observational	HMV (pressure or volume NOS)	-Emergency phone number
Cheung, 2010[12] RCT	BPAP ST versus CPAP	-Nurse hotline NOS
McEvoy, 2009[43] RCT	BPAP S	-Telephone calls answered by nurses as needed
Casanova, 2000[10] RCT	BPAP S	-"Close contact was maintained" for first 3 weeks
Garrod, 2000 [30] RCT	BPAP S	-Phone call every 2 weeks to encourage use
Clini, 1996[14] 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[37] RCT	BPAP ST	-24 hour hotline staffed by health-care providers and specialized nurses
Gay, 1996[28] RCT	BPAP ST	-Regular phone calls to ensure compliance.
Durao, 2018[25] RCT	HMV/BPAP mix	-Smoking cessation NOS
Tsolaki, 2011[66], 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[20] Observational	BPAP NOS	-Telephone helpline (24 hours)
Tsolaki, 2011[66] 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[60] 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[54] 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[20] Observational	BPAP NOS	-Telephone helpline (24 hours)	

Author, Year, Study Design	Device/Mode	Respiratory Services delivered in the home (including by whom and how frequently)	Outcome
Nauffal, 2002[47] Observational	BPAP NOS	-Telephone helpline (24 hours)	
Gonzalez-Bermejo, 2013[32] Observational	BPAP ST	-Instruction on assisted cough techniques including mechanical insufflation-exufflation by a respiratory physiotherapist	
Sancho, 2014[58] 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[17] 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[7] RCT	BPAP ST	-Multidisciplinary clinical team review, education about assisted cough techniques, posture, bed raisers, adjustable beds, palliative care, hospice as needed.	
Sancho, 2017[59] 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[26] 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[66]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[40 41] 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[6] 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[66] 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[44] Observational	NMD/TRD	HMV volume assist/control mode versus HMV volume control mode	-Telephone helpline
Chiang, 2003[13] 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

Appendix G. Guidelines

Table G.1. Guidelines for all conditions

Organization	Topic	KQ	Statement
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device initiation criteria	KQ1	Generally NIV should be commenced when there is evidence of: Daytime hypercapnia, PaCO2 ≥45mmHg and/or Evidence of nocturnal hypoventilation (in order of recommendation), such as: A rise in PaCO2 of ≥ 8mmHg between evening and morning ABGs or other accurate CO2 surrogate An acute peak rise of ≥ 8mmHg in TcCO2 or ETCO2 A rise in TcCO2 or ETCO2 > 50mmHg for more than 50% of total sleep time Whilst not ideal - when a measure of CO2 is not available - nocturnal oximetry demonstrates sustained oxygen desaturation ≤ 88% for 5 consecutive minutes or SpO2 <90% for >10% of total sleep time and Symptoms of significant sleep disordered breathing associated with nocturnal obstructive or hypopneoic events and/or Otherwise unexplained potential co-morbidity of sleep disorders, such as refractory hypertension, pulmonary hypertension, right heart failure, polycythaemia, cardiovascular disease or stroke.
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device initiation, monitoring, and candidate selection	KQ1	Hypoxia, hypercapnia, or an elevation in serum bicarbonate indicate the need for additional respiratory assessments and interventions. Polysomnography should be performed where there is a history suggestive of sleep disordered breathing or where FVC <40% predicted, base excess > +4mmols/L on arterial blood gases or erect to supine fall in VC of ≥ 25%. Consideration for polysomnography also includes symptoms of impaired sleep quality (such as daytime somnolence, waking headache or grogginess, fatigue, impaired cognition, impaired short-term memory, irritability, anxiety and depression) or symptoms of sleep-disordered breathing (such as frequent awakening, snoring, choking, gasping, waking dry mouth, waking dyspnea or witnessed apneas). Where there is no overt sign of respiratory compromise, serial VC, respiratory muscle testing, peak cough flow and oximetry should be performed to track baseline pulmonary function in suspected individuals. The minimum requirement for identifying sleep hypoventilation is overnight monitoring of oxygen saturation and, where possible, non-invasive carbon dioxide along with evening to morning arterial blood gases. When daytime indicators for NIV have already been met, a full diagnostic PSG measuring sleep quality is not an essential element in determining the need for NIV. Periodic nocturnal studies to identify unexpected problems or correct identified ones is indicated, with the frequency influenced by current response to therapy and the nature of the patient's underlying disorder. Minimum skills and level of knowledge need to be acquired by patients and / or their carers during the process of acclimatisation to NIV. Acclimatisation and education for domiciliary NIV should occur at institutions where there is a sufficient through-put of patients requiring long term NIV. The patient and/or carer are aware who to contact for medical and technical difficulties.
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device initiation, monitoring, and candidate selection	KQ1	The candidate (for home invasive or noninvasive ventilation) should be medically stable without constant or frequent monitoring, tests or treatment changes. The candidate and family must be motivated: Ventilator assisted individuals (VAIs) must express interest in transitioning/living in the community The family should express commitment to having the VAI live in the community.

Organization	Topic	KQ	Statement
			The family is willing to provide support (physical, emotional and financial). The candidate must have an adequate home setting : Identifiable home to live in, suitable to the needs of the VAI. Home is adaptable as necessary. The candidate must have sufficient caregiver support: Caregivers identified and committed to provide sufficient hours of care to meet the needs of the VAI. Available government-funded care hours identified. The candidate must have access to adequate financial resources: Sources of financial assistance identified and accessed. Sufficient financial resources available to meet projected costs The candidate must have access to equipment appropriate for the needs: Appropriate equipment selected and ordered. The candidate must have access to health care support in the community: Follow-up care available as appropriate (tracheotomy tube changes, ventilator reassessments and assessment of the ongoing effectiveness of the ventilatory support). Medical follow-up to allow for appropriate changes to the mode of ventilation (i.e., from invasive to noninvasive and vice versa, from continuous to nocturnal and vice versa). Professional services available post discharge. A government-funded ventilatory service is necessary to provide appropriate access to equipment and respiratory care.
British Thoracic Society, Guidelines for Home Oxygen Use in Adults, 2015[73]	Device initiation, monitoring, and candidate selection	KQ1	A blood gas assessment should be undertaken to exclude worsening hypercapnia and respiratory acidosis. Treatment with modalities of ventilatory support should be considered for patients who are hypercapnic. Patients with baseline hypercapnia can undergo LTOT assessment without adverse outcome but require monitoring of pH and PCO2 levels during and at the end of assessment. Patients with baseline hypercapnia should be monitored for the development of respiratory acidosis and worsening hypercapnia using ABGs after each titration of flow rate, as well as ABG sampling after oxygen titration is complete. Patients who develop a respiratory acidosis and/or a rise in PaCO2 of >1 kPa (7.5 mm Hg) during an LTOT assessment may have clinically unstable disease. These patients should undergo further medical optimization and be reassessed after 4 weeks. Patients who develop a respiratory acidosis and/or a rise in PaCO2 of >1 kPa (7.5 mm Hg) during an LTOT assessment on two repeated occasions, while apparently clinically stable, should only have domiciliary oxygen ordered in conjunction with nocturnal ventilatory support.
Clinical Indications for Noninvasive Positive Pressure Ventilation in Chronic Respiratory Failure due to Restrictive Lung Disease, COPD, and Nocturnal Hypoventilation—A Consensus Conference	Device initiation, monitoring, and candidate selection	KQ1	Certificate of Medical Necessity Should Document diagnosis Document indications Provide required settings Inspiratory parameters (such as tidal volume, pressure, inspiratory time, cycle) Expiratory pressures Rate (as clinically indicated) Supplemental oxygen (flow rate or fraction of inspired oxygen) Alarms (as clinically indicated)

Organization	Topic	KQ	Statement
Report, American Academy of Home Care Physicians, American College of Chest Physicians, American College of Physicians, American Sleep Disorders Association, American Thoracic Society, National Association for Medical Direction of Respiratory Care, 1999[74]			
Clinical Indications for Noninvasive Positive Pressure Ventilation in Chronic Respiratory Failure due to Restrictive Lung Disease, COPD, and Nocturnal Hypoventilation—A Consensus Conference Report, American Academy of Home Care Physicians, American College of Chest Physicians, American College of Physicians, American Sleep Disorders Association, American Thoracic Society, National Association for Medical Direction of Respiratory Care, 1999[74]	Device continuation, compliance, and outcomes	KQ1 and KQ2	It is expected that initial settings may be adjusted by personnel experienced and skilled in the treatment of NIPPV under the direction of the treating physician. At the 60-day reassessment, final settings must be documented Monitoring of Effectiveness Physician reassessment of patient adherence with the use of NIPPV at 30 to 60 days (documentation of machine usage average of ≥20 h/week) Ongoing monitoring and yearly recertification by physician
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device continuation, compliance, and outcomes	KQ1 and KQ2	Usage throughout all sleep periods should be recommended. Once established on therapy, regular monitoring of compliance data should be performed and compliance is deemed adequate at > 4 - 6 hours per night. Patients can be reviewed at 6 to 8 weeks following the commencement of NIV to determine the clinical response to therapy. After initiation of NIV, clinical review should occur within the first 2 to 3 months to assess symptoms, technical problems, ventilator settings, compliance and success. Further clinical reviews should be performed by a Sleep Physician / Respiratory Physician or Respiratory

Organization	Topic	KQ	Statement
			Failure clinic every 6 to 12 months, again assessing symptoms, compliance, technical problems, lung function, oximetry and further investigations (including ABGs and overnight oximetry or PSG) as required. At any time, when there are indications of unsatisfactory results like the recurrence of clinical symptoms or awake blood gases deteriorate despite clinical stability (e.g. absence of recent pulmonary infection) and adequate compliance, then inadequate ventilation must be suspected and objective evaluation during sleep must be undertaken. Outcome measures should include awake ABGs, nocturnal SpO2 and assessment of daytime sleepiness, breathlessness and health related quality of life.
German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010[75]	Device continuation, compliance, and outcomes	KQ1 and KQ2	Initialization of HMV must take place in a centre for HMV. The aim of the therapy is to eliminate hypoventilation under mechanical ventilation, as well as to reduce CO2 to the point of normocapnia during daytime spontaneous breathing. Once optimal ventilation has been achieved, criteria for supplementary oxygen supply must be assessed. The first ventilation control visit must occur in the short-term (4–8 weeks) and therapeutic success is evaluated according to subjective, clinical and technically-measurable parameters. Modifications to the ventilation system (e. g. parameters, ventilation-interface) must take place exclusively in conjunction with the centre for HMV. Identically-built machines with the same settings can be exchanged outside the hospital, whereas different machines must be exchanged under hospital conditions in the centre for HMV.
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device characteristics and titration	KQ3	Simple bilevel devices are suitable for individuals requiring nocturnal and limited daytime ventilatory support only. However, more sophisticated volume or hybrid devices are indicated for patients requiring more than 18 hours/day or where bilevel devices have proven to be inadequate. Ventilator dependent individuals should be titrated on and use ventilators which have been approved for life support and have an alternative battery source to mains power. They also should be supplied with an appropriate back-up ventilator. Machines with “mask off” or “low pressure” and “power failure” alarms are recommended for ventilator dependent patients and in disorders where there is a potential inability to arouse from an interruption to ventilation or when there is an absence of ventilatory responses when awake. Titration for long term NIV settings should occur when the patient is chronically stable (pH>7.35) and free from exacerbation. Adequate IPAP-EPAP difference is required to ameliorate hypoventilation. A Bi-level ventilation should be commenced in the spontaneous mode, unless there is specific evidence that the patient is unable to trigger the machine once baseline leak and settings have been optimized. Complete correction of sleep disordered breathing during the initial titration night is not necessary for improvement of daytime blood gases and symptoms to occur. Spontaneous-timed mode flow generator, or a ventilator, to be provided if Spontaneous mode device does not allow correction of sustained hypercapnia in the presence of central apnea or persisting hypoventilation. Ventilators using flow triggering or volume-cycled mandatory ventilation may be required for patients experiencing difficulty in triggering inspiration.
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute	Device characteristics and titration	KQ3	Pressure-targeted ventilators are the devices of choice for acute NIV. A full face mask (FFM) should usually be the first type of interface used. A range of masks and sizes is required and staff involved in delivering NIV need training in and experience of using them. NIV circuits must allow adequate clearance of exhaled air through an exhalation valve or an integral exhalation

Organization	Topic	KQ	Statement
Hypercapnic Respiratory Failure in Adults, 2016[76]			port on the mask. As patients recover from acute hypercapnic respiratory failure, ventilator requirements change and ventilator settings should be reviewed regularly.
German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010[75]	Device characteristics and titration	KQ3	A second ventilator and an external battery pack are necessary if ventilation periods exceed 16 hours/day. Every non-invasively-ventilated patient requires at least one reserve mask A humidifier is a mandatory requirement for invasive ventilation and is also useful for non-invasive ventilation if typical symptoms are present. In NMD patients with cough insufficiency and in children, selective use of a pulse oximeter is necessary.
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Respiratory services	KQ4	Education and preventive strategies in airway clearance must precede the need for mechanical ventilation whenever possible. In the absence of contraindications, lung volume recruitment (i.e. air stacking) techniques should be introduced with the measurement of peak cough flows and maximum insufflation capacity in those with peak cough flows <270 L/min. Manually assisted coughing is recommended alone or in addition to lung volume recruitment to increase peak cough flows to >270 L/min. In the absence of contraindications, mechanical in-exsufflation should be recommended for patients unable to achieve peak cough flows >270 L/min with lung volume recruitment and/or manually assisted coughing, particularly during respiratory infection. A government-funded ventilatory service is necessary to provide appropriate access to equipment and respiratory care.

ABG: arterial blood gases, CO2: carbon dioxide, ETCO2: end tidal carbon dioxide, EPAP: expiratory positive airway pressure, FVC: Forced vital capacity, HMV: home mechanical ventilation, IPAP: inspiratory positive airway pressure, kPa: kilopascal, KQ: key question, LTOT: long term oxygen therapy, mmHg: millimeters of mercury (pressure), mmols: millimoles, NIPPV/NPPV: non-invasive positive pressure ventilation, NIV: non-invasive ventilation, NMD: neuromuscular disease, PCO2/PaCO2: partial pressure of arterial carbon dioxide, pH: potential of hydrogen, PSG: polysomnogram, SpO2: Blood oxygen saturation level, TcCO2/tCO2: transcutaneous carbon dioxide pressure, VAI: ventilator assisted individual, VC: vital capacity

Table G.2. Guidelines for COPD

Organization	Topic	KQ	Statement
German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010[75]	Device initiation criteria	KQ1	Symptoms that indicate CRF and reduced quality of life in COPD patients as well as one of the following criteria (at least 1 criterion must be fulfilled) indicate the need for HMV: Chronic daytime hypercapnia with PaCO2 ≥ 50mmHg Nocturnal hypercapnia with PaCO2 ≥ 55mmHg Stable daytime hypercapnia with 46–50mmHg and a rise in PTcCO2 to ≥ 10mmHg during sleep. Stable daytime hypercapnia with PaCO2 46–50mmHg and at least 2 acute exacerbations accompanied by respiratory acidosis that required hospitalization within the last 12 months Following an acute exacerbation needing ventilatory support, according to clinical estimation. Poor compliance with medication intake and/or LTOT are relative contraindications. Complete discontinuation of nicotine abuse should be aspired to. NIV is the primary treatment option for HMV of COPD patients with CRF. The most important criteria for the advent of long-term NIV are the presence of hypercapnia in combination with the typical symptoms of ventilatory failure, recurring exacerbations and the reduction in quality of life.
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device initiation criteria	KQ1	Nocturnal NIV is indicated in COPD with PaCO2 > 50 mmHg, where there is evidence of signs and symptoms of sleep disordered breathing, and full PSG demonstrates nocturnal hypoventilation (based on a measure of PaCO2) that is not corrected or made worse by LTOT alone.
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device initiation criteria	KQ1	The use of long-term NIPPV cannot be widely recommended in patients with stable COPD. Long-term NIPPV in COPD should only be considered on an individual basis. One subgroup of patients with COPD in which long-term NIPPV could be considered are those with severe hypercapnia (PaCO2 >55 mmHg) experiencing repeated episodes of acute hypercapnic respiratory failure that require in-hospital ventilatory support. However, definitive proof of efficacy of long-term NIPPV in these patients will need to await future studies. The overlap syndrome, and concomitant COPD and OSA syndrome, should be differentiated from chronic respiratory failure that is solely due to advanced COPD.
8th International Conference on Management and Rehabilitation of Chronic Respiratory Failure, Pescara, Italy, 2015[77]	Device initiation criteria	KQ1	The role of long-term non invasive positive pressure ventilation in improving survival in COPD patients with CRF (chronic respiratory failure) is still discussed. There is simply not enough evidence to support it. Long-term non invasive ventilation should be reserved to individual patients. Once stable hypercapnia is proven, NIPPV may improve survival and health status. Therefore, despite recent studies adding some new data, the authors cannot recommend the widespread use of this therapeutic intervention after an episode of acute-on-chronic respiratory failure in COPD. Long-term night non invasive ventilation in these patients has some physiological and clinical benefits.
Clinical Indications for Noninvasive Positive Pressure Ventilation in Chronic Respiratory Failure due to Restrictive Lung Disease, COPD, and Nocturnal Hypoventilation—A Consensus	Device initiation criteria	KQ1	Indications for usage: symptoms (such as fatigue, dyspnea, morning headache, etc.) and physiologic criteria (one of the following): PaCO2 > 55 mm Hg; PaCO2 of 50 to 54 mm Hg and nocturnal desaturation (oxygen saturation by pulse oximeter 88% for 5 continuous minutes while receiving oxygen therapy 2 L/ min); or PaCO2 of 50 to 54 mm Hg and hospitalization related to recurrent (2 in a 12- month period) episodes of hypercapnic respiratory failure

