Appendixes

Contents

Appendixes ..................................................................................................................................... 0
Appendix A. Flow Chart ............................................................................................................. A-1
Appendix B. Search Strategy ...................................................................................................... B-1
Appendix C. Excluded Studies ................................................................................................... C-1
Appendix D. Characteristics of Included Studies ....................................................................... D-1
Appendix E. Risk of Bias ............................................................................................................. E-1
Appendix F. Results from the Included Studies .......................................................................... F-1
Appendix G. Guidelines .............................................................................................................. G-1
Appendix H. Figures ................................................................................................................... H-1
Appendix I. References for Appendixes ....................................................................................... I-1

Tables
Table D.1. Characteristics of included studies ............................................................................ D-1
Table E.1. Risk of bias for RCTs (Cochrane ROB tool) for included studies ............................. E-1
Table E.2. Risk of bias for observational studies (Newcastle-Ottawa Quality Assessment Scale) for included studies ……………………………………………………………………………………. E-2
Table F.1. COPD - New initiation of home device ........................................................................ F-1
Table F.2. COPD - Established home device use ........................................................................ F-6
Table F.3. Thoracic Restrictive Disorders - New initiation of home device .............................. F-7
Table F.4. Thoracic Restrictive Disorders – Established home device use .............................. F-8
Table F.5. Neuromuscular Disease - New initiation of home device ........................................ F-9
Table F.6. Neuromuscular Disease – Established home device use ........................................ F-11
Table F.7. Obesity Hypoventilation Syndrome - New initiation of home device .................... F-12
Table F.8. Other Respiratory Diseases - New initiation of home device ................................ F-15
Table F.9. Other Respiratory Diseases – Established home device use .................................. F-15
Table F.10. Mixed diseases – New initiation of home device .................................................. F-15
Table F.11. Mixed diseases – Established home device use .................................................... F-17
Table F.12. COPD – Effectiveness of home devices ................................................................. F-18
Table F.13. Thoracic Restrictive Disorders – Effectiveness of home devices ........................ F-40
Table F.14. Neuromuscular Disorder – Effectiveness of home devices .................................... F-41
Table F.15. Obesity Hypoventilation Syndrome – Effectiveness of home devices ................. F-45
Table F.16. Other Respiratory Diseases – Effectiveness of home devices .............................. F-49
Table F.17. Mixed Diseases – Effectiveness of home devices .................................................. F-49
Table F.18. COPD – Equipment parameters .......................................................................... F-51
Table F.19. Thoracic Restrictive Disorders – Equipment parameters ...................................... F-57
Table F.20. Neuromuscular Disease – Equipment parameters ................................................. F-59
Table F.21. Obesity Hypoventilation Syndrome – Equipment parameters ............................ F-63
Table F.22. Other Respiratory Diseases – Equipment parameters .......................................... F-65
Table F.23. Mixed Diseases – Equipment parameters ............................................................. F-65
Table F.24. COPD – Respiratory services .............................................................................. F-68
Table F.25. Thoracic Restrictive Disorders – Respiratory services .......................................... F-69
Table F.26. Neuromuscular Disease – Respiratory services .................................................. F-69
Table F.27. Obesity Hypoventilation Syndrome – Respiratory services ................................... F-71
Table F.28. Mixed Diseases – Respiratory Services ................................................................. F-71
Table G.1. Guidelines for all conditions .................................................................................. G-1
Table G.2. Guidelines for COPD ............................................................................................. G-6
Table G.3. Guidelines for Neuromuscular Disease ................................................................. G-10
Table G.4. Guidelines for Thoracic Restrictive Disorders ..................................................... G-23
Table G.5. Guidelines for Obesity Hypoventilation Syndrome ............................................... G-27
Table G.6. Guidelines for Other Respiratory Diseases ......................................................... G-30
Table H.1. Strength of evidence for COPD studies ................................................................. H-1
Table H.2. Strength of evidence for Neuromuscular Disease studies ..................................... H-2
Table H.3. Strength of evidence for Obesity Hypoventilation Syndrome studies .................. H-2

Figures
Figure A.1. Flow Chart ........................................................................................................... A-1
Figure H.1. 6 Minute Walk Test-BPAP versus No Device in COPD patients .......................... H-1
Figure H.2. Activities of Daily Living -BPAP versus No Device in COPD patients ............. H-2
Figure H.3. Dyspnea-BPAP versus No Device in COPD patients ....................................... H-3
Figure H.4. Exacerbation-BPAP versus No Device in COPD patients ................................. H-4
Figure H.5. ICU admissions-BPAP versus No Device in COPD patients .............................. H-5
Figure H.6. Need for Intubation-BPAP versus No Device in COPD patients ....................... H-6
Figure H.7. Mortality-BPAP versus No Device in COPD patients ........................................ H-7
Figure H.8. Quality of Life-BPAP versus No Device in COPD patients ............................... H-8
Figure H.9. Hospital Readmission-BPAP versus No Device in COPD patients .................... H-9
Figure H.10. Sleep Quality-BPAP versus No Device in COPD patients ............................... H-10
Appendix A. Flow Chart

Figure A.1. Flow chart

Records identified through database searching  
N = 6,097

Additional records identified through grey literature, reference mining, and Stakeholders/Key Informants  
N = 63

Records screened  
N = 6,160

Title and abstract review excluded  
N = 5,092

Full-text articles excluded  
N = 972
- Abstract/conference proceeding: 249
- Duplicate study: 3
- Foreign language: 155
- Irrelevant comparison or no comparison: 59
- Irrelevant intervention or time frame: 114
- Irrelevant outcome: 31
- Irrelevant patient (age or medication condition): 119
- Irrelevant setting/location: 22
- Irrelevant study design: 115
- Outcomes not reported by device intervention: 19
- Published before 1995: 54
- Unclear device: 15
- Clinical trials: 17

Relevant ongoing clinical trials  
N = 8

Relevant systematic review/meta-analysis N=12

Full-text articles assessed for eligibility N =1,068

Guidelines included in the review  
N = 13

Original studies included in the review  
N = 61 (63 articles)
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/
   (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.

2  1 or 2

3  1 or 2

4  *Amyotrophic Lateral Sclerosis/

5  *Bronchiectasis/

6  *Cystic Fibrosis/

7  *Hypercarnia/

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/
   ("Amyotrophic lateral sclerosis" or "Atrophic Muscular Disorder*" or Bronchiectasis or
   "Chronic Obstructive Pulmonary Disease*" or "congenital structural myopathy*" or "Cystic
   Fibrosis" or hypercapnia or hypoventilation or "Interstitial Lung Disease*" or kyphosis or
30 "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 myopathy*").ti.

31 or/4-30
32 3 and 31
   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]
   limit 33 to (adult <18 to 64 years> or aged <65+ years>) [Limit not valid in
34 CCTR,CDSR,Ovid MEDLINE(R),Ovid MEDLINE(R) Daily Update,Ovid MEDLINE(R) In-
   Process,Ovid MEDLINE(R) Publisher; records were retained]
   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]
   limit 35 to (embryo or infant or child or preschool child <1 to 6 years> or school child <7 to
36 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/
    ((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")
73 adj3 (study or survey or analysis or design)) or retrospective* or "prospective study" or
"prospective survey" or "prospective analysis" or prospective* 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 analysis*)) or "case control study" or "case base 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
   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 structural myopathy*" 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 myopathy*"")

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 (pragmatic clinical trial) or (random* W/1 allocat*) or (double* W/1 blind*) or (double* 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 retrospective* or "prospective study" or "prospective survey" or "prospective analysis" or prospectiv* or (population W/3 (study* or survey* or research)) or "incidence study" or "incidence survey" or "incidence analysis" or (("follow-up" or followup) W/1 (study* or survey or analysis) or (observation or observational) W/1 (study or survey or analysis)) or (clinical* W/1 trial) or "clinical trial" or "clinical trial" or "evaluation study" or "evaluation survey" or "evaluation analysis" or "quantitative study" or "quantitative analysis*" or "numeric study" or "validation study" or "validation survey" or "validation analysis" or "validation study" or "validation survey" or "validation analysis" or "quasi experimental study" or "quasi experimental analysis" or "quasieperimental 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 referent study" or "case referent 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


National Guidelines Clearinghouse

("Amyotrophic lateral sclerosis" or "Atrophic Muscular Disorder**" or Bronchiectasis or "Chronic Obstructive Pulmonary Disease**" or "congenital structural myopathy**" 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 Injury**" or "Spinal Muscular Atrophy" or "structural congenital myopathy**") 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**")

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 structural myopathy**" 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 Injury**" or "Spinal Muscular Atrophy" or "structural congenital myopathy**") 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/
   (((domestic or home or domiciliary) adj3 (residence or residences or setting or settings or care or nurses* or help or service* or treatment* or therapy* or "respiratory care" or "respiratory treatment*" or "respiratory therapy*" or "respiratory service*" or "respiratory assist*" or ventilator*)) or "assisted living" or homecare).ti,ab,hw,kw.
2 "nursing home*".ti,ab,hw,kw.
3 (1 or 2) not 3
4 exp Respiration, Artificial/
   (((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 Tracheostomy* or ventilation or ventilator*).ti,ab,hw,kw.
5 5 or 6
6 4 and 7
   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]
7 5 or 6
8 4 and 7
   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]
9 8 and 11
   limit 10 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]
10 11 not 9
11 12 not 10
12 8 not 13
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/

((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 "replication survey" 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 referent study" or "case referent study" or "case referent study" or "case compare peer 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 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 nur* 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 (randomised W/3 trial) 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 retrospective* or "prospective study" or "prospective survey" or "prospective analysis" or prospectiv* or (population W/3 (stud* or study or survey or research)) or "concurrent study" or "concurrent survey" or "concurrent analysis" or "incidence study" or "incidence survey" or "incidence analysis" or ("random* W/1 allocat*) or ("random* W/1 mask") or ("random* W/1 blind") or (("random* W/1 mask") or ("random* W/1 blind") or ("random* W/1 allocat") or ("random* W/1 blind") or ("random* W/1 mask") or "random* 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


4. [Searching the literature for non-invasive positive pressure ventilation for neuromuscular diseases]. Revue des Maladies Respiratoires. 2006 Nov;23(5 Pt 4):1451-53. PMID: 17151546. [Foreign language study].


23. Alison Galbraith AfHR, Quality CHA, Harvard School of Public Health HPHC. Evaluating Sequential Strategies to Reduce Readmission in a Diverse Population. 2011 October. PMID: NCT01619098. [Irrelevant patient (age, medical condition)].


(NIV) treatment for COPD patients with a history of NIV-treated exacerbation; a randomized, controlled, multi-center study. BMC Pulmonary Medicine. 2016 Feb 12;16:32. PMID: 26867542. [Irrelevant outcome].


43. Armstrong A, Ross J, Pieri-Davies S. Development and implementation of standard carer training guidelines for the care of long term ventilated patients in the

44. Assistance Publique - Hôpitaux de Paris AFcIM. Efficacy and Tolerance of Early Launching of Nocturnal Non Invasive. 2010 October. PMID: NCT01225614. [Irrelevant patient (age, medical condition)].


46. Associazione Riabilitatori Insufficienza R. Effects of Home-based Pulmonary Rehabilitation in Patients With Severe or Very Severe Chronic Obstructive Pulmonary Disease (COPD). 2010 September. PMID: NCT01198288. [Irrelevant intervention (or time frame)].


PMID: 614770522. [Abstract/conference proceeding].


71. Bayside Health NH, Medical Research Council A, Monash University CFFA. Non-invasive Ventilation and Oxygen Therapy in Cystic Fibrosis Patients With Nocturnal Oxygen Desaturation. 2003 March. PMID: NCT00157183. [Completed Clinical Trial].


75. Belfast Health SCT, Queen's University B. pRotective vEntilation With Veno-venouS Lung assisT in Respiratory Failure. 2016 March. PMID: NCT02654327. [Irrelevant patient (age, medical condition)].


Outcomes not reported per device intervention.


93. Boehringer I. Multiple Dose Comparison of Tiotropium Inhalation Capsules, Salmeterol Inhalation Aerosol and Placebo in Patients With Chronic Obstructive Pulmonary Disease (COPD). 1999 February. PMID: NCT02172287. [Irrelevant intervention (or time frame)].


2010 Jul;55(7):885-94. PMID: 20587101. [Irrelevant intervention (or time frame)].


137. Cambridge University Hospitals NHSFT, University of C, King's College L. Trial of a Breathlessness Intervention Service for Intractable Breathlessness. 2008 August. PMID: NCT00678405. [Irrelevant patient (age, medical condition)].


140. Cano NJM, Roth H, Court-Ortune I, et al. Nutritional depletion in patients on long-


149. Cazzolli PA, Oppenheimer EA. Home mechanical ventilation for amyotrophic lateral sclerosis: nasal compared to tracheostomy-intermittent positive pressure ventilation. Journal of the Neurological Sciences. 1996 Aug;139
150. Centocor I. A Study of Safety and Efficacy of CNTO 148 in Patients With Severe Persistent Asthma. 2004 August. PMID: NCT00207740. [Irrelevant patient (age, medical condition)].

151. Centre d'Investigation Clinique et T. Concordance Between ETCO2, PTCO2 and PaCO2 in the Home-ventilated Neuromuscular Patient. 2014 April. PMID: NCT02068911. [Completed Clinical Trial].

152. Centre d'Investigation Clinique et T. Long-Term Effect of LIAM on Respiratory Performance in NIV Patients Suffering From Neuromuscular Disease. 2015 October. PMID: NCT02288299. [Irrelevant study design].

153. Centre d'Investigation Clinique et T. Natural History of Cardiac and Respiration Function in Patients With Muscular Dystrophies on Home Mechanical Ventilation. 2016 July. PMID: NCT02501083. [Completed Clinical Trial].

154. Centre Hospitalier Universitaire Saint P. Impact of Telemonitoring to Improve Adherence in Continuous Positive Airway Pressure (CPAP)-Treated Patients. 2016 April. PMID: NCT02773953. [Irrelevant patient (age, medical condition)].


163. Chang Gung Memorial H. Respiratory Muscle Exercise Training in COPD Patients. 2012 December. PMID: NCT01747694. [Irrelevant intervention (or time frame)].

164. Chang Gung Memorial H. The Impacts of Pulmonary Rehabilitation Therapy on Patients After Thoracic Surgery. 2016 April. PMID: NCT02757092. [Irrelevant patient (age, medical condition)].


174. Cherniack RM, Svanhill E. Long-term use of intermittent positive-pressure


196. Columbia U. Efficacy of Noninvasive Ventilation in Amyotrophic Lateral Sclerosis (ALS). 2007 May. PMID: NCT00537641. [Completed Clinical Trial].


1. PMID: 26323938. [Irrelevant patient (age, medical condition)].


232. Domicile IPS. Initiation of Non-Invasive Ventilation at Home Versus Hospital Among Patients With Overlap Syndrome. 2015 February. PMID: NCT02363413. [Completed Clinical Trial].


the additive effect of spinal surgery and home nocturnal ventilation in improving survival. Neuromuscular Disorders. 2007 Jun;17(6):470-5. PMID: 17490881. [Irrelevant patient (age, medical condition)].


256. Efficacy and Safety Study of Benralizumab in Patients With Uncontrolled Asthma on Medium to High Dose Inhaled Corticosteroid Plus LABA (MIRACLE). 2017 September 15. PMID: NCT03186209. [Irrelevant patient (age, medical condition)].


ATS. 2012;185(no pagination). PMID: 71987703. [Abstract/ conference proceeding].


287. Fisher PH. CPAP In-home Assessment Australia. 2016 July. PMID: NCT02809794. [Irrelevant patient (age, medical condition)].
288. Fisher PH. CPAP In-home Assessment NZ. 2016 July. PMID: NCT02804919. [Irrelevant patient (age, medical condition)].


291. Fondazione Don Carlo Gnocchi O, Fondazione Salvatore M. "New Perspectives of Adaptation to NIV in ALS". 2015 January. PMID: NCT02537132. [Completed Clinical Trial].


295. Frederiksberg University H, Foundation T, The TF, et al. The Virtual Hospital - a Clinical Trial. 2010 June. PMID: NCT01155856. [Irrelevant patient (age, medical condition)].


299. Fundació Institut de Recerca de l'Hospital de la Santa Creu i Sant P. Home Mechanical Ventilation Effectiveness and Air Leaks. 2006 June. PMID: NCT01090986. [Irrelevant study design].

300. Fundació Institut de Recerca de l'Hospital de la Santa Creu i Sant P. Home Mechanical Ventilation in Patients With Chronic Obstructive Pulmonary Disease (COPD) and Hypercapnic Response. 2005 October. PMID: NCT01120574. [Completed Clinical Trial].


311. Garuti G. Home rehabilitation and therapy. [Italian]. Rassegna di Patologia dell'Apparato Respiratorio. 2010


321. Girault C. Noninvasive ventilation and acute respiratory failure. [French]. Revue

323. Goldstein R. Evaluating the end-stage patient for appropriate oxygen delivery system and identifying appropriate candidates for home ventilation. Seminars in Respiratory and Critical Care Medicine. 1996;17(6):477-89. PMID: 27045149. [Irrelevant patient (age, medical condition)].