Organization	Topic	KQ	Statement
Conference Report, American Academy of Home Care Physicians, American College of Chest Physicians, American College of Physicians, American Sleep Disorders Association, American Thoracic Society, National Association for Medical Direction of Respiratory Care, 1999[74]			
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Device initiation criteria	KQ1	In acute hypercapnic respiratory failure in the hospital, NIV should be started when pH<7.35 and pCO2 >6.5 kPa persist or develop despite optimal medical therapy. In acute hypercapnic respiratory failure in the hospital, NIV can be discontinued when there has been normalization of pH and pCO2 and a general improvement in the patient's condition.
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device initiation, monitoring, and candidate selection	KQ1	Recurrent hospitalizations (2 or more in a year) for acute hypercapnic respiratory failure (especially life threatening events) or difficulty weaning from invasive ventilation are an indicator for assessment for domiciliary NIV.
Clinical Indications for Noninvasive Positive Pressure Ventilation in Chronic Respiratory Failure due to Restrictive Lung Disease, COPD, and Nocturnal Hypoventilation—A Consensus Conference Report, American Academy of Home Care Physicians, American College of Chest Physicians, American College of Physicians, American Sleep Disorders Association, American Thoracic Society, National Association for Medical Direction of Respiratory Care, 1999[74]	Device initiation, monitoring, and candidate selection	KQ1	Before considering a COPD patient for NIPPV, a physician with skills and experience in NIPPV must establish and document an appropriate diagnosis on the basis of history, physical examination, and results of diagnostic tests, and assure optimal management of COPD with such treatments as bronchodilators, oxygen when indicated, and optimal management of other underlying disorders (such as performing a multichannel sleep study to exclude associated sleep apnea if clinically indicated)
United States Department of Veterans Affairs, the Department of Defense, and the National Guideline	Device initiation, monitoring, and candidate selection	KQ1	In the absence of other contributors (e.g., sleep apnea), we suggest referral for a pulmonary consultation in patients with stable, confirmed COPD and hypercapnia.

Organization	Topic	KQ	Statement
Clearinghouse Clinical Practice Guideline for the Management of Chronic Obstructive Pulmonary Disease, 2014[78]			
United Kingdom National Institute for Health and Care Excellence (NICE), Chronic Obstructive Pulmonary Disease in Over 16s: Diagnosis and Management, 2010[79]	Device initiation, monitoring, and candidate selection	KQ1	Adequately treated patients with chronic hypercapnic ventilatory failure who have required assisted ventilation (whether invasive or non-invasive) during an exacerbation or who are hypercapnic or acidotic on LTOT should be referred to a specialist centre for consideration of long-term NIV. Patients with severe disease requiring interventions such as long-term non-invasive ventilation should be reviewed regularly by specialists.
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device continuation, compliance, and outcomes	KQ1 and KQ2	Changes in awake blood gases are not the best measure of effectiveness of NIV in chronic hypercapnic COPD. Changes in symptoms including exertional dyspnoea, control of nocturnal hypoventilation, reduction in hospital admissions and QoL (SF-36) are better indicators of the patient's response to therapy.
German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010[75]	Device characteristics and titration	KQ3	The aim of the ventilation is to normalize PaCO2; sufficiently high ventilation pressures are required to achieve this. Controlled ventilation mode with ventilation pressures from 20 to 40 mbar. Pressure escalation until normocapnia or maximum tolerance is reached. Rapid increase in inspiratory pressure (0.1 to 0.2 seconds) PEEP can be useful for assisted- or assisted-controlled ventilation. Minimal duration of therapy: 4.5 hours/day The introduction of NIV in the hospital can take up to two weeks.
Clinical Indications for Noninvasive Positive Pressure Ventilation in Chronic Respiratory Failure due to Restrictive Lung Disease, COPD, and Nocturnal Hypoventilation—A Consensus Conference Report, American Academy of Home Care Physicians, American College of Chest Physicians, American College of Physicians, American Sleep Disorders Association, American Thoracic Society, National Association for Medical Direction of Respiratory Care, 1999[74]	Device characteristics and titration	KQ3	NIPPV appears to be better tolerated in this patient population than negative pressure ventilation. In addition, advantages of ease of administration and portability as well as the ability to eliminate obstructive sleep apneas make NIPPV the noninvasive mode of first choice.
8th International Conference	Respiratory		Telemonitoring in ventilator dependent patients: 1) Home mechanical ventilators may be equipped with

Organization	Topic	KQ	Statement
on Management and Rehabilitation of Chronic Respiratory Failure, Pescara, Italy, 2015[77]	services		remote monitoring tools in order to improve physician supervision, with the aim to adapt settings to the needs and comfort of the patient. 2) Economic, regulatory and legal impacts of home telemonitoring will be important in its adaption by health care systems. 3) Relevant issues are prescription criteria, modalities of follow-up, team expertise, technologies, adherence, bundling of services, and outcomes

COPD: chronic obstructive pulmonary disease, CRF: chronic respiratory failure, HMV: home mechanical ventilation, kPa: kilopascal, KQ: key question, LTOT: long term oxygen therapy, NIPPV/NPPV: non-invasive positive pressure ventilation, NIV: non-invasive ventilation, OSA: obstructive sleep apnea, PCO2/PaCO2: partial pressure of arterial carbon dioxide, PEEP: positive end expiratory pressure, pH: potential of hydrogen, PSG: polysomnogram, QoL: quality of life, SF-36: Medical Outcomes Study Questionnaire Short Form, tCO2/ PTcCO2: transcutaneous carbon dioxide pressure

Table G.3. Guidelines for Neuromuscular Disease

Organization	Topic	KQ	Statement
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device initiation criteria	KQ1	The institution of NIV is recommended in patients with rapidly progressive respiratory muscle weakness associated with orthopnoea, hypercapnia or symptomatic sleep hypoventilation (sleep fragmentation/ daytime hypersomnolence/ morning headaches and cognitive dysfunction).
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Device initiation criteria	KQ1	Planned elective domiciliary NIV is preferable to crisis management in NMD and chest wall disorders. This reduces the risk of acute presentation and provides a proven alternative to invasive mechanical ventilation which risks prolonged or permanent tracheostomy ventilation. NIV should almost always be trialled in the acutely unwell patients with NMD or CWD with hypercapnia. Do not wait for acidosis to develop. In patients with NMD or CWD, NIV should be considered in acute illness when vital capacity (VC) is known to be <1 L and RR >20, even if normocapnic. In patients with NMD or CWD, nocturnal NIV should usually be continued following an episode of AHRF, pending discussion with a home ventilation service.
United Kingdom National Institute for Health and Care Excellence (NICE), Motor Neuron Disease: Assessment and Management, 2016[80]	Device initiation criteria	KQ1	If the person's SpO2 (measured at rest and breathing room air) is greater than 94%, or 92% for those with lung disease, but they have sleep-related respiratory symptoms: Consider referring them to a respiratory ventilation service for continuous nocturnal (overnight) oximetry and/or a limited sleep study and discuss both the impact of respiratory impairment and treatment options with the patient and (if the person agrees) their family and carers. If the person's arterial partial pressure of carbon dioxide (PaCO2) is greater than 6 kPa: refer them urgently to a respiratory ventilation service (to be seen within 1 week) and explain the reasons for and implications of the urgent referral to the person and (if the person agrees) their family and carers. If the person's PaCO2 is less than or equal to 6 kPa but they have any symptoms or signs of respiratory impairment, particularly orthopnoea refer them to a respiratory ventilation service for nocturnal (overnight) oximetry and/or a limited sleep study and discuss both the impact of respiratory impairment and treatment options with the person and (if the person agrees) their family and/or carers (as appropriate). If any of the results listed in box 2 is obtained, discuss with the person and (if appropriate) their family and carers: their respiratory impairment their treatment options possible referral to a respiratory ventilation service for further assessment based on discussion with the person, and their wishes.

Organization	Topic	KQ	Statement
Clinical Indications for Noninvasive Positive Pressure Ventilation in Chronic Respiratory Failure due to Restrictive Lung Disease, COPD, and Nocturnal Hypoventilation—A Consensus Conference Report, American Academy of Home Care Physicians, American College of Chest Physicians, American College of Physicians, American Sleep Disorders Association, American Thoracic Society, National Association for Medical Direction of Respiratory Care, 1999[74]	Device initiation criteria	KQ1	Indications for usage Symptoms (such as fatigue, dyspnea, morning headache, etc.) and one of the following Physiologic criteria (one of the following PaCO2 ≥ 45 mm Hg Nocturnal oximetry demonstrating oxygen saturation ≤ 88% for 5 consecutive minutes For progressive neuromuscular disease, maximal inspiratory pressures < 60 cm H2O or FVC <50% predicted
German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010[75]	Device initiation criteria	KQ1	NIV of NMD patients with clinical signs of CRF is indicated by the following (at least 1 criterion should be fulfilled): Chronic daytime hypercapnia with PaCO2 ≥ 45mmHg Nocturnal hypercapnia with PaCO2 ≥ 50mmHg Daytime normocapnia with a rise in PTcCO2 of ≥ 10mmHg during the night A rapid, significant reduction in VC At the first signs of nocturnal hypercapnia, the patient should be offered NIV therapy rather than waiting until the hypercapnia extends into the daytime period. There are no indications for prophylactic mechanical ventilation in the absence of symptoms or hypoventilation. NIV is also indicated prior to elective vertebral column correction surgery when VC < 60% target value and FEV1 < 40% target value, respectively, or during pregnancy with restricted lung function, as well as palliative care of dyspnea. Patients with NMD should undergo clinical assessment and assessment of VC at 3–12 month-intervals. Polygraphy and PTcCO2-measurement are indicated when VC is < 70%. NIV is the primary treatment option for HMV of NMD patients with CRF; in cases of inviability, failure or rejection of NIV, invasive HMV should only be established in accordance with the explicit wishes of the patient and custodian, respectively. The most important criteria for the initiation of NIV are hypercapnia in combination with the characteristic symptoms of ventilatory failure, and a reduction in quality of life.
American Academy of Neurology, Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: multidisciplinary care, symptom management, and cognitive/behavioral impairment (an evidence-based review), 2009[81]	Device initiation Criteria (ALS)	KQ1	NIV may be considered at the earliest sign of nocturnal hypoventilation or respiratory insufficiency in order to improve compliance with NIV in patients with ALS. NIV may be considered to enhance QOL in patients with ALS who have respiratory insufficiency.

Organization	Topic	KQ	Statement
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device initiation Criteria (ALS)	KQ1	NIV should be offered to patients with any one of the following: Orthopnea Daytime hypercapnia Symptomatic sleep disordered breathing FVC <50% predicted SNP <40 cmH2O or PImax<40 cmH2O
American Thoracic Society, Respiratory Care of the Patient with Duchenne Muscular Dystrophy, 2004[82]	Device initiation criteria (Duchenne Muscular Dystrophy)	KQ1	Consider daytime ventilation when measured waking Pco2 exceeds 50 mm Hg or when hemoglobin saturation remains < 92% while awake.
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device initiation criteria (Duchenne Muscular Dystrophy)		Offer nocturnal NIV to patients with diurnal hypercapnia (daytime arterial PCO2 >45 mmHg), or when there is documented nocturnal hypercapnia and the presence of symptoms consistent with hypoventilation. Institution of NIV during sleep should be offered to patients demonstrating a major degree of nocturnal hypoxemia, even if asymptomatic.
German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010[75]	Device initiation, monitoring, and candidate selection	KQ1	Specific aspects in the ventilation of patients with NMD comprise: Muscle weakness in the oropharyngeal area, carrying the risk of reduced ability or complete inability to close the mouth Bulbar symptoms with the risk of recurrent aspiration Hypersalivation; therapy with anti-cholinergics (e. g. Scopolamine patch, amitryptiline or botulinum toxin injections into the salivary glands) Coughing weakness, with the development of acute decompensation
British Thoracic Society, Guidelines for Home Oxygen Use in Adults, 2015[73]	Device initiation, monitoring, and candidate selection	KQ1	Non-invasive ventilation (NIV) should be the treatment of choice for patients with NMD or chest wall disease causing type 2 respiratory failure.
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Device initiation, monitoring, and candidate selection	KQ1	In patients with NMD or CWD, senior/experienced input is needed in care planning and is essential if differences in opinion exist or develop between medical staff and patient representatives. In patients with NMD, it should be anticipated that bulbar dysfunction and communication difficulties, if present, will make NIV delivery difficult, and may make it impossible. Discussion about NIV and IMV, and patients' wishes with respect to cardiopulmonary resuscitation, should occur as part of routine care of patients with NMD or CWD. In patients with NMD or chest wall diseases, senior staff should be involved in decision-making, in conjunction with home mechanical ventilation specialists, if experience is limited, and especially when the appropriateness of invasive mechanical ventilation is questioned. Domiciliary NIV is effective in treating chronic hypercapnia, improves long-term survival and preserves a good or acceptable QoL

Organization	Topic	KQ	Statement
United Kingdom National Institute for Health and Care Excellence (NICE), Motor Neuron Disease: Assessment and Management, 2016[80]	Device initiation, monitoring, and candidate selection	KQ1	Assess and monitor the person's respiratory function and symptoms. Treat people with NMD and worsening respiratory impairment for reversible causes (for example, respiratory tract infections or secretion problems) before considering other treatments. Offer non-invasive ventilation as treatment for people with respiratory impairment. Decisions to offer non-invasive ventilation should be made by the multidisciplinary team in conjunction with the respiratory ventilation service, and the person. Consider urgent introduction of non-invasive ventilation for people with NMD who develop worsening respiratory impairment and are not already using non-invasive ventilation. As part of the initial assessment to diagnose NMD, or soon after diagnosis, a healthcare professional from the multidisciplinary team who has appropriate competencies should perform the following tests (or arrange for them to be performed) to establish the person's baseline respiratory function: oxygen saturation measured by pulse oximetry (SpO2): this should be a single measurement of SpO2 with the person at rest and breathing room air if it is not possible to perform pulse oximetry locally, refer the person to a respiratory ventilation service. Then one or both of the following: forced vital capacity (FVC) or vital capacity (VC) sniff nasal inspiratory pressure (SNIP) and/or maximal inspiratory pressure (MIP). If the person has severe bulbar impairment or severe cognitive problems that may be related to respiratory impairment: ensure that SpO2 is measured (at rest and breathing room air) do not perform the other respiratory function tests (FVC, VC, SNIP and MIP) if interfaces are not suitable for the person. A healthcare professional with appropriate competencies should perform the respiratory function tests every 2–3 months, although tests may be performed more or less often depending on: whether there are any symptoms and signs of respiratory impairment (see box 1) the rate of progression of NMD the person's preference and circumstances. [2010, amended 2016] Perform arterial or capillary blood gas analysis if the person's SpO2 (measured at rest and breathing room air): is less than or equal to 92% if they have known lung disease is less than or equal to 94% if they do not have lung disease. If it is not possible to perform arterial or capillary blood gas analysis locally, refer the person to a respiratory ventilation service.