325. Golish J, Ioachimescu OC. Ambulatory titration of continuous positive airway pressure was as effective as polysomnography for obstructive sleep apnoea. Evidence-Based Medicine. 2007 October;12(5):148. PMID: 47611221. [Abstract/ conference proceeding].


332. Gonzalez Rodriguez CI, Jimenez Bermejo F, Rubio T, et al. [Non-invasive home mechanical ventilation in the COPD patient]. Anales del Sistema Sanitario de


353. Hannover Medical S. Long-Term Study On Home Spirometry After Lung Transplantation. 2000 January. PMID: NCT00743171. [Irrelevant patient (age, medical condition)].


365. Heral A, Stalenheim G, Boman G. Effects of positive expiratory pressure (PEP), continuous positive airway pressure (CPAP) and hyperventilation in COPD patients with chronic hypercapnea. Upsala Journal of Medical Sciences. 1995;100(3):223-32. PMID: 8808185. [Irrelevant intervention (or time frame)].


370. Highcock MP, Shneerson JM, Smith IE. Increased ventilation with NIIPPV does not necessarily improve exercise capacity in COPD. European Respiratory Journal. 2003 Jul;22(1):100-5. PMID: 12882458. [Irrelevant intervention (or time frame)].


376. Howard M, Berlowitz D, Batchelder I, et al. Day implementation model for non-


385. Icadom PR. Wearable Noninvasive Positive Pressure Ventilation Device in COPD. 2017 January. PMID: NCT03130361. [Irrelevant study design].


387. Imperial College L, The Health F. Assessment of the Effects of an Intermediate Care Package in Preventing Hospitalisation of Patients With COPD. 2003 December. PMID: NCT00129779. [Irrelevant intervention (or time frame)].


412. Kaohsiung Medical University Chung-Ho Memorial H. The Efficacy of Pulmonary Rehabilitation Exercise in Home Care for the Non-invasive Ventilator-dependent Elderly With COPD. 2016 July. PMID: NCT02836912. [Completed Clinical Trial].


419. Katholieke Universiteit Leuven QsU, Kingston O. Effects of Inspiratory Muscle Training on Dyspnea Perception During Exercise in Patients With COPD. 2013 July. PMID: NCT01900873. [Irrelevant intervention (or time frame)].


sclerosis (ALS) and their family members. [German]. Journal fur Anasthesie und Intensivbehandlung. 2002;9(1):34-5. PMID: 34875226. [Foreign language study].


7. PMID: 27025993. [Foreign language study].


PMID: 70275093. [Abstract/ conference proceeding].


443. Kohler D, Criere CP, Raschke F. [Guidelines for home oxygen and home ventilation therapy. German Society of Pneumology, German Society of Sleep Medicine, Working Group of Nocturnal Respiratory and Cardiovascular Disorders, Working Circle of Home and Long-Term


454. Krempf M, Barel P, Adoue D, et al. [Application of residual positive pressure during assisted ventilation at home using
pressure relaxator devices (author's transl)].
Revue Francaise des Maladies Respiratoires.
[Foreign language study].

455. Kruse N, Bulow HH. Noninvasive ventilation of patients with respiratory insufficiency: Six months experience.
[Foreign language study].

[Irrelevant intervention (or time frame)].

[Irrelevant study design].


[Irrelevant intervention (or time frame)].

[Irrelevant comparison or no comparison].


[Published before 1995].


466. Laier-Groeneveld G. [Home and Long-Term Ventilation Study Group: guidelines...


469. Lane A, Harlow S, Murray P. A local domiciliary non-invasive ventilation (NIV) service reduces length of hospital stay for patients unable to wean from NIV. Thorax. 2015 December;70:A175. PMID: 72199778. [Abstract/ conference proceeding].


prolongs survival in subjects with ALS. Amyotrophic Lateral Sclerosis. 2007 Jun;8(3):185-8. PMID: 17538782. [Outcomes not reported per device intervention].


509. Lyager S, Steffensen B, Juhl B. Indicators of need for mechanical ventilation in Duchenne muscular dystrophy and spinal muscular atrophy. Chest. 1995 Sep;108(3):779-85. PMID: 7656633. [Irrelevant comparison or no comparison].

511. Maastricht University Medical Center MMC, Erasmus Medical Center TNAF, Stichting Astma Bestrijding TN, et al. (Cost-)Effectiveness Interdisciplinary Community-Based COPD Management Program (INTERCOM). 2002 January. PMID: NCT00840892. [Irrelevant intervention (or time frame)].


514. Mahidol U. Randomized Cross-over TRD and CPAP for OSA. 2016 January. PMID: NCT02788487. [Irrelevant patient (age, medical condition)].


553. Michailidis V, Steiropoulos P, Perantoni E, et al. Validation of the Greek version of the severe respiratory insufficiency


563. Moizuddin M, Vujnic S, Janssen W, et al. Preliminary report on the impact of continuous positive airway pressure (CPAP) therapy on health-related quality of life in patients with obstructive sleep apnea and


583. Naestved Hospital UHR, Hvidovre University Hospital AU, Bispebjerg Hospital T, et al. Breathing Exercises in Asthma Targeting Dysfunctional Breathing. 2017 April 27. PMID: NCT03127059. [Irrelevant patient (age, medical condition)].


591. National Guideline C. Care of dying adults in the last days of life. 2015. [Irrelevant patient (age, medical condition)].

592. National Guideline C. Care of the hospitalized patient with acute exacerbation of COPD. 2016. [Irrelevant patient (age, medical condition)].

593. National Guideline C. Chronic pain disorder medical treatment guideline. 2017. [Irrelevant patient (age, medical condition)].


596. National Guideline C. Idiopathic pulmonary fibrosis. The diagnosis and management of suspected idiopathic pulmonary fibrosis. 2013. [Irrelevant intervention (or time frame)].

597. National Guideline C. Metastatic spinal cord compression. Diagnosis and management of adults at risk of and with
metastatic spinal cord compression. 2008. [Irrelevant patient (age, medical condition)].


599. National Guideline C. Palliative care for adults. 2013. [Irrelevant patient (age, medical condition)].

600. National Guideline C. Tuberculosis. 2016. [Irrelevant patient (age, medical condition)].


605. Network for Engineering ER. Effect of a Pilot Group Hand-washing Program on Handwashing Behaviors Among Elementary School Children in Assam. 2015 November. PMID: NCT02617225. [Irrelevant patient (age, medical condition)].


619. Odense University Hospital T, CoLab Denmark RoSD, University of Southern Denmark DLA. Telemedical Training for
Chronically Ill COPD Patients: a Cross Sectoral Study. 2016 August. PMID: NCT02754232. [Irrelevant patient (age, medical condition)].


term Outcome and Prognostic Factors. Archivos de Bronconeumologia. 2014;17. PMID: 53243547. [Foreign language study].


639. Oscroft NS, Quinnell TG, Shneerson JM, et al. A prospective, randomised, single-


641. Oxford University Hospitals NHST. Factors Associated With Chronic Respiratory Failure in Obesity. 2011 June. PMID: NCT01380418. [Irrelevant intervention (or time frame)].


663. Patrick Murphy Gs, St Thomas' Charity Ri, ResMed RF, et al. Home Mechanical Ventilation vs Home Oxygen Therapy in COPD. 2009 October. PMID: NCT00990132. [Completed Clinical Trial].


695. Qu Y, Peng H, Chen P, et al. [Combination of chest physiotherapy and intermittent non-invasive mechanical ventilation for chronic obstructive


705. Raffenberg M. Noninvasive positive pressure ventilation in chronic respiratory insufficiency. [German]. Deutsche Medizinische Wochenschrift. 2000 25


717. Rappard S, Hickey J. Just the Berries. Use of CPAP and BiPAP in acute respiratory failure. Canadian family physician Medecin


720. ResMed CRITCRIG. Registry of Stable Hypercapnic Chronic Obstructive Pulmonary Disease Treated With Non-Invasive Ventilation. 2016 July. PMID: NCT02811588. [Completed Clinical Trial].


vs oxygen supplementation in adult patients with cystic fibrosis. Pediatric Pulmonology. 2016 October;51:375. PMID: 612358533. [Abstract/ conference proceeding].


747. Rutgers TSUoNJ, National Heart L. RCT of Effects of Device-guided Breathing on Ambulatory BP. 2008 May. PMID: NCT01184755. [Irrelevant patient (age, medical condition)].


750. Saeed El Hoshy M, Ahmed Eshmawey H, Sayed El Tawab S. Outcome of pulmonary rehabilitation in patients with COPD: Comparison between patients receiving exercise training and those receiving exercise training and CPAP. Egyptian Journal
of Chest Diseases and Tuberculosis. 2017 October;66(4):609-16. PMID: 618796402. [Irrelevant intervention (or time frame)].


770. Scales DC. The implications of a tracheostomy for discharge destination. Am Thoracic Soc; 2015. [Irrelevant intervention (or time frame)].


772. Schaefer IL, Dorschner S. "Quality of life means acting independently". How do


795. Shneerson JM, Smith IE. Long term survival after hospital discharge from a Progressive Care Programme (PCP) for
patients failing to wean from invasive ventilation on ICU. Thorax. 1997;52(SUPPL. 6). [Abstract/ conference proceeding].


805. Simonds AK. Recent advances in respiratory care for neuromuscular disease. Chest. 2006;130(6):1879-86. [Irrelevant study design].


review. JAMA. 2003 Nov 05;290(17):2301-12. PMID: 14600189. [Irrelevant intervention (or time frame)].

808. Sinuff T, Keenan SP, Department of Medicine MU. Clinical practice guideline for the use of noninvasive positive pressure ventilation in COPD patients with acute respiratory failure. Journal of Critical Care. 2004 Jun;19(2):82-91. PMID: 15236140. [Irrelevant setting/location].


817. State University of New York at B. Repeat Emergency Department Visits Among Patients With Asthma and COPD. 2016 January. PMID: NCT02499887. [Irrelevant patient (age, medical condition)].


841. Tang Q, Qin G. [Therapeutic effect of long-term noninvasive positive pressure

842. Tanque AAG, Nolido RT, Benedicto JP. Noninvasive positive-pressure ventilation to prevent postextubation respiratory failure: A meta-analysis. Respiratory. 2010 November;15:51. PMID: 70313298. [Irrelevant patient (age, medical condition)].


848. Thammasat U. Patient Characteristics, Feasibility, and Outcomes of a Home Mechanical Ventilation Program in a Developing Country. 2014 November. PMID: NCT02927613. [Irrelevant comparison or no comparison].


874. Universidad Autonoma de M. Effects of Manual Therapy and Respiratory Muscle Training on the Maximal Inspiratory Pressure in Patients With Asthma. 2015 December. PMID: NCT02690831. [Irrelevant patient (age, medical condition)].
876. Universidad Autonoma de M. Video Game Exercise Effectiveness of a Domiciliary Pulmonary Rehabilitation Program in Cystic Fibrosis Patients. 2015 July. PMID: NCT02552043. [Irrelevant intervention (or time frame)].

877. Universidad de G. Physiotherapy in Patients With Stable Chronic Obstructive Pulmonary Disease. 2015 September. PMID: NCT02517411. [Irrelevant intervention (or time frame)].

878. Università degli Studi di F. Exercise Training in Chronic Obstructive Pulmonary Disease (COPD). 2008 January. PMID: NCT01218282. [Irrelevant patient (age, medical condition)].


881. University Hospital G. COPD-EXA-REHAB. Early Pulmonary Rehabilitation of Patients With Acute Exacerbation of COPD. 2013 November. PMID: NCT02987439. [Irrelevant intervention (or time frame)].

882. University Hospital M. Effects of a Therapeutic Education Program on Treatment Adherence Among Patients Prescribed At-home CPAP or At-home NIV. 2017 May 12. PMID: NCT03151317. [Completed Clinical Trial].

883. University Hospital of South Manchester NHSFT, University of M, Imperial College L, et al. The Use of Home-monitoring and mHealth Systems to Predict Asthma Control and the Occurrence of Asthma Exacerbations. 2016 April. PMID: NCT02774772. [Irrelevant patient (age, medical condition)].

884. University Hospital R. Change in Breathing Pattern on Non-invasive Ventilation of COPD Patients Under Home Mechanical Ventilation. 2017 January. PMID: NCT03018470. [Irrelevant comparison or no comparison].

885. University Hospital R. Home Non Invasive Ventilation for COPD Patients. 2011 December. PMID: NCT03221101. [Completed Clinical Trial].

886. University Hospital R. Home Non-invasive Ventilation for Chronic Obstructive Pulmonary Disease Patients. 2011 December. PMID: NCT01526642. [Irrelevant study design].

887. University of Alabama at Birmingham UoC, San Francisco VAoOR, Development UoM. IVR-Enhanced Care Transition Support for Complex Patients. 2010 February. PMID: NCT01135381. [Irrelevant intervention (or time frame)].

888. University of C. The Effect of Yoga on Cardiac Sympathetic Innervation Evaluated by I-123 mIBG. 2017 August 31. PMID: NCT03227393. [Irrelevant patient (age, medical condition)].

889. University of D, Chiesi Farmaceutici SpA. Effects of Particle Size in Small Airways Dysfunction. 2013 July. PMID:
NCT01892787. [Irrelevant patient (age, medical condition)].


891. University of M. Home-Based Diagnosis and Management of Sleep-Related Breathing Disorders in Spinal Cord Injury. 2011 October. PMID: NCT01882257. [Completed Clinical Trial].

892. University of M-C, Tyco Healthcare G. Prospective Study Comparing Different Modalities of Oxygen Delivery During Assessment of Functional Exercise Capacity. 2006 May. PMID: NCT00484562. [Irrelevant intervention (or time frame)].

893. University of W. Stroke and CPAP Outcome Study 2. 2016 June. PMID: NCT02809430. [Irrelevant patient (age, medical condition)].

894. University of Wisconsin M. Brief Trainings to Buffer Against Acute Stress Effects. 2014 October. PMID: NCT02214264. [Irrelevant patient (age, medical condition)].

895. University RA. SAS in Patients With Bronchial Carcinoma. 2014 April. PMID: NCT02270853. [Irrelevant patient (age, medical condition)].

896. Use of Respiratory Therapists (RTs) to Improve Outcomes and Quality of Life in Patients With COPD. 2014 March. PMID: NCT02078622. [Irrelevant patient (age, medical condition)].


902. Veale D, Gonzalez-Bermejo J, Borel JC, et al. [Initiation of long-term non-invasive


916. Volpato E, Banfi P, Pagnini F. A psychological intervention to promote acceptance and adherence to non-invasive ventilation in people with chronic obstructive pulmonary disease: Study protocol of a randomised controlled trial. Trials. 2017;18(1). [Irrelevant study design].


918. Wake Forest U, Wake Forest University Health S. Supportive Intervention Programs Study. 2011 June. PMID: NCT01590147. [Irrelevant patient (age, medical condition)].

919. Wang CG, Liu Y, Cao Z. Sequential non invasive ventilation (NIV) following short term invasive ventilation (IV) in COPD induced hypercapnic respiratory failure-the concept of pulmonary infection control window (PIC window) and its value for being used as the switching point: a prospective, randomised controlled study [Abstract]. European respiratory journal. 2003;22(Suppl 45):Abstract P2599. PMID: CN-00486358 UPDATE. [Abstract/conference proceeding].


945. Winterholler MG, Erbguth FJ, Hecht MJ, et al. [Survival with artificial respiration at home. An open, prospective study on home ventilation for neuromuscular diseases, in particular, the situation of ALS patients]. Nervenarzt. 2001 Apr;72(4):293-301. PMID: 11320865. [Abstract/conference proceeding].


947. Wissenschaftliches Institut Bethanien e V. Course and Complications of Invasive Out-of-hospital Ventilation. 2009 January. PMID: NCT01755039. [Irrelevant intervention (or time frame)].


959. Yale University RWJF, ResMed Foundation VAOoR. Diagnosis and Treatment of Sleep-Disordered Breathing in the Homes of Patients With Transient Ischemic Attack. 2004 November. PMID: NCT00251290. [Irrelevant patient (age, medical condition)].