Organization	Topic	KQ	Statement
Clinical Indications for Noninvasive Positive Pressure Ventilation in Chronic Respiratory Failure due to Restrictive Lung Disease, COPD, and Nocturnal Hypoventilation—A Consensus Conference Report, American Academy of Home Care Physicians, American College of Chest Physicians, American College of Physicians, American Sleep Disorders Association, American Thoracic Society, National Association for Medical Direction of Respiratory Care, 1999[74]	Device initiation, monitoring, and candidate selection	KQ1	Disease documentation Before considering a restrictive thoracic patient for NIPPV, a physician with skills and experience in NIPPV must establish and document an appropriate diagnosis on the basis of history, physical examination, and diagnostic tests and assure optimal treatment of other underlying disorders (such as performing a multichannel sleep study to detect associated sleep apnea if clinically indicated) The most common disorders would include sequelae of polio, spinal cord injury, neuropathies, myopathies and dystrophies, ALS, chest wall deformities, and kyphoscoliosis.
British Thoracic Society, Guidelines for Home Oxygen Use in Adults, 2015[73]	Device initiation, monitoring, and candidate selection	KQ1	Patients with neuromuscular weakness affecting respiratory muscles should not have nocturnal oxygen therapy alone ordered. It can be considered in patients with evidence of established ventilatory failure, where it should be given with NIV support.

Organization	Topic	KQ	Statement
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device initiation, monitoring, and candidate selection	KQ1	Subjects with progressive respiratory muscle weakness and other restrictive thoracic disorders should be observed regularly with lung function (VC, MIP, MEP, SNP and PCF) and oximetry. An arterial blood gas should be performed especially if VC < 40% predicted or MIP < 60 cmH2O. <u>Slowly progressive NMD</u> Hypoxia, hypercapnia, or an elevation in serum bicarbonate indicate the need for additional respiratory assessments and interventions. All subjects with DMD should be referred for clinical assessment initially to a paediatric specialist unit for assessment and then care transferred to an adult centre when age >18 years. Assessment as to the risk of development of progressive respiratory failure should be considered in all subjects with other progressive neuromuscular disorders. Referral to a specialist centre should occur if significant respiratory muscle weakness or sleep disordered breathing occurs. Patients should have access to other specialist health providers, including medical specialists and allied health professionals, preferably in a well co-ordinated multidisciplinary team. <u>Rapidly progressive NMD</u> Patients with NMD are recommended to have 3 monthly clinical evaluation to monitor for symptoms and signs of respiratory and sleep complications. Sniff nasal inspiratory pressure and overnight oximetry are the initial investigations of choice for the assessment of early respiratory muscle compromise and nocturnal hypoventilation. A diagnostic polysomnogram should be reserved for patients in whom co-existent upper airway obstruction is suspected on clinical grounds with inconclusive nocturnal oximetry. While NMD patients with significant bulbar dysfunction should still have the option to trial NIV, it should be recognized that this group of patients may have reduced tolerance to and derive less benefit from NIV. The progression to tracheostomy intermittent positive pressure ventilation (TIPPV) should be made on an individual basis, weighing the longer survival advantage with a significantly greater burden of care and cost to the patient, carer and/or community and recognizing that QoL improvements associated with the use of NIV may not be seen with TIPPV to an equivalent degree. The elective commencement of NIV is preferred over non-elective TIPPV despite the improved survival advantage. Patients with NMD should be managed in a multidisciplinary clinic as this improves survival and QoL, and facilitates earlier uptake of interventions including NIV and PEG insertion.

Organization	Topic	KQ	Statement
European Federation of Neurological Societies (EFNS) Guidelines on the Clinical Management of Amyotrophic Lateral Sclerosis, 2012[83]	Device initiation, monitoring, and candidate selection (ALS)	KQ1	Symptoms or signs of respiratory insufficiency (including symptoms of nocturnal hypoventilation) should be checked at each visit. Forced vital capacity and vital capacity are the most available and practical tests for the regular monitoring of respiratory function. Sniff nasal pressure may be used for monitoring, particularly in bulbar patients with weak lips. Percutaneous nocturnal oximetry is recommended as a screening test and for monitoring respiratory function. Symptoms or signs of respiratory insufficiency should prompt discussions with the patient and caregivers about treatment options and the terminal phase. Early discussions are needed to allow advance planning and directives. NIPPV should be considered in preference to IMV in patients with symptoms or signs of respiratory insufficiency. NIPPV can prolong survival for many months and may improve the patient's quality of life IMV has a major impact upon caregivers and should be initiated only after informed discussion. Unplanned (emergency) IMV should be avoided through an early discussion of end-of-life issues, coordination with palliative care teams and appropriate advance directives. Oxygen therapy alone should be avoided as it may exacerbate carbon dioxide retention and oral dryness. Use oxygen only if symptomatic hypoxia is present.
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device initiation, monitoring, and candidate selection (ALS)	KQ1	Regular monitoring of ALS patients is advised from the time of diagnosis every two to six months and varies with anticipated rapidity of disease progression and should include the following: Symptom review to include orthopnea, dyspnea, poor sleep, excessive daytime sleepiness, poor concentration, morning headache. Measurement of sitting FVC. Measurement of one or more of the following: supine VC, sniff nasal pressure, Pimax (MIP). Measurement of ABGs or end tidal CO2 (ETCO2) when hypercapnia is suspected. Nocturnal oximetry ± transcutaneous CO2 (tCO2) when symptomatic sleep disordered breathing is suspected. Measurement of peak cough flow. NIV should be considered the preferred option for ventilation even when ventilation is required 24 h per day. Elective tracheostomy ventilation may be considered, and is dependent on regional resources and careful discussion with the patient and caregivers. Long-term invasive ventilation can be offered after acute respiratory failure requiring invasive ventilation, if the patient and caregivers fully understand the consequences and appropriate support is available.

Organization	Topic	KQ	Statement
American Thoracic Society, Respiratory Care of the Patient with Duchenne Muscular Dystrophy, 2004[82]	Device initiation, monitoring, and candidate selection (Duchenne Muscular Dystrophy)	KQ1	In centers with appropriate expertise, consider mouthpiece intermittent positive pressure ventilation or other forms of noninvasive daytime ventilation. Consider tracheostomy when contraindications or patient aversion to noninvasive ventilation are present. Patients receiving noninvasive ventilation should have regular (at least annual) noninvasive monitoring of gas exchange, including oxygen saturation and end-tidal Pco2 levels. Discussions regarding ventilatory support for each patient should involve the patient, caregivers, and medical team. Where CO2 monitoring is not available, overnight pulse oximetry can be used to detect nighttime oxyhemoglobin desaturation. Simple oximetry provides, at best, only indirect information on ventilation, and should be used to assess need for ventilatory support only when better alternatives are unavailable. Schedule periodic reassessment as appropriate to stage of disease. Follow-up visits should include monitoring for the development of daytime hypoventilation, which may necessitate around-the-clock ventilation. Use nasal intermittent positive pressure ventilation to treat sleep-related upper airway obstruction and chronic respiratory insufficiency in patients with DMD. Negative-pressure ventilators should be used with caution in patients with DMD due to the risk of precipitating upper airway obstruction and hypoxemia. Do not use oxygen to treat sleep-related hypoventilation without ventilatory assistance. Objective evaluation at each clinic visit should include: oxyhemoglobin saturation by pulse oximetry, spirometric measurements of FVC, FEV1, and maximal mid-expiratory flow rate, maximum inspiratory and expiratory pressures, and peak cough flow. Awake carbon dioxide tension should be evaluated at least annually in conjunction with spirometry. Where available, capnography is ideal for this purpose. Arterial blood gas analysis is not necessary for routine follow-up of patients with DMD. If capnography is not available, then a venous or capillary blood sample should be obtained to assess for the presence of alveolar hypoventilation. Additional measures of pulmonary function and gas exchange may be useful, including lung volumes, assisted cough peak flow, and maximum insufflation capacity. Carefully evaluate patients for evidence of other respiratory disorders, such as obstructive sleep apnea, oropharyngeal aspiration, gastroesophageal reflux, and asthma. Annual laboratory studies in patients requiring a wheelchair for ambulation should include a complete blood count, serum bicarbonate concentration, and a chest radiograph.
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device initiation, monitoring, and candidate selection (Duchenne Muscular Dystrophy)	KQ1	Carefully question and educate patients to report symptoms consistent with hypoventilation, including disturbed sleep, excessive daytime sleepiness, morning headache and weight loss. -Measure VC, MIP, maximal expiratory pressure, peak cough flow and awake oxyhemoglobin saturation by pulse oximetry at least yearly; if VC <40% predicted, also monitor awake CO2 tension by noninvasive methods or ABG analysis. Perform an evaluation of ventilation during sleep if there are symptoms consistent with nocturnal hypoventilation or other forms of sleep disordered breathing. In the absence of such symptoms, periodic screening for sleep disordered breathing should also be considered once FEV1 or FVC is <40% predicted.

Organization	Topic	KQ	Statement
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device initiation, monitoring, and candidate selection (Other myopathies)	KQ1	Obtain periodic clinical assessment and spirometry at six- to 12-month intervals, including sitting (plus supine if diaphragmatic weakness is suspected) spirometric testing. Consider monitoring for sleep disordered breathing in patients with VC <60%. Consider ABGs or nocturnal measure of CO2 in patients with VC <40% to exclude hypercapnia. NIV should be offered when there is daytime hypercapnia or symptomatic nocturnal hypoventilation. Assess airway clearance ability with peak cough flows and implement cough-assistance strategies
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device initiation, monitoring, and candidate selection (Myotonic dystrophy)	KQ1	Obtain six to 12 monthly clinical assessment of symptoms of daytime or nocturnal hypoventilation. Obtain yearly VC and consider daytime *Pa*CO2 measurement, even with mild reductions of VC when patients exhibit symptoms of hypoventilation. Consider overnight oximetry or polysomnography when there are symptoms of nocturnal hypoventilation. Long-term NIV should be offered to patients with daytime hypercapnia or symptomatic nocturnal hypoventilation as for other NMDs Carefully assess motivation and ability to adhere to treatment with patients and their caregivers before initiating long-term ventilatory support. Reassess every six months to verify treatment adherence and provide extra help and motivation as needed.
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device initiation, monitoring, and candidate selection (Post-polio syndrome)	KQ1	Yearly assessment of VC is recommended from the time of presentation of post polio syndrome. If VC >50% with symptoms of hypoventilation, perform measurements of daytime ABGs, overnight oximetry and consider polysomnography. When VC <50%, perform ABG analysis and/or nocturnal oximetry yearly. With confirmation of the presence of chronic hypoventilation, offer NIV.
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device initiation, monitoring, and candidate selection (Spinal cord injury)	KQ1	Each patient must be individually evaluated for the need for long-term ventilation either acutely or in follow-up. Noninvasive support is preferable to invasive ventilation. Phrenic nerve pacing is recommended in selected individuals as an alternative to positive pressure ventilation alone. In the long term, individuals with SCI require regular monitoring to identify the development of sleep disordered breathing or respiratory failure and evaluate the need for NIV.
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device initiation, monitoring, and candidate selection (Spinal cord injury)	KQ1	NIV is indicated when there is intractable or refractory sputum retention, atelectasis, respiratory tract infection or type-I respiratory failure (PaO2 < 80 mmHg, SpO2 <95%). NIV is indicated when there is intolerance of CPAP for treatment of OSA, especially in cases of SCI at C6 or above. Use of an abdominal binder may be considered as the initial intervention in cases of mild hypoventilation, or as an adjunct to the use of NIV. The implementation of NIV should occur in a specialised centre where there is access to a spinal unit, accredited pulmonary function and sleep laboratory, physician experienced in the use of NIV, NIV service and physiotherapy service trained in secretion removal in patients with spinal cord injury.

Organization	Topic	KQ	Statement
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device characteristics and titration (Duchenne Muscular Dystrophy)	KQ3	When bilevel ventilation is used, backup respiratory rates are recommended during sleep while on NIV to reduce the work of breathing associated with breath initiation. Individualize the decision about the transition from nocturnal NIV to daytime ventilation by carefully evaluating patient factors (symptoms, bulbar involvement, patient preference, etc.) and available resources. In patients requiring daytime ventilation, strongly consider mouthpiece ventilation as an alternative to invasive tracheostomy.
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Device characteristics and titration	KQ3	In patients with NMD or CWD, consider controlled ventilation as triggering may be ineffective.
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device characteristics and titration (ALS)	KQ3	Ventilator settings should be adjusted for optimal patient comfort and improvement of symptoms. ABGs and/or nocturnal oximetry and/or polysomnography are not required, but may be helpful in some circumstances. When bilevel pressure ventilators are used for NIV, a backup rate is recommended.
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Respiratory services	KQ4	Ability to generate PCF of at least 160 L/min is necessary for non-invasive management of pulmonary secretions. Baseline assisted PCF <270 L/min are likely to decrease to <160 L/min during chest infections, increasing the likelihood of pneumonia and respiratory failure. Patients with a baseline PCF < 270 L/min should have access to equipment which can provide insufflation and a mechanical cough in-exsufflation. Training of insufflation should commence when VC < 2L or 50% predicted. As manual assisted coughing techniques (e.g. abdominal thrust) further enhance PCF, they should be incorporated with insufflation or mechanical in-exsufflation techniques, where possible. For patients with VC < 1 to 1.5L, insufflations should precede manual assisted coughing techniques (e.g. abdominal thrusts). In adults, mechanical in-exsufflation settings of +40 cmH2O and – 40 cmH2O appear to safely provide adequate PCF for the majority of patients with neuromuscular disease. Mechanical in-exsufflation can be ineffective in patients with very poor bulbar dysfunction with insufflation capacity >1L, where dynamic airway collapse occurs. Techniques of insufflation, manual assisted coughing and mechanical in-exsufflation require substantial acclimatisation and should be trained when the patient is well and ideally prior to an acute infective requirement.
German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010[75]	Respiratory services	KQ4	A reduced cough impulse (peak cough flow; PCF < 270 l/min) can lead to acute decompensations and increased incidence of aspiration pneumonia. Measures to eliminate secretions should therefore be taken when SaO2< 95%, or a 2–3% drop in the patient's individual best value occurs. Step-based secretion management consists of measures to increase intrapulmonary volume via air stacking, frog breathing or manual hyperinflation, as well as assisted coughing techniques or mechanical cough assistants (CoughAssist ®, Pegaso Cough®) The measurement of coughing capacity in NMD patients is obligatory. Coughing weakness (PCF < 270 l/min) indicates the need for the initiation of secretion management.

Organization	Topic	KQ	Statement
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Respiratory services	KQ4	In patients with neuromuscular disease (NMD), mechanical insufflation and exsufflation should be used, in addition to standard physiotherapy techniques, when cough is ineffective and there is sputum retention.
American Academy of Neurology, Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: multidisciplinary care, symptom management, and cognitive/behavioral impairment (an evidence-based review), 2009[81]	Respiratory services	KQ4	Mechanical insufflation/exsufflation) may be considered to clear secretions in patients with ALS who have reduced peak cough flow, particularly during an acute chest infection. There are insufficient data to support or refute high frequency chest wall oscillation for clearing airway secretions in patients with ALS.
United Kingdom National Institute for Health and Care Excellence (NICE), Motor Neuron Disease: Assessment and Management, 2016[80]	Respiratory services	KQ4	Offer cough augmentation techniques such as manual assisted cough to people with NMD who cannot cough effectively. Consider unassisted breath stacking and/or manual assisted cough as the first-line treatment for people with NMD who have an ineffective cough For patients with bulbar dysfunction, or whose cough is ineffective with unassisted breath stacking, consider assisted breath stacking (for example, using a lung volume recruitment bag). Consider a mechanical cough assist device if assisted breath stacking is not effective, and/or during a respiratory tract infection. Consider opioids as an option to relieve symptoms of breathlessness. Take into account the route of administration and acquisition cost of medicines. Consider benzodiazepines to manage breathlessness that is exacerbated by anxiety. Take into account the route of administration and acquisition cost of medicines.
Canadian Thoracic Society 2011[72]	Respiratory services (ALS)	KQ4	Lung volume recruitment maneuvers should be introduced with declining VC. Methods to assist secretion clearance should be initiated when PCF is <4.25 L/s or the Norris bulbar core is <29.