## Appendix D. Characteristics of Included Studies

<table>
<thead>
<tr>
<th>Author, Year</th>
<th>Study Country, Study Design, Study Period</th>
<th>Risk of Bias</th>
<th>Inclusion / Exclusion Criteria</th>
<th>Intervention and comparisons (Groups)</th>
<th>Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.</th>
<th>Patient Characteristics</th>
<th>Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Benhamou, 1997¹</td>
<td>Observational comparative case-control study in France</td>
<td>High ROB</td>
<td>Inclusion: Treated by home non-invasive mechanical ventilation &amp; LTOT for severe chronic respiratory failure from diffuse bronchiectasis.</td>
<td>HMV (volume assist control ventilation)</td>
<td>HMV – Not FDA approved Monnal D; Taema (Antony, France)</td>
<td>14 Patients aged 64±10</td>
<td>Other</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Oxygen alone</td>
<td>No PAP</td>
<td>14 Patients aged 66±9</td>
</tr>
<tr>
<td>Bertella, 2017²</td>
<td>RCT in Italy, 03/2011-03/2014</td>
<td>Low ROB</td>
<td>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 &gt;40 km.</td>
<td>BPAP volume assured pressure support ventilation inpatient initiation</td>
<td>BPAP – not FDA approved Trend II ST 30; Hoffrichter (Schwerin, Germany)</td>
<td>25 patients aged 65.92±10.18, 32% female</td>
<td>NMD</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>BPAP volume assured pressure support ventilation outpatient initiation</td>
<td>BiPAP Synchrony II, Philips Respironics (Murrysville, PA, USA)</td>
<td>25 patients aged 61.26±8.64, 44% female</td>
</tr>
<tr>
<td>Bhatt, 2013³</td>
<td>RCT in USA</td>
<td>High ROB</td>
<td>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&lt;35 years, diseases limiting life expectancy &lt;2 years, active malignancies in previous 2 years, process precluding a nasal mask.</td>
<td>BPAP NOS</td>
<td>BPAP - FDA approved BiPAP Synchrony; Respironics Inc.</td>
<td>15 Patients, 47% female</td>
<td>COPD</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>No BPAP</td>
<td>No PAP</td>
<td>12 Patients aged 68 (IQR 65-78), 0% female</td>
</tr>
<tr>
<td>Author, Year</td>
<td>Study Country, Study Design, Study Period</td>
<td>Risk of Bias</td>
<td>Inclusion / Exclusion Criteria</td>
<td>Intervention and comparisons (Groups)</td>
<td>Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.</td>
<td>Patient Characteristics</td>
<td>Disease</td>
</tr>
<tr>
<td>-------------</td>
<td>------------------------------------------</td>
<td>--------------</td>
<td>--------------------------------</td>
<td>--------------------------------------</td>
<td>---------------------------------------------------------------</td>
<td>------------------------</td>
<td>---------</td>
</tr>
<tr>
<td>Blankenburg, 2017&lt;sup&gt;4&lt;/sup&gt;</td>
<td>Observational, Prospective in Germany, 01/01/2011 to 12/31/2011</td>
<td>Moderate ROB</td>
<td>Inclusion: COPD (GOLD criteria NOS), PaCO2&gt;7.0kPa, pH&gt;7.35, stable disease (no exacerbation in 2 weeks prior) Exclusion: decompensated heart failure or “other conditions” that cause chronic respiratory failure, systemic corticosteroids</td>
<td>HMV (pressure controlled ventilation or pressure support ventilation)</td>
<td>HMV – FDA approved VS III; ResMed (Saime SA, France)</td>
<td>51 patients aged 66.9 (SE 1.3), 37% female</td>
<td>COPD</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Inclusion: OHS (BMI &gt;30kg/m², chronic daytime hypercapnia PaCO2&gt;6.7kPa, pH&gt;7.35), symptoms of hypercapnia NOS) Exclusion: decompensated heart failure or “other conditions” that cause chronic respiratory failure</td>
<td></td>
<td></td>
<td>34 patients aged 65.4 (SE 1.8), 50% female</td>
<td>OHS</td>
</tr>
<tr>
<td>Borel, 2011&lt;sup&gt;3&lt;/sup&gt;</td>
<td>RCT in Switzerland</td>
<td>High ROB</td>
<td>Inclusion: Age 20-75 years, BMI &gt;30 Exclusion: Declined or presented any significant airway obstruction, scoliosis, cardiac failure, progressive NMD.</td>
<td>BPAP ST</td>
<td>BPAP - FDA approved GoodKnight-425ST; Covidien</td>
<td>19 Patients aged 58±11, 56% female</td>
<td>OHS</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Lifestyle counseling</td>
<td>No PAP</td>
<td>18 Patients aged 54±6, 59% female</td>
</tr>
<tr>
<td>Bourke, 2006&lt;sup&gt;6&lt;/sup&gt;</td>
<td>RCT in United Kingdom, 03/2000 to 12/2003</td>
<td>High ROB</td>
<td>Inclusion: Current or previous NIPPV use, significant comorbidities, age&gt;75 years, inability to complete quality of life assessment.</td>
<td>BPAP ST (full cohort)</td>
<td>BPAP - FDA approved VPAP STII; ResMed United Kingdom Ltd (Abingdon, United Kingdom)</td>
<td>22 Patients aged 63.7±10.3, 36% female</td>
<td>NMD</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>No BPAP ST (full cohort)</td>
<td>No PAP</td>
<td>19 Patients aged 63±8.1, 47% female</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>BPAP ST (good bulbar patients)</td>
<td>BPAP good bulbar</td>
<td>11 Patients</td>
</tr>
<tr>
<td>Author, Year</td>
<td>Study Country, Study Design, Study Period</td>
<td>Risk of Bias</td>
<td>Inclusion / Exclusion Criteria</td>
<td>Intervention and comparisons (Groups)</td>
<td>Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.</td>
<td>Patient Characteristics</td>
<td>Disease</td>
</tr>
<tr>
<td>-------------</td>
<td>------------------------------------------</td>
<td>-------------</td>
<td>--------------------------------</td>
<td>--------------------------------------</td>
<td>---------------------------------------------------------------</td>
<td>------------------------</td>
<td>---------</td>
</tr>
<tr>
<td>Budweiser, 2007</td>
<td>Observational Prospective in Germany, 01/2002 to 12/2005</td>
<td>Low ROB</td>
<td>Inclusion: Less than 80 years old, severe COPD (GOLD IV), FEV1/FVC &lt;70%, FEV1 &lt;50% predicted, PaCO2 &gt; 50mmHg after therapy/treatment for exacerbation Exclusion: Malignancy diagnosis within prior 5 years, intubation or tracheostomy prior to NIPPV</td>
<td>BPAP (pressure controlled ventilation)</td>
<td>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</td>
<td>99 Patients aged 34.2±8.4, 36.4% female</td>
<td>COPD</td>
</tr>
<tr>
<td>Buyse, 2003</td>
<td>Observational Retrospective in Belgium, 09/1990 to 03/2001</td>
<td>Moderate ROB</td>
<td>Inclusion: Consecutive kyphoscoliosis &amp; respiratory insufficiency patients started on LTOT and/or NIPPV at center.</td>
<td>HMV (volume or pressure cycled ventilator NOS) + oxygen</td>
<td>HMV - FDA approved Ecole 3 (volume-cycled ventilator); Saime (Savigny-Le-Temple, France) or O’nyx (pressure cycled ventilator); Nellcor Puritan Bennet (Villers-les-Nancy, France)</td>
<td>18 Patients aged 61±7, 77.8% female</td>
<td>TRD</td>
</tr>
<tr>
<td>Casanova, 2000</td>
<td>RCT in Spain, 1995 to 1997</td>
<td>High ROB</td>
<td>Inclusion: Age 45-75 years, smoking history 20 pack years, clinically stable</td>
<td>BPAP S + standard care</td>
<td>BPAP – Not FDA approved DP-90; Taema (Paris, France)</td>
<td>20 Patients aged 64±5, 0% female</td>
<td>COPD</td>
</tr>
<tr>
<td>Author, Year</td>
<td>Study Country, Study Design, Study Period</td>
<td>Risk of Bias</td>
<td>Inclusion / Exclusion Criteria</td>
<td>Intervention and comparisons (Groups)</td>
<td>Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.</td>
<td>Patient Characteristics</td>
<td>Disease</td>
</tr>
<tr>
<td>--------------</td>
<td>------------------------------------------</td>
<td>--------------</td>
<td>--------------------------------</td>
<td>--------------------------------------</td>
<td>---------------------------------------------------------------</td>
<td>------------------------</td>
<td>---------</td>
</tr>
<tr>
<td>Castillejo, 2014&lt;sup&gt;10&lt;/sup&gt;</td>
<td>Observational Prospective in Spain, 1998 to 2010</td>
<td>High ROB</td>
<td>Inclusion: OHS &amp; BMI &gt;30 Exclusion: Obstructive disease with FEV1/FVC ratio &lt;70%, NMD with respiratory involvement, respiratory disease other than OHS.</td>
<td>BPAP ST in OHS without OSA</td>
<td>BPAP OHS OSA - FDA approved Harmony BiPAP; Respironics (Louisville, USA)</td>
<td>50 Patients aged 64.6±9.8, 82% female</td>
<td>OHS</td>
</tr>
<tr>
<td>Cheung, 2010&lt;sup&gt;11&lt;/sup&gt;</td>
<td>RCT in China, 01/2007 to 03/2009</td>
<td>High ROB</td>
<td>Inclusion: Severe exacerbation with persistent respiratory acidosis (despite treatment with bronchodilators, corticosteroids, antibiotics), required NIPPV treatment Exclusion: Active smokers, RF from non-COPD cause, evidence of pneumonia, transmissible infections, requiring long-term systemic steroids, comorbidity giving life expectancy &lt;1 year, significant OSA, already on home NIPPV, inability to comply with study protocol.</td>
<td>CPAP</td>
<td>CPAP - FDA approved BiPAP Synchrony; Respironics Inc. (Murrysville, USA)</td>
<td>24 Patients aged 71±7.7, 8.3% female</td>
<td>COPD</td>
</tr>
<tr>
<td>Chiang, 2003&lt;sup&gt;12&lt;/sup&gt;</td>
<td>RCT in Taiwan, 06/2001 to 11/2002</td>
<td>Moderate ROB</td>
<td>Inclusion: Diagnosed with COPD and asthma and bronchiectasis, repeat admission due to lung deterioration despite treatment, well-motivated, sleepy during</td>
<td>BPAP NOS</td>
<td>BPAP - FDA approved Respironics (Murrysville, USA)</td>
<td>13 Patients aged 62.5±11.5, 23.1% female</td>
<td>COPD, Other</td>
</tr>
<tr>
<td>Author, Year</td>
<td>Study Country, Study Design, Study Period</td>
<td>Risk of Bias</td>
<td>Inclusion / Exclusion Criteria</td>
<td>Intervention and comparisons (Groups)</td>
<td>Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.</td>
<td>Patient Characteristics</td>
<td>Disease</td>
</tr>
<tr>
<td>--------------</td>
<td>------------------------------------------</td>
<td>--------------</td>
<td>--------------------------------</td>
<td>-------------------------------------</td>
<td>---------------------------------------------------------------</td>
<td>------------------------</td>
<td>---------</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>COPD</td>
</tr>
<tr>
<td>Clinis, 1996&lt;sup&gt;13&lt;/sup&gt; Observational Prospective in Italy, 12/1991 to 09/1992</td>
<td>High ROB</td>
<td>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.</td>
<td>no BPAP NOS</td>
<td>No PAP</td>
<td>14 Patients aged 65.5±10, 35.7% female</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Clinis, 1998&lt;sup&gt;14&lt;/sup&gt; Observational Prospective in Italy, 12/1991 to 12/1994</td>
<td>Low ROB</td>
<td>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</td>
<td>BPAP ST + home care + oxygen</td>
<td>BPAP - FDA approved BiPAP; Respironics (Murrysville, USA)</td>
<td>17 Patients aged 62±3, 29.4% female</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Clinis, 2002&lt;sup&gt;13&lt;/sup&gt; RCT in Italy/France, 06/1996 to 01/2000</td>
<td>High ROB</td>
<td>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.</td>
<td>BPAP ST + oxygen</td>
<td>BPAP - FDA approved BiPAP; Respironics (Murrysville, USA)</td>
<td>28 Patients aged 66±6, 21.4% female</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>COPD</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>COPD</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>COPD</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>COPD</td>
</tr>
</tbody>
</table>