Organization	Topic	KQ	Statement
European Federation of Neurological Societies (EFNS) Guidelines on the Clinical Management of Amyotrophic Lateral Sclerosis, 2012[83]	Respiratory services (ALS)	KQ4	Active management of secretions and provision of cough-assist devices can increase the effectiveness of assisted ventilation in ALS. For bronchial secretions: A mucolytic including N-acetylcysteine, 200–400 mg three times daily, may be beneficial. Beta-receptor antagonists and a nebulizer with saline and/or an anticholinergic bronchodilator and/or a mucolytic and/or furosemide may be used in combination. Mucolytics should only be used if sufficient cough flow is present. The patient and carer should be taught the technique of assisting expiratory movements using a manual-assisted cough (can also be performed by a physical therapist). The use of a mechanical insufflator–exsufflator may be helpful, particularly in the setting of an acute respiratory infection. A portable home suction device and a room humidifier may be of use. The medical treatment of intermittent dyspnoea should involve: a for short dyspnoeic bouts: relieve anxiety and give lorazepam 0.5–2.5 mg sublingually; b for longer phases of dyspnoea (>30 minutes): give morphine 2.5 mg orally or s.c. For the medical treatment of chronic dyspnoea, start with morphine 2.5 mg orally four to six times daily. For severe dyspnoea, give morphine s.c. or as an i.v. infusion. Start with 0.5 mg/h and titrate. If needed, add midazolam (2.5–5 mg) or diazepam for nocturnal symptom control and to relieve anxiety.
American Thoracic Society, Respiratory Care of the Patient with Duchenne Muscular Dystrophy, 2004[82]	Respiratory services (Duchenne Muscular Dystrophy)	KQ4	Patients with DMD should be taught strategies to improve airway clearance and how to employ those techniques early and aggressively. Use assisted cough technologies in patients whose clinical history suggests difficulty in airway clearance, or whose peak cough flow is less than 270 L/minute and/or whose maximal expiratory pressures are less than 60 cm H2O. The committee strongly supports use of mechanical insufflation-exsufflation in patients with DMD and also recommends further studies of this modality. Home pulse oximetry is useful to monitor the effectiveness of airway clearance during respiratory illnesses and to identify patients with DMD needing hospitalization Individuals who require mechanically assisted airway clearance therapy or mechanically assisted ventilation should see a pulmonologist every 3 to 6 months or as indicated for routine follow-up.
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Respiratory services (Duchenne Muscular Dystrophy)	KQ4	Lung volume recruitment maneuvers should be introduced with declining VC. -Methods to assist secretion clearance should be initiated when PCF <270 L/min.
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Respiratory services (All NMD except ALS and Duchenne Muscular Dystrophy)	KQ4	Assess airway clearance ability with peak cough flows and implement cough-assistance strategies

Organization	Topic	KQ	Statement
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Respiratory services (Spinal cord injury)	KQ4	Regular airway clearance techniques (lung volume recruitment, manually assisted coughing, and mechanical in-exsufflation), clinical assessment and ongoing monitoring of pulmonary function is recommended to ensure adequate airway clearance.

ABG: arterial blood gases, AHRF: acute hypercapnic respiratory failure, ALS: amyotrophic lateral sclerosis, cmH2O: centimeters of water (pressure), CO2: carbon dioxide, CPAP: continuous positive airway pressure, CRF: chronic respiratory failure, CWD: chest wall deformity, DMD: Duchenne muscular dystrophy, ETCO2: end tidal carbon dioxide, FEV1: Forced expiratory volume in one second, FVC: Forced vital capacity, HMV: home mechanical ventilation, IMV: invasive mechanical ventilation, kPa: kilopascal, KQ: key question, MEP: maximal expiratory pressure, mg: milligram, MIP: maximal inspiratory pressure, mmHg: millimeters of mercury (pressure), NIPPV/NPPV: non-invasive positive pressure ventilation, NIV: non-invasive ventilation, NMD: neuromuscular disease, OSA: obstructive sleep apnea, PaO2: partial pressure of arterial oxygen, PCO2/PaCO2: partial pressure of arterial carbon dioxide, PCF: peak cough flow, PEG: Polyethylene glycol, PImax: Maximal inspiratory mouth pressures, QOL: quality of life, RR: respiratory rate, s.c: subcutaneous, SCI: spinal cord injury, SNP/SNIP: sniff nasal inspiratory pressure, SpO2: Blood oxygen saturation level, tCO2/PTcCO2: transcutaneous carbon dioxide, TIPPV: tracheostomy intermittent positive pressure ventilation, VC: vital capacity

Table G.4. Guidelines for Thoracic Restrictive Disorders

Organization	Topic	KQ	Statement
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Device initiation criteria	KQ1	Planned elective domiciliary NIV is preferable to crisis management in NMD and chest wall disorders. This reduces the risk of acute presentation and provides a proven alternative to invasive mechanical ventilation which risks prolonged or permanent tracheostomy ventilation. NIV should almost always be trialled in the acutely unwell patients with NMD or CWD with hypercapnia. Do not wait for acidosis to develop. In patients with NMD or CWD, NIV should be considered in acute illness when vital capacity (VC) is known to be <1 L and RR >20, even if normocapnic. In patients with NMD or CWD, nocturnal NIV should usually be continued following an episode of AHRF, pending discussion with a home ventilation service.
Clinical Indications for Noninvasive Positive Pressure Ventilation in Chronic Respiratory Failure due to Restrictive Lung Disease, COPD, and Nocturnal Hypoventilation—A Consensus Conference Report, American Academy of Home Care Physicians, American College of Chest Physicians, American College of Physicians, American Sleep Disorders Association, American Thoracic Society, National Association for Medical Direction of Respiratory Care, 1999[74]	Device initiation criteria	KQ1	Indications for usage Symptoms (such as fatigue, dyspnea, morning headache, etc.) and one of the following Physiologic criteria (one of the following PaCO2 ≥ 45 mm Hg Nocturnal oximetry demonstrating oxygen saturation ≤ 88% for 5 consecutive minutes For progressive neuromuscular disease, maximal inspiratory pressures < 60 cmH2O or FVC <50% predicted
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device initiation criteria	KQ1	Patients with kyphoscoliosis should undergo periodical spirometry testing, and if FVC is <50%, ongoing review, assessing for evidence of hypercapnic respiratory failure should be instituted. Long-term nocturnal NIV should be offered to all patients with kyphoscoliosis who have developed chronic hypercapnic respiratory failure. Patients with hypoxemia but without hypercapnia may be managed cautiously with oxygen therapy alone while monitoring for development of hypercapnia.
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device initiation criteria	KQ1	NIV in patients with respiratory insufficiency from chest wall disease provides greater physiological and symptomatic relief over oxygen alone. NIV should be trialled in all patients with chest wall disorders with evidence of nocturnal hypoventilation.

Organization	Topic	KQ	Statement
German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010[75]	Device initiation criteria	KQ1	The following indication criteria are valid when symptoms of CRF and a reduced quality of life are present (at least 1 criterion must be fulfilled): Chronic daytime hypercapnia with $PaCO_2 \geq 45$mmHg Nocturnal hypercapnia with $PaCO_2 \geq 50$mmHg Daytime normocapnia with a rise in $PTcCO_2$ of ≥ 10mmHg during the night Patients without manifest hypercapnia but with severe, restrictive ventilatory dysfunction (VC < 50% predicted), must undergo a short-term (within 3 months) clinical control examination including polygraphy. NIV is the primary treatment option for HMV of restrictive thoracic disease patients with CRF. The most important criteria for the advent of long-term NIV are hypercapnia in combination with the typical symptoms of ventilatory insufficiency, and the reduction in quality of life. For symptoms of hypoventilation in the absence of hypercapnia, a somnological examination should take place. Patients with severe, restrictive ventilatory dysfunction in the absence of manifest hypercapnia must be closely monitored.
British Thoracic Society, Guidelines for Home Oxygen Use in Adults, 2015[73]	Device initiation, monitoring, and candidate selection	KQ1	Non-invasive ventilation (NIV) should be the treatment of choice for patients with NMD or chest wall disease causing type 2 respiratory failure.
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Device initiation, monitoring, and candidate selection	KQ1	In patients with NMD or CWD, senior/experienced input is needed in care planning and is essential if differences in opinion exist or develop between medical staff and patient representatives. In patients with NMD, it should be anticipated that bulbar dysfunction and communication difficulties, if present, will make NIV delivery difficult, and may make it impossible. Discussion about NIV and IMV, and patients' wishes with respect to cardiopulmonary resuscitation, should occur as part of routine care of patients with NMD or CWD. In patients with NMD or chest wall diseases, senior staff should be involved in decision-making, in conjunction with home mechanical ventilation specialists, if experience is limited, and especially when the appropriateness of invasive mechanical ventilation is questioned. Domiciliary NIV is effective in treating chronic hypercapnia, improves long-term survival and preserves a good or acceptable QoL

Organization	Topic	KQ	Statement
Clinical Indications for Noninvasive Positive Pressure Ventilation in Chronic Respiratory Failure due to Restrictive Lung Disease, COPD, and Nocturnal Hypoventilation—A Consensus Conference Report, American Academy of Home Care Physicians, American College of Chest Physicians, American College of Physicians, American Sleep Disorders Association, American Thoracic Society, National Association for Medical Direction of Respiratory Care, 1999[74]	Device initiation, monitoring, and candidate selection	KQ1	Disease documentation Before considering a restrictive thoracic patient for NIPPV, a physician with skills and experience in NIPPV must establish and document an appropriate diagnosis on the basis of history, physical examination, and diagnostic tests and assure optimal treatment of other underlying disorders (such as performing a multichannel sleep study to detect associated sleep apnea if clinically indicated) The most common disorders would include sequelae of polio, spinal cord injury, neuropathies, myopathies and dystrophies, ALS, chest wall deformities, and kyphoscoliosis.
British Thoracic Society, Guidelines for Home Oxygen Use in Adults, 2015[73]	Device initiation, monitoring, and candidate selection	KQ1	NIV should be the treatment of choice for patients with chest wall or neuromuscular disease causing type 2 respiratory failure. Additional LTOT (long term oxygen therapy) may be required in case of hypoxaemia not corrected with NIV.
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Device characteristics and titration	KQ3	In patients with NMD or CWD, consider controlled ventilation as triggering may be ineffective.
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device characteristics and titration	KQ3	Both pressure and volume preset ventilation is likely to be equally effective in chest wall disease, but there is a subset of patients which may demonstrate the need for volume ventilation if adequately titrated pressure preset fails to significantly improve diurnal hypercapnia.
German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010[75]	Device characteristics and titration	KQ3	NIV in pressure- and volume-limited modes is feasible. With set pressure, maximal ventilation pressure often reaches 20–25 mbar. Changeover from set pressure to set volume should be taken into account in order to improve ventilation. EPAP is generally not necessary if bronchial obstructions are absent.

Organization	Topic	KQ	Statement
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Respiratory services	KQ4	In patients with neuromuscular disease (NMD), mechanical insufflation and exsufflation should be used, in addition to standard physiotherapy techniques, when cough is ineffective and there is sputum retention.
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Respiratory services	KQ4	Methods to assist secretion clearance should be initiated when peak cough flow is <270 L/min

AHRF: acute hypercapnic respiratory failure, ALS: amyotrophic lateral sclerosis, cmH2O: centimeters of water (pressure), CRF: chronic respiratory failure, CWD: chest wall deformity, EPAP: expiratory positive airway pressure, FVC: Forced vital capacity, IMV: invasive mechanical ventilation, KQ: key question, LTOT: long term oxygen therapy, mbar: megabar (pressure), mmHg: millimeters of mercury (pressure), NIPPV/NPPV: non-invasive positive pressure ventilation, NIV: non-invasive ventilation, NMD: neuromuscular disease, PCO2/PaCO2: partial pressure of arterial carbon dioxide, QOL: quality of life, RR: respiratory rate, tCO2/PTcCO2: transcutaneous carbon dioxide pressure, VC: vital capacity

Table G.5. Guidelines for Obesity Hypoventilation Syndrome

Organization	Topic	KQ	Statement
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Device initiation criteria	KQ1	In patients with OHS, NIV should be started in acute hypercapnic respiratory failure using the same criteria as in AECOPD (pH<7.35 and pCO2 >6.5 kPa persist or develop despite optimal medical therapy). Many patients with acute hypercapnic respiratory failure secondary to OHS will require long-term domiciliary support (CPAP or NIV). Following an episode of acute hypercapnic respiratory failure referral to a home ventilation service is recommended. Patients with OSA, OHS or overlap syndrome should not have nocturnal oxygen therapy alone ordered. It can be considered in patients with evidence of established ventilatory failure, where it should be given with NIV support
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device initiation criteria	KQ1	NIV is the treatment of choice for OHS. In patients with OHS who have a minor degree of nocturnal desaturation and no nocturnal rise in PaCO2, CPAP is a reasonable initial therapy provided that follow-up is arranged within one to three months to evaluate response to therapy. Polysomnography is useful for titrating and confirming efficacy of bilevel pressures. Under circumstances when access to more than one device (bilevel PAP or CPAP) is limited, bilevel therapy is recommended. In patients with OHS who experience significant nocturnal desaturation or a nocturnal increase in PaCO2, bilevel PAP remains the therapy of choice.
Clinical Indications for Noninvasive Positive Pressure Ventilation in Chronic Respiratory Failure due to Restrictive Lung Disease, COPD, and Nocturnal Hypoventilation—A Consensus Conference Report, American Academy of Home Care Physicians, American College of Chest Physicians, American College of Physicians, American Sleep Disorders Association, American Thoracic Society, National Association for Medical Direction of Respiratory Care, 1999[74]	Device initiation criteria	KQ1	Before considering NIPPV for a patient with nocturnal hypoventilation from causes other than COPD or neuromuscular disease (criteria as outlined in part 1 and 2), a physician with demonstrated skills and experience in NIPPV must establish and document an appropriate diagnosis from this category on the basis of history and physical examination. A polysomnogram (PSG) is required for diagnosis of sleep apnea. A CPAP trial is recommended if OSA is documented unless a previous CPAP trial was unsuccessful or there is significant hypoventilation that is believed to be unlikely to respond to CPAP alone. Indications for usage of NIPPV PSG criteria for OSA not responsive to CPAP PSG criteria for mixed sleep apnea not responsive to CPAP Central sleep apnea Other forms of nocturnal hypoventilation
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device initiation criteria	KQ1	Indications for NIV in OHS include an awake PaCO2 >45mmHg and failure of CPAP therapy as evidence by either sustained oxygen desaturation during sleep or an increase in nocturnal daytime or nocturnal CO2 >8mmHg.
German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for	Device initiation criteria	KQ1	Due to the high prevalence of an accompanying obstructive sleep apnea syndrome (90% of cases), primary sleep diagnostics by means of polysomnography are necessary. The indication of NIV for patients with symptomatic CRF under adequate CPAP therapy yields to the following situations: A ≥ 5 minute-long increase in nocturnal PTcCO2 > 55mmHg and in PaCO2 ≥ 10 mmHg, respectively, in

Organization	Topic	KQ	Statement
Treatment of Chronic Respiratory Failure, 2010[75]			comparison to the awake state or Desaturations < 80% SaO2 over ≥ 10 minutes In the case of severe hypercapnia or symptomatic, severe co-morbidity, primary NIV can be implemented according to the physician's assessment. If the first control visit (including poly(somno)graphy under CPAP therapy) reveals no improvement in the characteristic symptoms of chronic hypoventilation or the absence of daytime normocapnia ("non-responder"), transfer of the patient to NIV is indicated. CPAP or NIV are the primary treatment options for HMV of patients with OHS. An accompanying loss of weight should also be aimed for. An initial attempt at CPAP treatment under polysomnographical conditions should take place in patients without significant co-morbidities. In the presence of significant co-morbidities, however, primary NIV therapy can be indicated. Persistent hypoventilation under CPAP (≥ 5 minute-long increase in PTcCO2 > 55mmHg and PaCO2 ≥ 10 mmHg, respectively, in comparison to normocapnia during the awake state, or desaturation < 80% over ≥ 10 minutes) is an indication for NIV. Significant weight loss can enable a change from NIV to CPAP therapy, or even an attempt at resting the treatment.
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device initiation, monitoring, and candidate selection	KQ1	Simple spirometry, SpO2 and serum bicarbonate should be performed in all patients referred for SDB assessment when BMI is greater than 35kg/m2. Arterial blood gases should be obtained in those individuals where SpO2 is ≤ 92% or where the serum bicarbonate is >27mmol/L to confirm the presence and severity of hypoventilation. Thyroid function should also be assessed and any airflow limitation treated appropriately. Positive airway pressure is first line therapy in patients with OHS, although adjunctive oxygen therapy is likely to be required, at least initially, for a significant number of patients. Autotitrating and home studies are not appropriate for this patient group. A full PSG should be performed during manual titration in order to identify the nature of the sleep disordered breathing and response to CPAP pressure. Many individuals will respond to initial intervention with CPAP. Titration should commence in CPAP mode to document the patient's response to abolition of upper airway obstruction alone.
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device continuation, compliance, and outcomes	KQ1 and KQ2	Individuals initially using bilevel support should be reviewed again after 3 months on therapy and CPAP retried, since a significant number may be switched to CPAP without clinical deterioration. In patients placed on CPAP in whom awake PaCO2 at baseline was 45-55mmHg, a clinical review at one month with repeat blood gases should be performed. Bilevel support should be used as initial therapy in patients presenting with acute decompensated respiratory failure. After 3 months, a CPAP titration should be undertaken to determine long term therapy. The need for and type of nocturnal PAP therapy should be reassessed if significant weight loss occurs.
German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010[75]	Device characteristics and titration	KQ3	Titration of CPAP pressure until hypoventilation is eliminated For NIV therapy, increase EPAP until obstructions are eliminated accompanied by titration of inspiratory pressure. In the case of considerable weight loss, a repeated attempt at CPAP, a change from NIV to CPAP, or a rest in treatment are all possible under poly(somno)graphical control. Weight loss should be part of the long-term treatment plan.

Organization	Topic	KQ	Statement
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device characteristics and titration (Central hypoventilation syndrome)	KQ3	CHS patients who require only nocturnal ventilator support may be managed by NIV with a backup rate or diaphragmatic pacing. Severe CHS, mainly seen in congenital CHS, requires continuous invasive ventilator support, but daytime diaphragmatic pacing can markedly improve mobility and, as the child matures, NIV may suffice.
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Device characteristics and titration	KQ3	High inspiratory positive airway pressure (IPAP) and expiratory positive airway pressure (EPAP) settings are commonly required in patients with OHS (e.g., IPAP>30, EPAP>8). Volume control (or volume assured) modes of providing NIV may be more effective when high inflation pressures are required.

AECOPD: acute exacerbation of chronic obstructive pulmonary disease, BMI: body mass index, CHS: central hypoventilation syndrome, CO2: carbon dioxide, COPD: chronic obstructive pulmonary disease, CPAP: continuous positive airway pressure, CRF: chronic respiratory failure, EPAP: expiratory positive airway pressure, HMV: home mechanical ventilation, IPAP: inspiratory positive airway pressure, kPa: kilopascal, KQ: key question, mmHg: millimeters of mercury (pressure), mmol: millimole, NIPPV/NPPV: non-invasive positive pressure ventilation, NIV: non-invasive ventilation, OHS: Obesity hypoventilation syndrome, OSA: obstructive sleep apnea, PAP: positive airway pressure, PCO2/PaCO2: partial pressure of arterial carbon dioxide, pH: potential of hydrogen, PSG: polysomnogram, SaO2: arterial blood oxygen saturation, SDB: sleep disordered breathing, SpO2: Blood oxygen saturation level, tCO2/PTcCO2: transcutaneous carbon dioxide

Table G.6. Guidelines for Other Respiratory Diseases

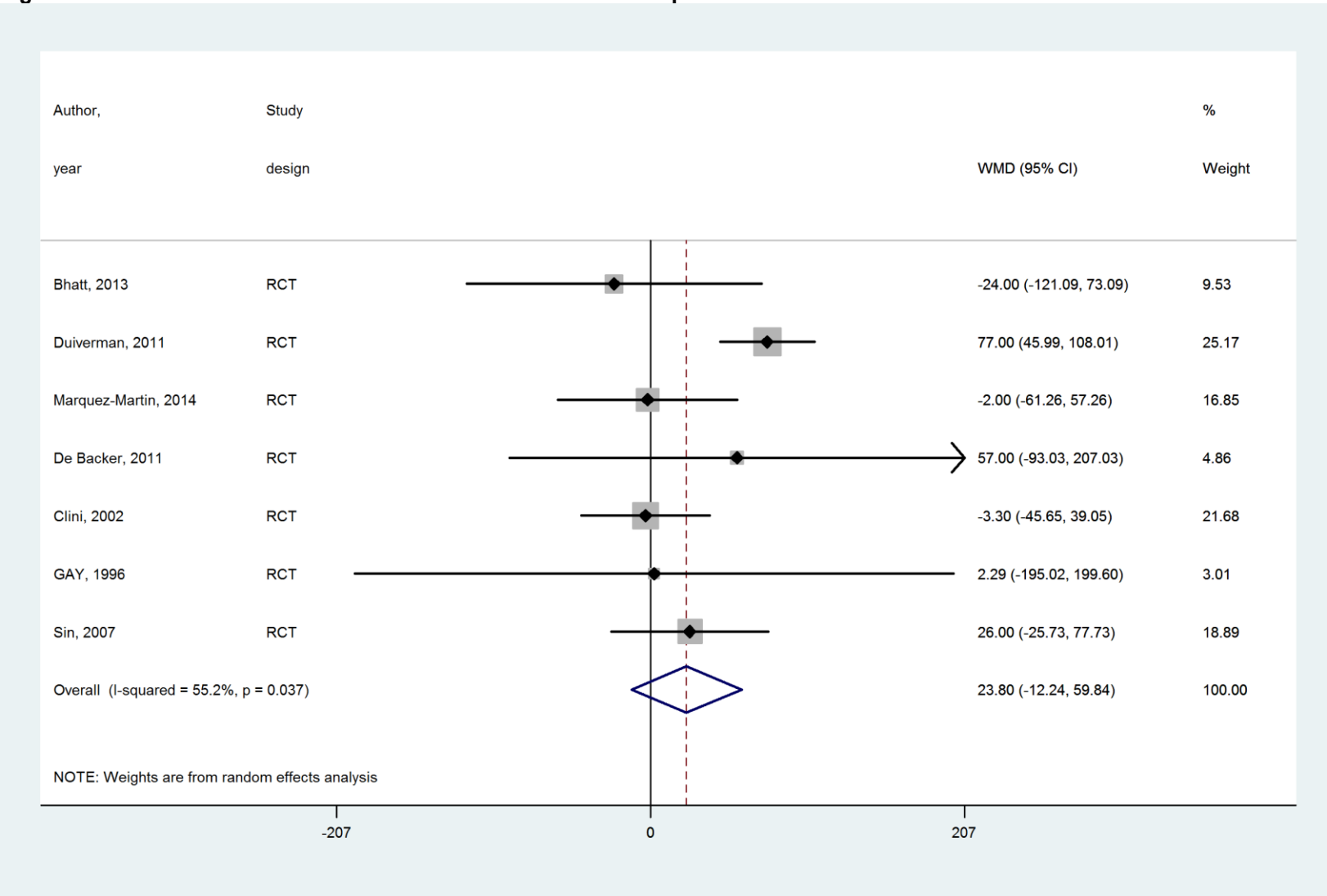
Organization	Topic	KQ	Statement
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Device initiation, monitoring, and candidate selection (Asthma)	KQ1	Acute (or acute on chronic) episodes of hypercapnia may complicate chronic asthma. This condition closely resembles COPD and should be managed as such.
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Device initiation, monitoring, and candidate selection (Bronchiectasis)	KQ1	In patients with non-CF bronchiectasis, NIV should be started in acute hypercapnic respiratory failure using the same criteria as in AECOPD (pH<7.35 and pCO2 >6.5 kPa persist or develop despite optimal medical therapy).
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Device initiation, monitoring, and candidate selection (Cystic fibrosis)	KQ1	In patients with cystic fibrosis, NIV is the treatment of choice when ventilatory support is needed.
British Thoracic Society, Guidelines for Home Oxygen Use in Adults, 2015[73]	Device initiation, monitoring, and candidate selection (Cystic fibrosis)	KQ1	Nocturnal oxygen therapy should not be given to CF patients with nocturnal hypoxaemia alone who do not fulfil LTOT criteria. It can be considered in patients with evidence of established ventilator failure, where it should be given with NIV support.
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device initiation, monitoring, and candidate selection (Cystic fibrosis)	KQ1	Individuals with awake SpO2<94% or spirometry (FEV1<65% predicted) are at risk of nocturnal oxygen desaturation. Overnight oximetry should be undertaken in individuals meeting these criteria. Non-invasive ventilation is indicated if daytime CO2>45mmHg and nocturnal gas exchange shows SpO2<90% for >5% of TST and/or a rise in TcCO2 / ETCO2 from NREM to REM >5mmHg during room air breathing occurs. Nocturnal NIV is more effective than oxygen therapy in controlling nocturnal hypoventilation in patients with hypercapnic CF lung disease. Bilevel ventilation should be trialled initially. Volume ventilation may offer additional benefits in some individuals especially if work of breathing is high. NIV does not appear to increase the incidence of pneumothorax, but this is a relatively common occurrence in this population. Therefore, patients need to be educated regarding the symptoms of pneumothorax and should seek immediate medical attention should these symptoms arise. Changes in awake blood gases are not the best measure of the effectiveness of NIV in CF. Changes in symptoms, exertional dyspnoea and exercise tolerance, and control of nocturnal hypoventilation are better indicators of the patient's response to therapy. NIV may be used in patients unsuitable for transplant to relieve symptoms and improve sleep quality. However, alternative methods of symptom relief need to be introduced at the appropriate time.
Agency for Clinical Innovation,	Device	KQ1	Awake PaCO2 > 45 mmHg in the absence of lung and chest wall abnormalities, skeletal malformations and

Organization	Topic	KQ	Statement
Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	initiation, monitoring, and candidate selection (Hypercapnic central sleep apnea)		neuromuscular disorders, in combination with symptoms consistent with sleep disordered breathing warrant a full polysomnogram. In patients with isolated sleep hypoventilation, titrate NIV settings in a spontaneous-timed mode, during a full polysomnogram. Where hypercapnic central apnoea is caused from pharmacological intake (e.g. opioid based derivatives), referrals to chronic pain team or relevant prescribing body should be made with the aim of reducing medication intake in order to improve central events and stabilise oxygen saturations. Overall patient management should be performed by specialised teams. Any signs of chest infection should be reviewed and managed promptly, especially in the case of CCHS where a lack of dyspnoea in response to pneumonia may mask severe respiratory compromise.
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Respiratory services (Cystic fibrosis)	KQ4	In patients with cystic fibrosis, specialised physiotherapy is needed to aid sputum clearance.

CCHS: congenital central hypoventilation syndrome, CF: cystic fibrosis, CO2: carbon dioxide, COPD: chronic obstructive pulmonary disease, ETCO2: end tidal carbon dioxide, FEV1: Forced expiratory volume in one second, kPa: kilopascal, KQ: key question, LTOT: long term oxygen therapy, mmHg: millimeters of mercury (pressure), NIV: non-invasive ventilation, NREM: non-rapid eye movement, PCO2/PaCO2: partial pressure of arterial carbon dioxide, pH: potential of hydrogen, REM: rapid eye movement, SpO2: Blood oxygen saturation level, tCO2/TcCO2: transcutaneous carbon dioxide pressure