D-5
<table>
<thead>
<tr>
<th>Author, Year</th>
<th>Study Country, Study Design, Study Period</th>
<th>Risk of Bias</th>
<th>Inclusion / Exclusion Criteria</th>
<th>Intervention and comparisons (Groups)</th>
<th>Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.</th>
<th>Patient Characteristics</th>
<th>Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Coco, 2006&lt;sup&gt;16&lt;/sup&gt;</td>
<td>Observational Prospective in Italy, 10/1999 to 07/2003</td>
<td>Low ROB</td>
<td>Exclusion: 15% increase FEV1 after salbutamol, pH ≤7.34, active smokers, history of OSA (defined by apnea-hypopnea index &gt;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.</td>
<td>LTOT</td>
<td>No PAP</td>
<td>47 Patients aged 66±14, 21.3% female</td>
<td>NMD</td>
</tr>
<tr>
<td>Crespo, 2009&lt;sup&gt;17&lt;/sup&gt;</td>
<td>Observational Retrospective in Spain, 1998 to 2001</td>
<td>High ROB</td>
<td>Inclusion: Definite/probable ALS Exclusion: Primary lateral sclerosis, diagnosis other than ALS during followup.</td>
<td>BPAP ST (use ≥ 4 hours/day)</td>
<td>BPAP 4 hour+ FDA approved BiPAP; Respironics (Vitalaire, Italy)</td>
<td>44 Patients aged 62.3±11.4, 31.8% female</td>
<td>COPD, TRD, NMD, OHS, Other</td>
</tr>
<tr>
<td>De Backer, 2011&lt;sup&gt;18&lt;/sup&gt;</td>
<td>RCT in Belgium</td>
<td>Moderate ROB</td>
<td>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.</td>
<td>HMV (pressure or volume NOS) in age ≥ 75 years old</td>
<td>HMV Age75</td>
<td>10 Patients aged 76.9±2.1, 30% female</td>
<td>COPD</td>
</tr>
<tr>
<td>Author, Year</td>
<td>Study Country, Study Design, Study Period</td>
<td>Risk of Bias</td>
<td>Inclusion / Exclusion Criteria</td>
<td>Intervention and comparisons (Groups)</td>
<td>Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.</td>
<td>Patient Characteristics</td>
<td>Disease</td>
</tr>
<tr>
<td>-------------</td>
<td>-----------------------------------------</td>
<td>--------------</td>
<td>--------------------------------</td>
<td>-------------------------------------</td>
<td>---------------------------------------------------------------</td>
<td>-------------------------</td>
<td>---------</td>
</tr>
</tbody>
</table>
| Domenéch-Clar, 2003<sup>19</sup> | Observational Prospective in Spain, 01/1997 to 11/2001 | High ROB | NIPPV before admission  
Exclusion: Invasive ventilation, asthmatic, restrictive lung disease, malignancy, heart failure, OSA. | Standard care | No PAP | 5 Patients aged 66±6 | TRD |
| Dreher, 2010<sup>20</sup> | RCT in Germany | High ROB | Inclusion: CHRF due to COPD stage IV  
Exclusion: Acute RF, invasive ventilation via tracheostomy, weaned from invasive ventilation, intubated during prior 3 months, other ventilatory support prior to study. | HMV (pressure assist/control) (time period 1)  
HMV (PSV ST) (time period 1)  
HMV (PSV ST) (time period 2)  
HMV (pressure assist/control) (time period 2) | HMV Pres Control - FDA approved Breas Vivo 40; Breas Medical AB (Molnlycke, Sweden) or Smart Air; Airox (Pau Cedex, France)  
HMV PSV ST - FDA approved Breas Vivo 40; Breas Medical AB (Molnlycke, Sweden) or Smart Air; Airox (Pau Cedex, France)  
HMV PSV ST  
HMV Pres Control | 9 Patients  
8 Patients  
8 Patients  
24 Patients aged 63±10, 33.3% female | COPD |
| Duiverman, 2011<sup>21</sup> | RCT in Netherlands | Moderate ROB | Inclusion: COPD stage III/IV, age 40-76 years, clinically stable, chronic hypercapnic RF  
Exclusion: cardiac/neuromuscular disease | BPAP ST + pulmonary rehabilitation | BPAP - FDA approved BiPAP Synchrony; Respironics Inc. | 24 Patients aged 63±10, 33.3% female | COPD |
<table>
<thead>
<tr>
<th>Author, Year</th>
<th>Study Country, Study Design, Study Period</th>
<th>Risk of Bias</th>
<th>Inclusion / Exclusion Criteria</th>
<th>Intervention and comparisons (Groups)</th>
<th>Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.</th>
<th>Patient Characteristics</th>
<th>Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Duiverman, 2017</td>
<td>RCT in Netherlands</td>
<td>High ROB</td>
<td>Limiting exercise tolerance, exposure to pulmonary rehab program (previous 18 months), previous exposure to chronic NIPPV ever, apnea-hypopnea index ≥10h.</td>
<td>Pulmonary rehabilitation alone</td>
<td>No PAP</td>
<td>32 Patients aged 61±8, 46.9% female</td>
<td>COPD</td>
</tr>
<tr>
<td>Durao, 2018</td>
<td>Observational Retrospective in Portugal, 08/1/2011 to 07/31/2014</td>
<td>Low ROB</td>
<td>Inclusion: COPD (GOLD III or IV), ≥ 2 AECOPD with acute hypercapnic respiratory failure (pH&lt;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&gt;7.35). Exclusion: TRD, NMD</td>
<td>HMV/BPAP mix (pressure controlled ventilation) (high intensity)</td>
<td>HMV – FDA approved Vivo 50; Breas Medical (Molndal, Sweden) BPAP – FDA approved Stellar 100; Resmed (Martinsried, Germany)</td>
<td>62 patients aged 64.8±10.4, 12.9% female</td>
<td>COPD</td>
</tr>
<tr>
<td>Durao, 2018</td>
<td>Observational Retrospective in Portugal, 08/1/2011 to 07/31/2014</td>
<td>Low ROB</td>
<td>Inclusion: COPD NOS Exclusion: No clinical assessment in prior 6 months, OSA with a history of noncompliance with CPAP</td>
<td>HMV/BPAP mix started in AECOPD</td>
<td>BPAP – FDA approved VPAP ST S9; Resmed VPAP ST STA; Resmed BiPAP PR1; Philips Respironics BiPAP A30; Philips Respironics BiPAP A40; Philips Respironics</td>
<td>47 patients aged 66.9±8.4, 17% female</td>
<td>COPD</td>
</tr>
<tr>
<td>Author, Year</td>
<td>Study Country, Study Design, Study Period</td>
<td>Risk of Bias</td>
<td>Inclusion / Exclusion Criteria</td>
<td>Intervention and comparisons (Groups)</td>
<td>Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.</td>
<td>Patient Characteristics</td>
<td>Disease</td>
</tr>
<tr>
<td>--------------</td>
<td>------------------------------------------</td>
<td>-------------</td>
<td>---------------------------------</td>
<td>--------------------------------------</td>
<td>---------------------------------------------------------------------</td>
<td>------------------------</td>
<td>---------</td>
</tr>
<tr>
<td>Funk, 2010&lt;sup&gt;25&lt;/sup&gt;</td>
<td>RCT in Austria, 04/01/2003 to 02/28/2007</td>
<td>Moderate ROB</td>
<td>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.</td>
<td>BPAP NOS</td>
<td>BPAP - Not reported (&quot;various types of patient-triggered bi-level positive pressure ventilators were used&quot;)</td>
<td>13 Patients aged 62±6, 46% female</td>
<td>COPD</td>
</tr>
<tr>
<td>Gad, 2014&lt;sup&gt;26&lt;/sup&gt;</td>
<td>Observational Prospective in Egypt, 10/2012 to 04/2014</td>
<td>Moderate ROB</td>
<td>Inclusion: Severe COPD stage III/IV, FEV1/FVC &lt;70%, clinically stable Exclusion: invasive mechanical ventilation, OSA, cardiac disease limiting exercise tolerance, NMDs, orthopedic impairment of shoulder girdle</td>
<td>BPAP ST + exercise program</td>
<td>Exercise program</td>
<td>No PAP</td>
<td>15 Patients aged 65.70±10, 40% female</td>
</tr>
<tr>
<td>Galli, 2014&lt;sup&gt;27&lt;/sup&gt;</td>
<td>Observational Retrospective in USA, 01/2011 to 12/2011</td>
<td>High ROB</td>
<td>Inclusion: Primary/secondary discharge diagnosis of AECOPD, hypercapnic RF during hospitalization Exclusion: discharged to hospice, no documented hypercapnia, not receiving NIPPV during hospitalization.</td>
<td>BPAP NOS post hospital admission</td>
<td>No BPAP post hospital admission</td>
<td>No PAP</td>
<td>88 Patients aged 64.9±10.8, 67% female</td>
</tr>
<tr>
<td>Garrod, 2000&lt;sup&gt;28&lt;/sup&gt;</td>
<td>RCT in England</td>
<td>High ROB</td>
<td>Inclusion: Severe COPD, all patients had limited exercise tolerance due to dyspnea and no previous exposure to NIPPV</td>