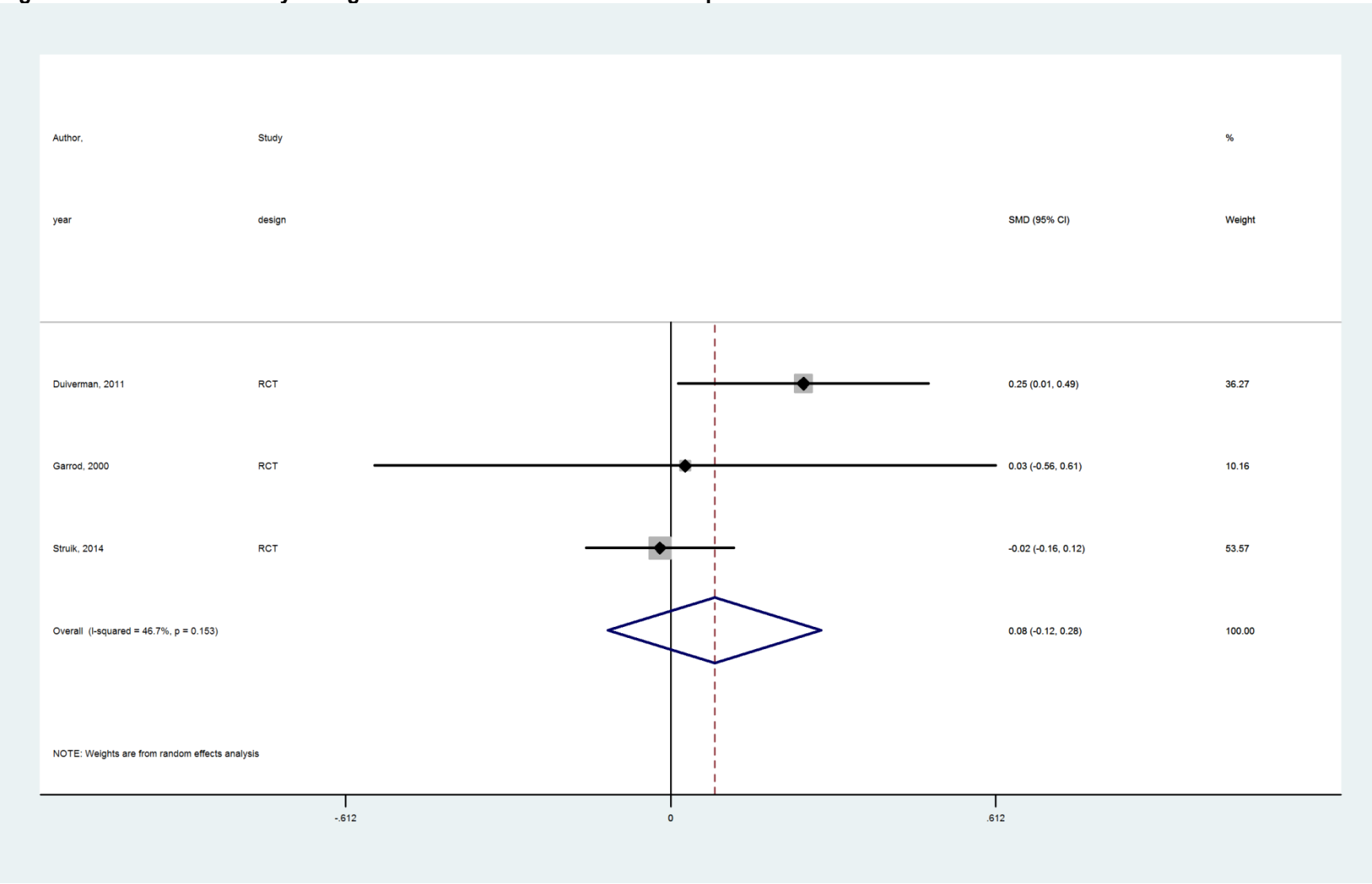
Appendix H. Figures

Figure H.1. 6 Minute Walk Test-BPAP versus No Device in COPD patients



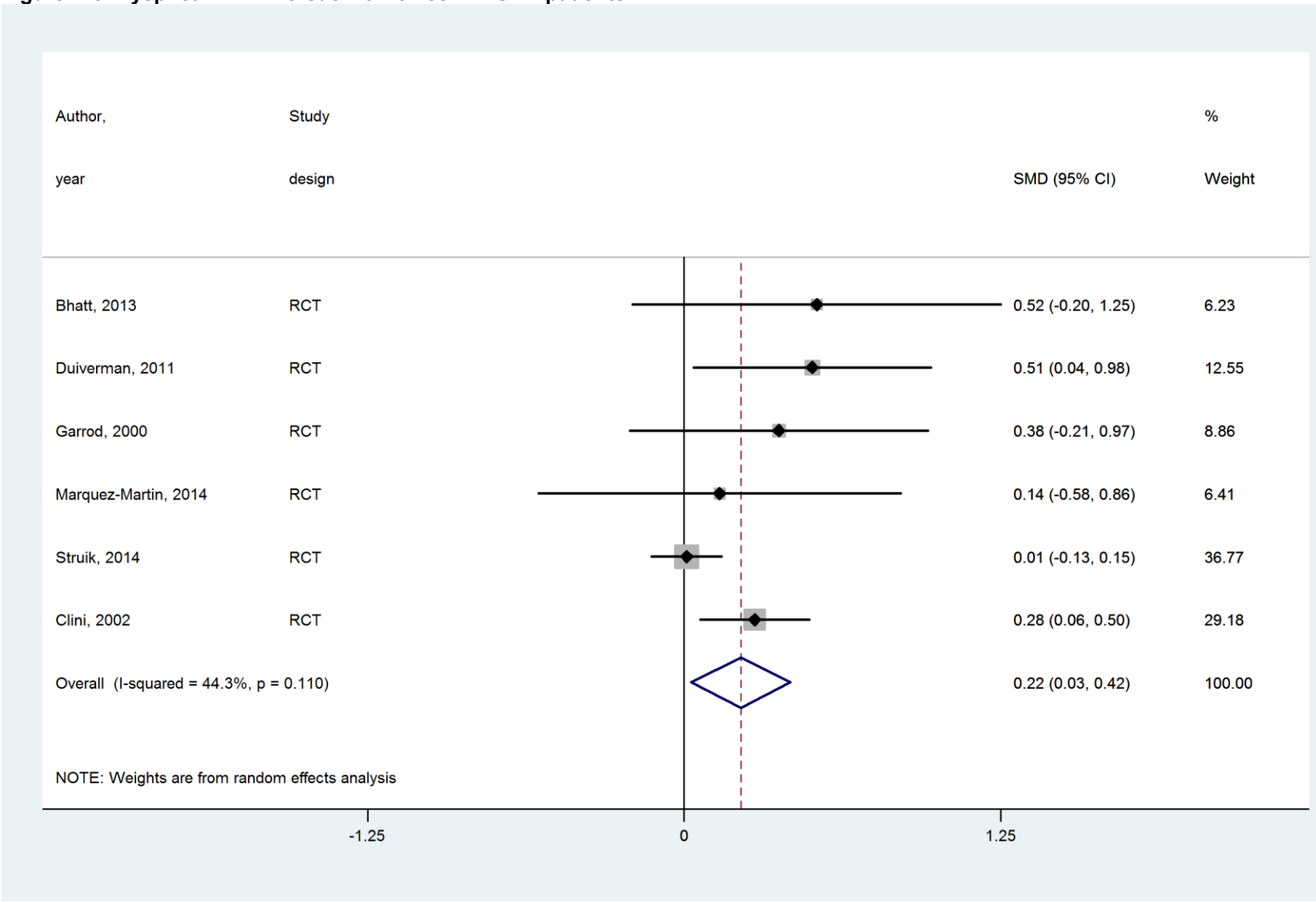
CI: Confidence interval; RCT: Randomized controlled trial; WMD: Weighted mean difference

Figure H.2. Activities of Daily Living-BPAP versus No Device in COPD patients



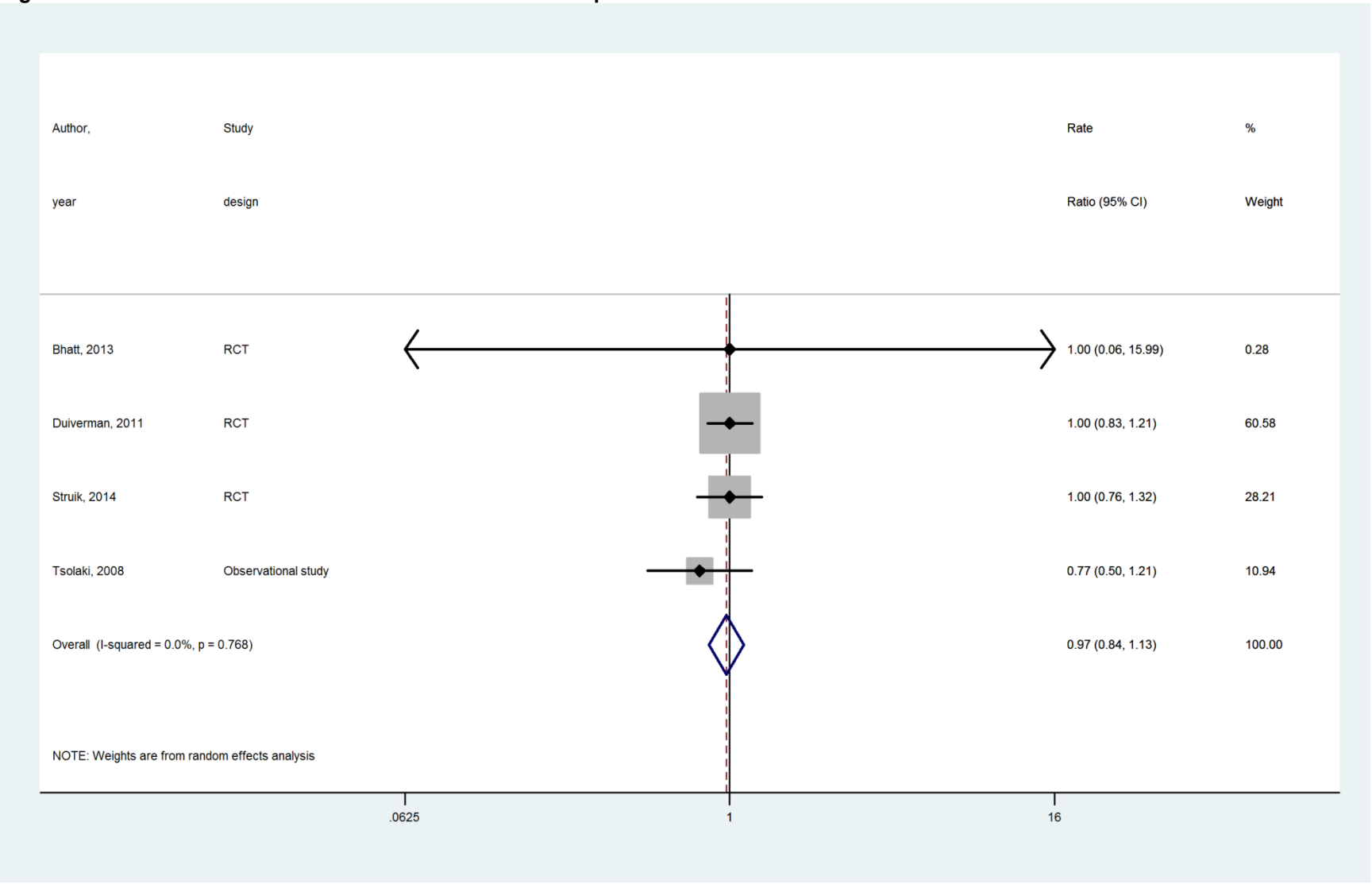
CI: Confidence interval; RCT: Randomized controlled trial; SMD: Standardized mean difference

Figure H.3. Dyspnea-BPAP versus No Device in COPD patients



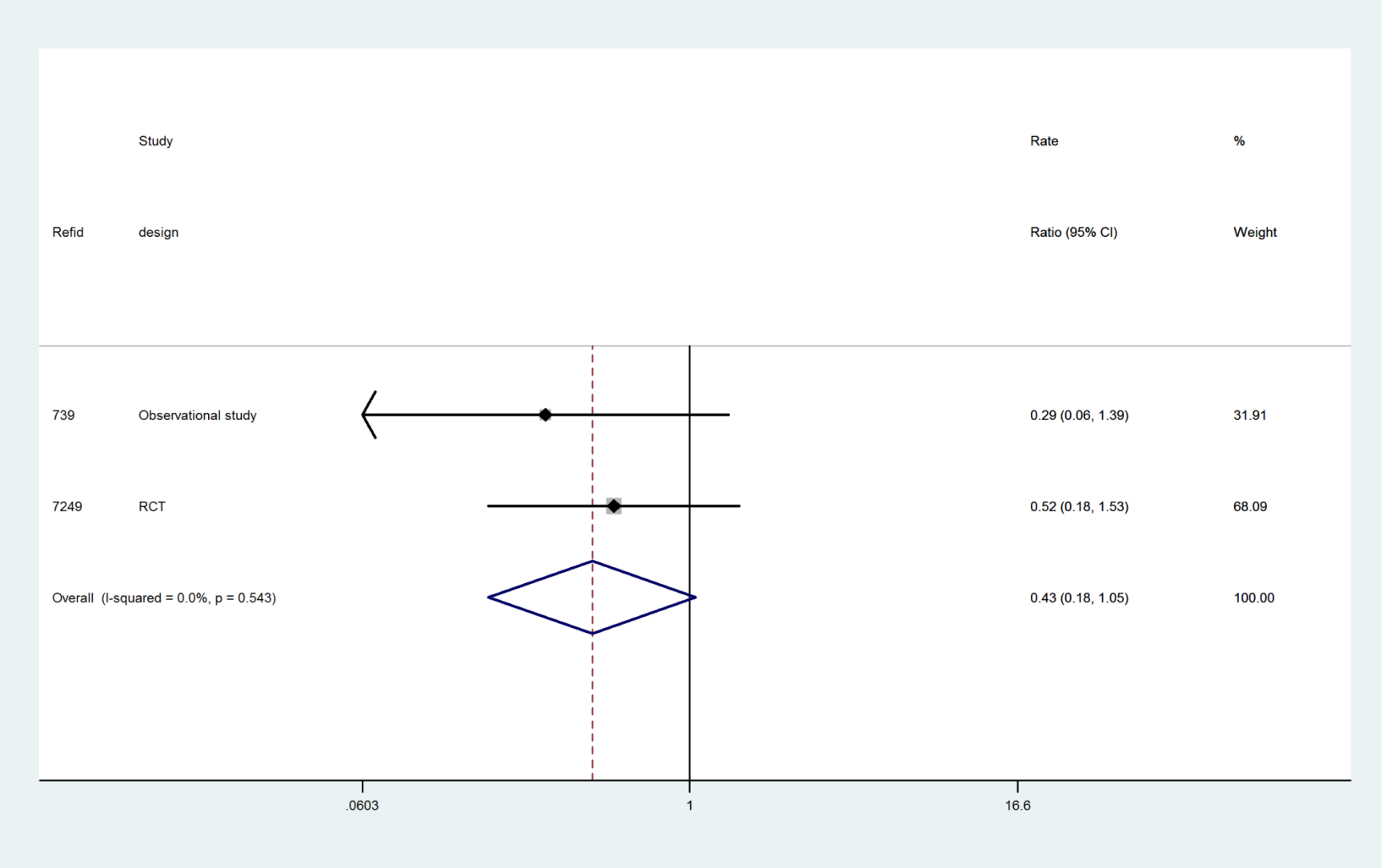
CI: Confidence interval; RCT: Randomized controlled trial; SMD: Standardized mean difference

Figure H.4. Exacerbation-BPAP versus No Device in COPD patients



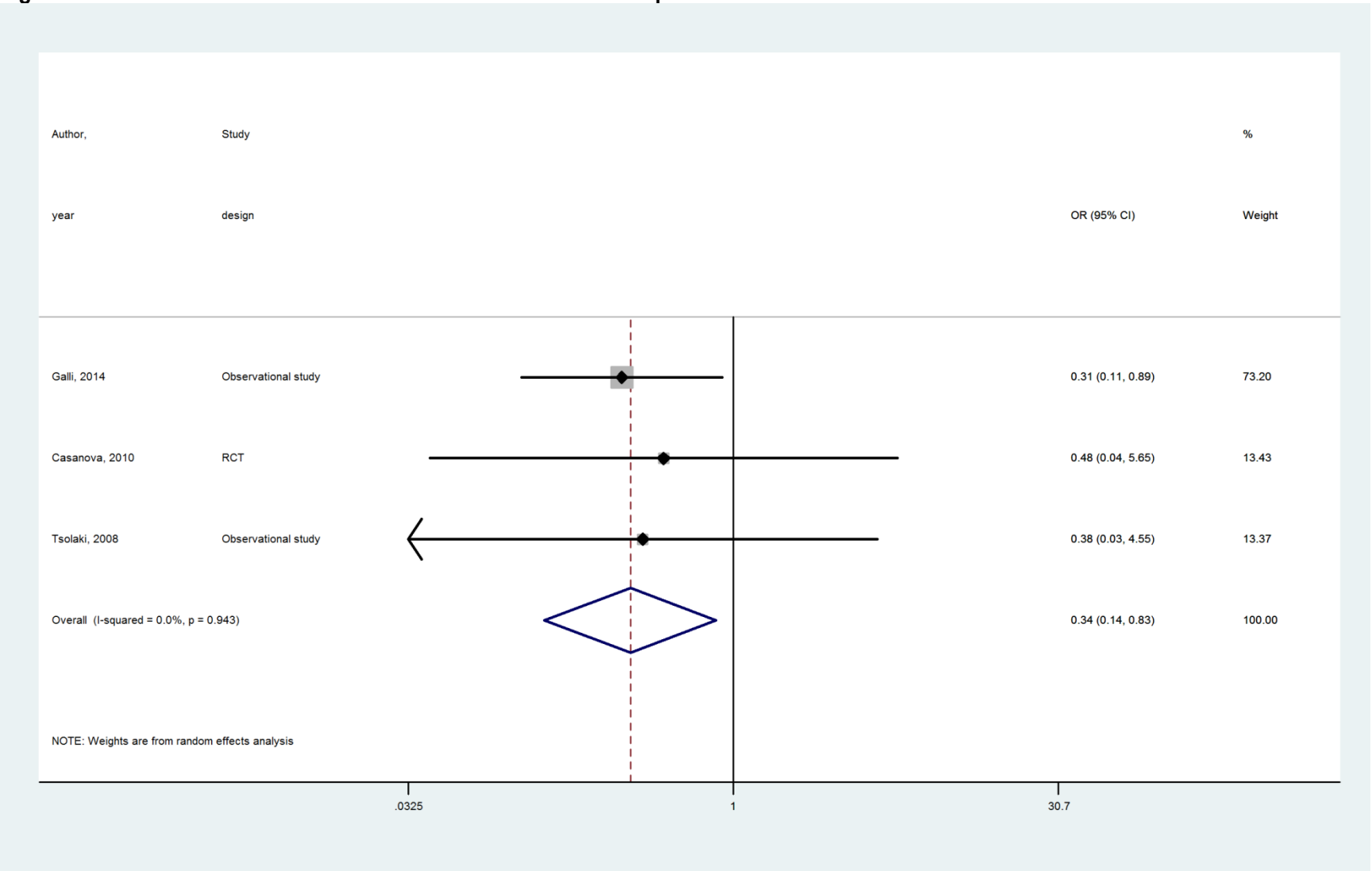
CI: Confidence interval; RCT: Randomized controlled trial

Figure H.5. ICU admissions-BPAP versus No Device in COPD patients



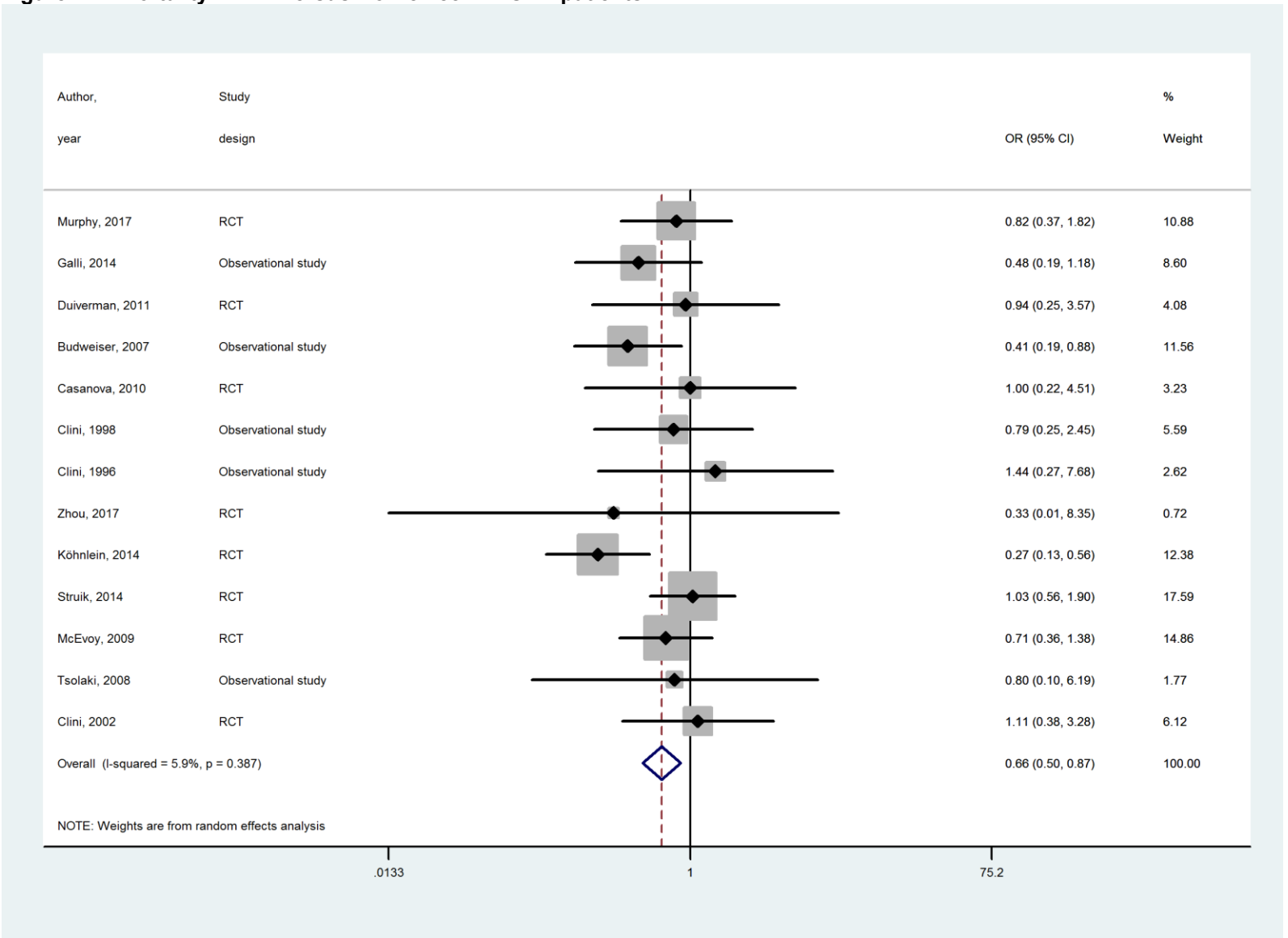
CI: Confidence interval; RCT: Randomized controlled trial

Figure H.6. Need for Intubation-BPAP versus No Device in COPD patients



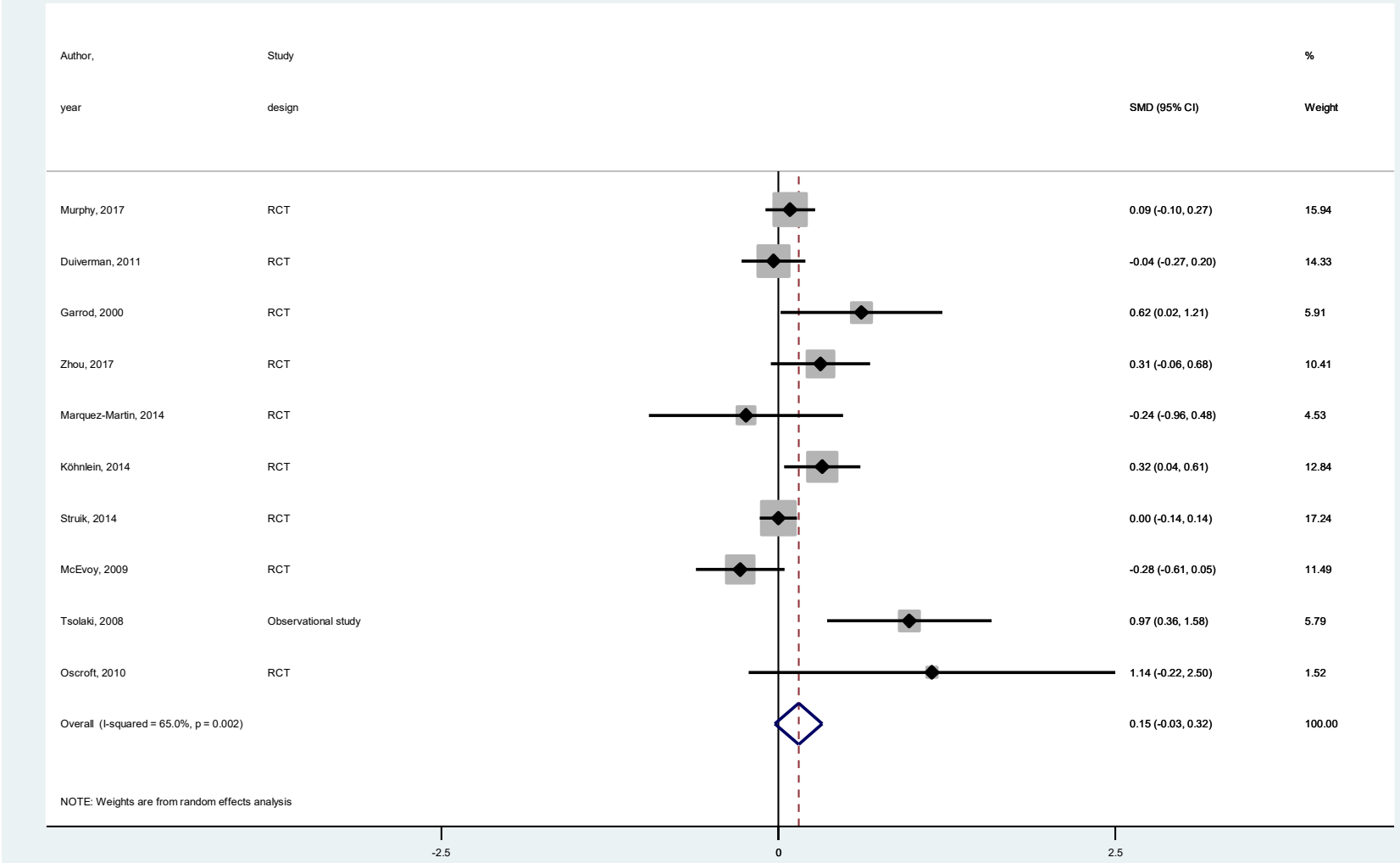
CI: Confidence interval; OR: Odds Ratio RCT: Randomized controlled trial

Figure H.7. Mortality-BPAP versus No Device in COPD patients



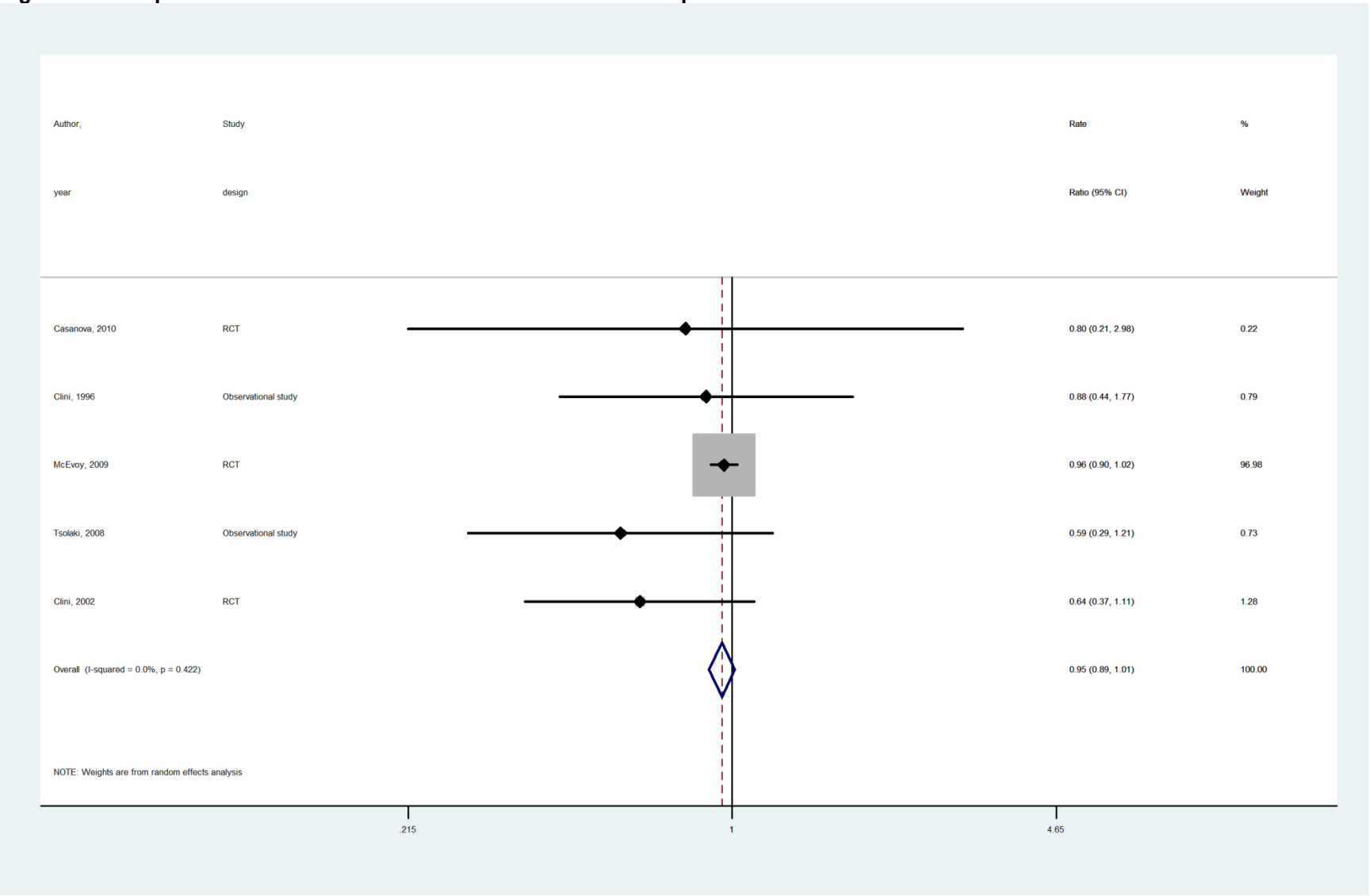
CI: Confidence interval; OR: Odds Ratio RCT: Randomized controlled trial

Figure H.8. Quality of Life-BPAP versus No Device in COPD patients



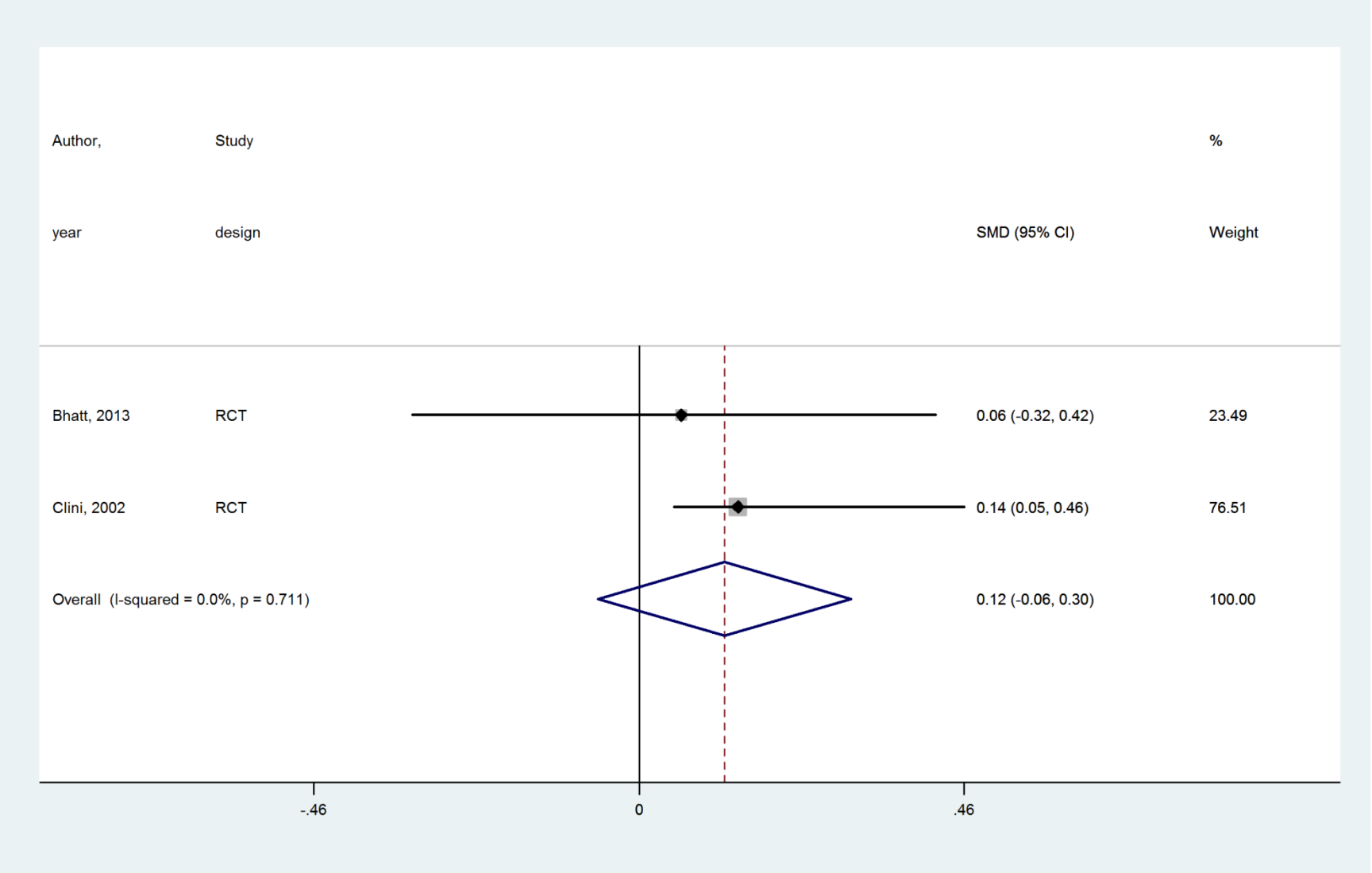
CI: Confidence interval; RCT: Randomized controlled trial; SMD: Standardized mean difference