<td>BPAP S + pulmonary rehabilitation</td>
<td>BPAP - FDA approved BiPAP ST 30; Respironics (Murrysville, USA)</td>
<td>23 Patients aged 63</td>
<td>COPD</td>
</tr>
<tr>
<td>Author, Year</td>
<td>Study Country, Study Design, Study Period</td>
<td>Risk of Bias</td>
<td>Inclusion / Exclusion Criteria</td>
<td>Intervention and comparisons (Groups)</td>
<td>Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.</td>
<td>Patient Characteristics</td>
<td>Disease</td>
</tr>
<tr>
<td>--------------</td>
<td>------------------------------------------</td>
<td>--------------</td>
<td>-----------------------------</td>
<td>--------------------------------------</td>
<td>---------------------------------------------------------------</td>
<td>------------------------</td>
<td>---------</td>
</tr>
<tr>
<td>Gay, 1996&lt;sup&gt;19&lt;/sup&gt;</td>
<td>RCT in USA, 1989 to 1992</td>
<td>High ROB</td>
<td>Exclusion: unstable angina, intermittent claudication, and other mobility-limiting conditions.</td>
<td>Pulmonary rehabilitation</td>
<td>No PAP</td>
<td>22 Patients aged 67</td>
<td>COPD</td>
</tr>
</tbody>
</table>
| Gonzalez-Bermejo, 2013<sup>16</sup> | Observational Retrospective in France, 01/01/2003 to 12/31/2007 | 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 - FDA approved BiPAP; Respirronics (Murrysville, USA) | 7 Patients aged 71.0±4.5, 28.6% female | NMD |
| Gonzalez-Bermejo, 2013<sup>16</sup> | Observational Retrospective in France, 01/01/2003 to 12/31/2007 | High ROB | Inclusion: 4h/night minimal adherence  
Exclusion: Use of other ventilator types, without integrated SpO2 monitoring. | BPAP ST "correctly ventilated patients" | BPAP Correct ventilated patients - FDA approved VPAP-III or VPAP-IV plus automatic ventilatory signal analysis (Reslink); Resmed (Sydney, Australia) | 40 Patients aged 63±12, 32.5% female | NMD |
<p>| Gonzalez-Bermejo, 2013&lt;sup&gt;16&lt;/sup&gt; | Observational Retrospective in France, 01/01/2003 to 12/31/2007 | High ROB | BPAP ST &quot;insufficiently ventilated patients&quot; | BPAP Insufficient ventilated patients - FDA approved VPAP-III or VPAP-IV plus automatic ventilatory signal analysis (Reslink); Resmed (Sydney, Australia) | 42 Patients aged 64±10, 17% female | NMD |</p>
<table>
<thead>
<tr>
<th>Author, Year</th>
<th>Study Country, Study Design, Study Period</th>
<th>Risk of Bias</th>
<th>Inclusion / Exclusion Criteria</th>
<th>Intervention and comparisons (Groups)</th>
<th>Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.</th>
<th>Patient Characteristics</th>
<th>Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hazenberg, 2014&lt;sup&gt;11&lt;/sup&gt;</td>
<td>RCT in Netherlands, 10/2008 to 10/2012</td>
<td>Moderate ROB</td>
<td>Inclusion: Chronic RF from NMD or thoracic cage disorder, orthopnea from diaphragm paralysis &amp; daytime normocapnia also included Exclusion: Strictly COPD patients, not mask naive, acute RF, age &lt; 18 years, invasive ventilation, nursing home residing. (77 total patients in both groups, 3 patients on volume control, 74 patients on pressure control)</td>
<td>HMV started at home pressure controlled ventilation with change to volume assist control ventilation if not tolerated</td>
<td>HMV Home - FDA approved Elisee 150; ResMed (Paris, France)</td>
<td>38 Patients aged 59.9±12.6, 47.4% female</td>
<td>NMD, TRD</td>
</tr>
<tr>
<td>Heinemann, 2011&lt;sup&gt;12&lt;/sup&gt;</td>
<td>Observational Retrospective in Germany, 01/2002 to 02/2008</td>
<td>High ROB</td>
<td>Inclusion: COPD, prolonged weaning from invasive mechanical ventilation Exclusion: Intubated from cardiogenic edema or cardiopulmonary resuscitation</td>
<td>BPAP (pressure controlled ventilation) No BPAP No PAP</td>
<td>No BPAP</td>
<td>39 Patients aged 64.6±10.8, 30.1% female</td>
<td>COPD</td>
</tr>
<tr>
<td>Hitzl, 2009&lt;sup&gt;13&lt;/sup&gt;</td>
<td>Observational Prospective in Germany</td>
<td>Low ROB</td>
<td>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.</td>
<td>HMV (pressure controlled ventilation) in COPD HMV (pressure controlled ventilation) in restrictive thoracic disease</td>
<td>HMV COPD</td>
<td>93 Patients aged 65.5±8, 30.1% female</td>
<td>COPD</td>
</tr>
<tr>
<td>Howard, 2016&lt;sup&gt;14&lt;/sup&gt;</td>
<td>RCT in Australia, 11/01/2011 to 12/31/2013</td>
<td>Moderate ROB</td>
<td>Inclusion: Primary OHS diagnosis Exclusion: Other conditions contributing to hypoventilation.</td>
<td>BPAP ST</td>
<td>BPAP - FDA approved Harmony; Philips Respironics (USA) or VPAP III STa; ResMed (Bella Vista, Australia)</td>
<td>29 Patients aged 53.2±10.7, 51.7% female</td>
<td>OHS</td>
</tr>
<tr>
<td>Author, Year</td>
<td>Study Country, Study Design, Study Period</td>
<td>Risk of Bias</td>
<td>Inclusion / Exclusion Criteria</td>
<td>Intervention and comparisons (Groups)</td>
<td>Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.</td>
<td>Patient Characteristics</td>
<td>Disease</td>
</tr>
<tr>
<td>--------------</td>
<td>------------------------------------------</td>
<td>-------------</td>
<td>-------------------------------</td>
<td>---------------------------------------</td>
<td>---------------------------------------------------------------</td>
<td>------------------------</td>
<td>---------</td>
</tr>
<tr>
<td>Köhnlein, 2014</td>
<td>RCT in Germany and Austria, 10/29/2004 to 07/31/2011</td>
<td>High ROB</td>
<td>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.</td>
<td>BPAP ST + standard care</td>
<td>CPAP - FDA approved Harmony; Philips Respironics (USA) or VPAP III St; ResMed (Bella Vista, Australia)</td>
<td>31 Patients aged 52.9±10, 41.9% female</td>
<td>COPD</td>
</tr>
<tr>
<td>Marquez-Martin, 2014</td>
<td>RCT in Spain, 05/2007 to 09/2011</td>
<td>Moderate ROB</td>
<td>Inclusion: Adults with COPD, clinically stable, chronic RF with hypoxemia.</td>
<td>BPAP ST</td>
<td>BPAP - FDA approved BiPAP; Respironics (Murrysville, USA)</td>
<td>15 Patients aged 69 (64-73)</td>
<td>COPD</td>
</tr>
<tr>
<td>Masa, 2000</td>
<td>Observational Prospective in Spain</td>
<td>Moderate ROB</td>
<td>Inclusion: OHS or kyphoscoliosis Exclusion: Apnea-hypopnea index &gt;20 events/h.</td>
<td>HMV (volume cycled or pressure cycled) in OHS</td>
<td>HMV OHS - Onyx FDA approved, Monal not FDA approved Monal DCC (volume control HMV); Taema (Paris, France). ** If could not tolerate Monal DCC, then patients were switched to a Onyx Plus Mallinckrodt SEFAM (Nancy, France) (which is a &quot;bilevel pressure device&quot;)</td>
<td>22 Patients aged 61±14, 81.8% female</td>
<td>OHS</td>
</tr>
<tr>
<td>Author, Year</td>
<td>Study Country, Study Design, Study Period</td>
<td>Risk of Bias</td>
<td>Inclusion / Exclusion Criteria</td>
<td>Intervention and comparisons (Groups)</td>
<td>Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.</td>
<td>Patient Characteristics</td>
<td>Disease</td>
</tr>
<tr>
<td>--------------</td>
<td>------------------------------------------</td>
<td>--------------</td>
<td>--------------------------------</td>
<td>---------------------------------------</td>
<td>---------------------------------------------------------------</td>
<td>------------------------</td>
<td>---------</td>
</tr>
</tbody>
</table>
| Masa, 2015<sup>39</sup> | 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 - FDA approved  
Breas Vivo 40 (General Electric, England), BiPAP AVAPS (Philips-Respironics, Netherlands), Trilogy 100 (Philips-Respironics, Netherlands), VS Ultra (ResMed, Australia, Monal T50 (Air Liquide, France), Puritan Bennett 560 (Puritan Bennett, USA) | 71 Patients aged 64±11, 85% female | OHS |
| Masa, 2015<sup>38</sup> | | | | | | 14 Patients aged 43±20, 50% female | TRD |