Figure H.9. Hospital Readmission-BPAP versus No Device in COPD patients



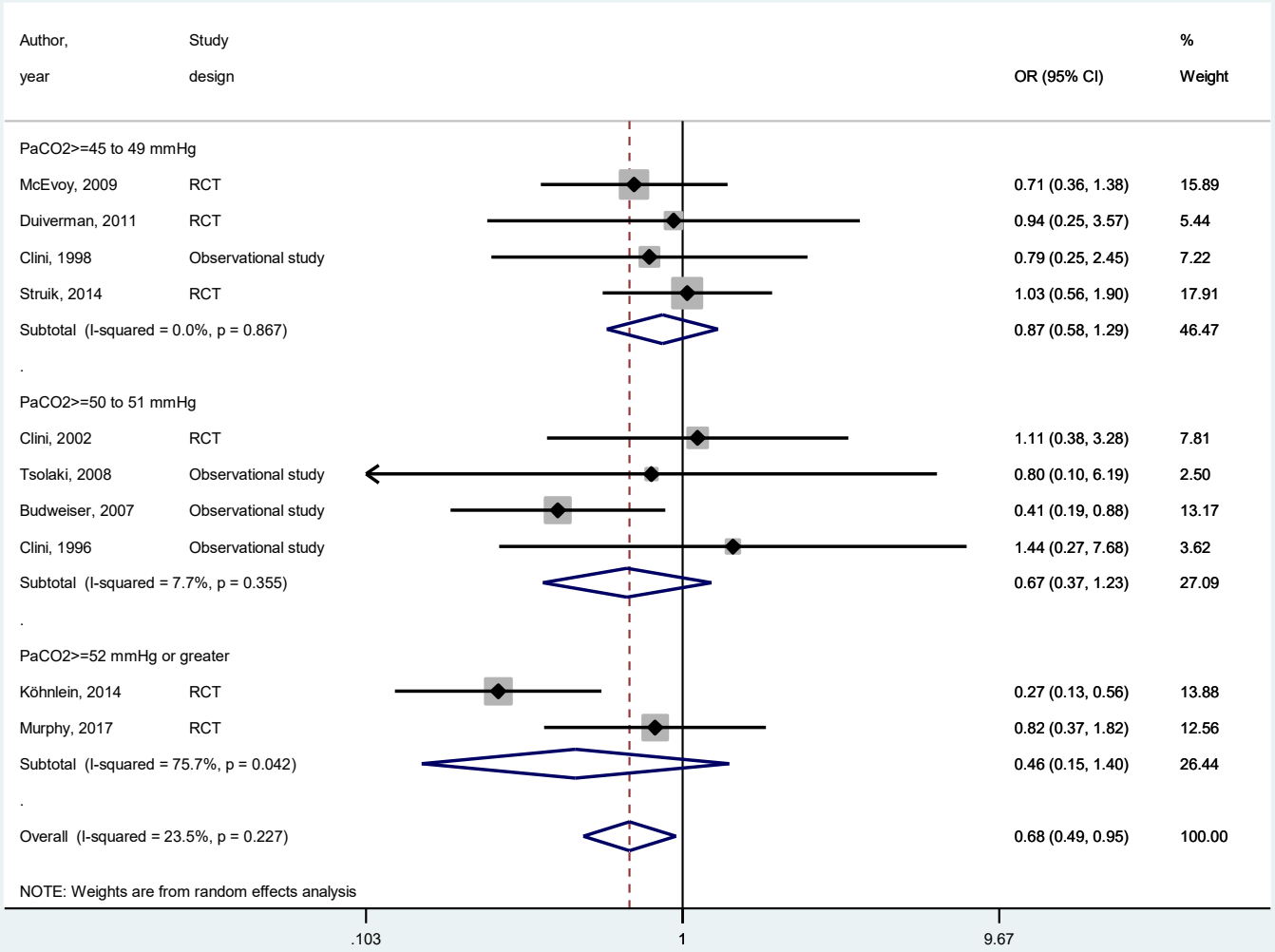
CI: Confidence interval; RCT: Randomized controlled trial

Figure H.10. Sleep Quality-BPAP versus No Device in COPD patients



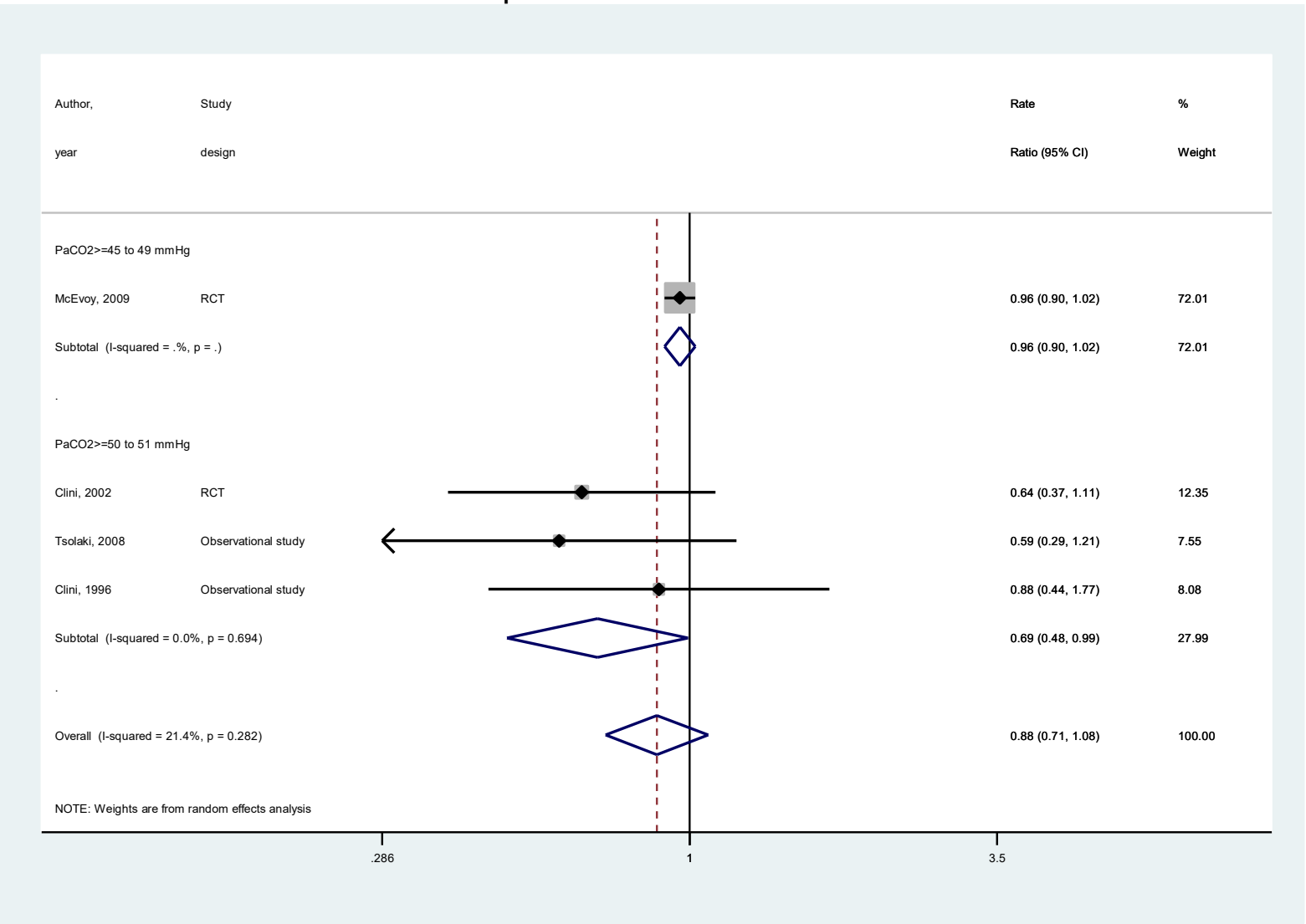
CI: Confidence interval; RCT: Randomized controlled trial; SMD: Standardized mean difference

Figure H.11. Subgroup Analysis: Level of Hypercapnia (PaCO2) used as an Initiation Criterion for Initiation of NIPPV on Mortality-BPAP



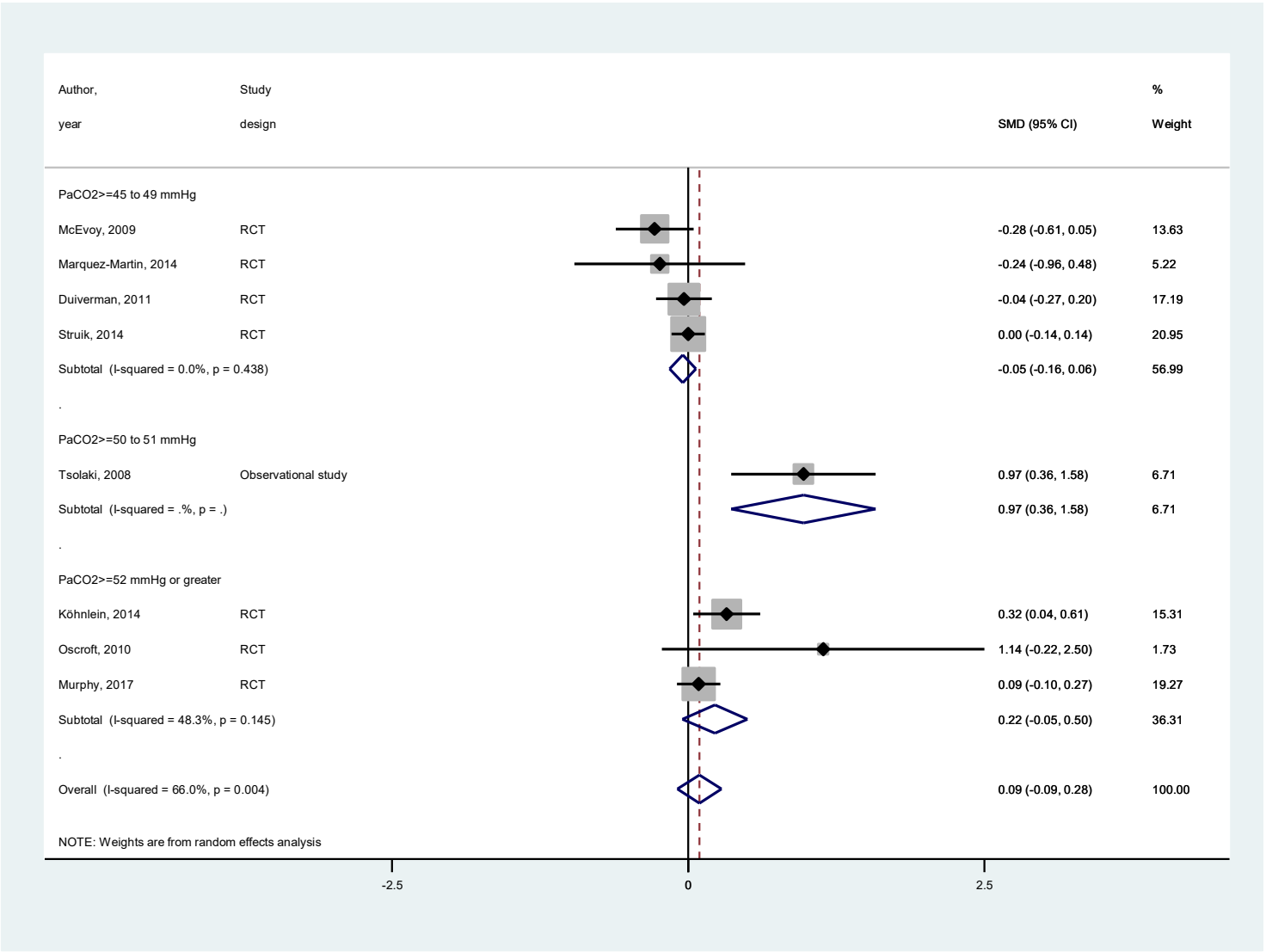
CI: Confidence interval; OR: Odds Ratio RCT: Randomized controlled trial

Figure H.12. Subgroup Analysis: Level of Hypercapnia (PaCO2) used as an Initiation Criterion for Initiation of NIPPV on Hospital Readmission-BPAP versus No Device in COPD patients



CI: Confidence interval; OR: Odds Ratio RCT: Randomized controlled trial

Figure H.13. Subgroup Analysis: Level of Hypercapnia (PaCO2) used as an Initiation Criterion for Initiation of NIPPV on Quality of Life-BPAP versus No Device in COPD Patients



CI: Confidence interval; RCT: Randomized controlled trial; SMD: Standardized mean difference

Appendix I. Post-hoc Subgroup Analysis

Post-hoc subgroup analysis of PaCO2 levels for starting NIPPV in patients with COPD

Background

In patients with chronic obstructive pulmonary disease (COPD), there is variability regarding the level of hypercapnia (PaCO2) that is considered as a prerequisite for initiation of noninvasive positive pressure ventilation (NIPPV). This variability exists in clinical practice, guideline recommendations, and patient enrollment criteria for comparative effectiveness studies.

For example, guidelines included in this review used the following criteria to consider initiation of NIPPV in patients with COPD: stable daytime PaCO2 >50mmHg, ≥ 50mmHg, and >55mmHg. Some guidelines used lower thresholds (i.e. PaCO2 46-50mmHg or PaCO2 50-54mmHg) when other characteristics were present (such as nocturnal hypercapnia, nocturnal hypoxia, recurrent exacerbations, or severe exacerbations requiring ventilatory support). Other guidelines did not specify which PaCO2 levels constituted "hypercapnia." In clinical practice, for example, the United States Centers for Medicare and Medicaid Services (CMS), uses a PaCO2 ≥ 52mmHg for initiation of BPAP for patients with COPD.[84] In this systematic review, we identified eleven studies (most of which were published in the past 10 years) that used PaCO2 levels of >45mmHg as inclusion criteria for initiating NIPPV, somewhat lower than was considered in many guidelines and clinical practices.

To evaluate the impact of PaCO2 initiation threshold on clinical outcomes, we searched for, but ultimately found no included studies which directly assessed this association. Based on reviewers' comments, we performed a post-hoc subgroup analysis of individual included studies to indirectly assess if higher PaCO2 thresholds to initiate NIPPV were associated with larger effect sizes for the 4 primary clinical outcomes (mortality, need for intubation, quality of life and all-cause hospital admissions).

Methods

We included studies which enrolled patients with COPD, reported one of the 4 primary outcomes (mortality, need for intubation, quality of life, and all-cause hospital admissions), and reported a daytime stable PaCO2 threshold for initiation of NIPPV. We excluded studies that did not report a PaCO2 threshold or that reported a PaCO2 threshold during an episode of acute respiratory failure. To evaluate if there was a dose response (higher cutoffs associated with increasingly better outcomes), and in the setting of PaCO2 ≥ 52mmHg threshold commonly used in the United States, we defined the PaCO2 threshold categories as: 1) PaCO2 ≥45 to 49 mmHg, 2) PaCO2 ≥50 to 51 mmHg, and 3) PaCO2 ≥52 mmHg or greater. The other methods and analysis were identical to the methods used in the main report.

Results

The post-hoc subgroup analysis was only possible for studies comparing BPAP use with no device use. When compared BPAP use to no device use in patients with COPD, 16 studies,[8, 10, 14, 15, 23, 29, 46, 70, 85 16, 30, 38, 43, 49, 64, 66] 11 RCTs, and 6 observational studies reported at least one of the 4 primary outcomes (mortality, need for intubation, quality of life and all-cause hospital admissions). We excluded 4[10, 29 30, 70]of these 16 studies from the subgroup analyses as two studies[30, 70] did not report PaCO2 threshold cutoffs and two studies[10, 29] measured PaCO2 cutoff during episodes of acute respiratory failure. Twelve studies were included in the subgroup analyses. The risk of bias of these 12 studies was rated as moderate to high similar to those in the main analysis. Findings are presented in Figures 1-3. These findings suggested that higher PaCO2 levels may be associated with improved quality of life compared to lower levels (PaCO2 ≥52 mmHg: SMD 0.22; 95% CI: -0.05 to 0.50 vs. PaCO2 ≥50 to 51: 0.97; 95% CI: 0.36, 1.58 vs. PaCO2 ≥45 to 49: -0.05; 95% CI: -0.16 to 0.06). The effect size for quality of life for cutoff PaCO2 ≥50 to 51 mmHg was also higher than the overall effect size (SMD: 0.97; 95% CI: 0.36 to 1.58 vs. SMD: 0.15, 95% CI: -0.03 to 0.32); however, this was driven by a single nonrandomized study. Differences in mortality and hospital readmissions favored higher initiation criteria but were not statistically different. There were no other significant difference between the subgroups and overall pooled effect sizes.

Conclusions

No included studies directly evaluated the association between clinical outcomes with different levels of hypercapnia as a criterion to initiate NIPPV in patients with COPD. In this post-hoc subgroup analysis of RCTs and observational studies, which compared BPAP use to no device use, there were no differences in mortality or all-cause hospital admissions based on PaCO2 threshold initiation criteria. There was a statistically significant larger improvement in quality of life with higher PaCO2 threshold initiation criteria (PaCO2≥50 to 51compared to PaCO2 ≥45 to 49). These findings suffer a high risk of bias and do not warrant high strength of evidence.

Appendix J. References for Appendixes

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