**Note:** HMV Kyphoscoliosis - Onyx FDA approved, Monal not FDA approved  
Monal DCC (volume control HMV); Taema (Paris, France). ** If could not tolerate Monal DCC, then patients were switched to a Onyx Plus Mallinckrodt SEFAM (Nancy, France) (which is a "bilevel pressure device")
<table>
<thead>
<tr>
<th>Author, Year</th>
<th>Study Country, Study Design, Study Period</th>
<th>Risk of Bias</th>
<th>Inclusion / Exclusion Criteria</th>
<th>Intervention and comparisons (Groups)</th>
<th>Device used (HMV, CPAP, BAP) manufacturer, brand name, model no.</th>
<th>Patient Characteristics</th>
<th>Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>McEvoy, 2009&lt;sup&gt;45&lt;/sup&gt;</td>
<td>RCT in Australia, 06/30/1998 to 05/15/2004</td>
<td>Moderate ROB</td>
<td>Inclusion: Age&lt;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&gt;40, evidence of sleep apnea.</td>
<td>BPAP S + Oxygen</td>
<td>BPAP - FDA approved VPAP S mode; ResMed (Sydney, Australia)</td>
<td>72 Patients aged 67.2 (IQR 65.3 to 69.1), 31% female</td>
<td>COPD</td>
</tr>
<tr>
<td>Munoz, 2005&lt;sup&gt;41&lt;/sup&gt;</td>
<td>Observational Retrospective in Spain, 1997 to 2001</td>
<td>Moderate ROB</td>
<td>Inclusion: Treated with non-invasive home volumetric ventilator, followup ≥1 year Exclusion: BPAP users.</td>
<td>HMV volume assist control ventilation</td>
<td>HMV volume assist/control mode</td>
<td>45 Patients aged 65.1±12.9, 48.9% female</td>
<td>NMD, TRD</td>
</tr>
<tr>
<td>Murphy, 2012&lt;sup&gt;42&lt;/sup&gt;</td>
<td>RCT in United Kingdom</td>
<td>Moderate ROB</td>
<td>Inclusion: BMI&gt;40, absence of other identifiable hypoventilation cause Exclusion: inability to provide written consent.</td>
<td>BPAP AVAPS</td>
<td>BPAP AVAPS - FDA approved BiPAP synchrony; Respironics (Murrysville, USA)</td>
<td>25 Patients aged 53±9, 52% female</td>
<td>OHS</td>
</tr>
<tr>
<td>Murphy, 2017&lt;sup&gt;43&lt;/sup&gt;</td>
<td>RCT in United Kingdom, 2010 to 2015</td>
<td>High ROB</td>
<td>Inclusion: Persistent hypercapnia and hypoxemia, &gt;30% sleep time &lt;90% oxygen saturation, arterial pH &gt;7.30 breathing room air Exclusion: BMI &gt;35, OSA, other RF causes.</td>
<td>BPAP ST + Home oxygen</td>
<td>BPAP – FDA approved Harmony 2; Philips Respironics or VPAP III STa; ResMed</td>
<td>57 Patients aged 66.4±10.2, 51% female</td>
<td>COPD</td>
</tr>
<tr>
<td>Mattos, 2018&lt;sup&gt;44&lt;/sup&gt;</td>
<td>RCT in United Kingdom, 2012 to 2016</td>
<td>High ROB</td>
<td>Inclusion: Persistent hypercapnia and hypoxemia, &gt;30% sleep time &lt;90% oxygen saturation, arterial pH &gt;7.30 breathing room air Exclusion: BMI &gt;35, OSA, other RF causes.</td>
<td>BPAP ST</td>
<td>BPAP - FDA approved VPAP S mode; ResMed (Sydney, Australia)</td>
<td>59 Patients aged 67.1±9.0, 54% female</td>
<td>COPD</td>
</tr>
<tr>
<td>Author, Year</td>
<td>Study Country, Study Design, Study Period</td>
<td>Risk of Bias</td>
<td>Inclusion / Exclusion Criteria</td>
<td>Intervention and comparisons (Groups)</td>
<td>Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.</td>
<td>Patient Characteristics</td>
<td>Disease</td>
</tr>
<tr>
<td>--------------</td>
<td>-------------------------------------------</td>
<td>--------------</td>
<td>-------------------------------</td>
<td>--------------------------------------</td>
<td>-------------------------------------------------</td>
<td>------------------------</td>
<td>---------</td>
</tr>
<tr>
<td>Nauffal, 2002</td>
<td>Observational Prospective in Spain, 01/1997 to 03/2000</td>
<td>Moderate ROB</td>
<td>Inclusion: Chronic hypoventilation due to kyphoscoliosis or NMD, moderate to severe restrictive ventilatory pattern, clinically stable.</td>
<td>BPAP NOS in kyphoscoliosis</td>
<td>BPAP Kyphoscoliosis – Not FDA approved DP-90; Taema (Paris, France)</td>
<td>35 Patients aged 55.9, 40% female</td>
<td>TRD</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>BPAP NOS in neuromuscular diseases</td>
<td>BPAP NMD – Not FDA approved DP-90; Taema (Paris, France)</td>
<td>27 Patients aged 42.5, 48% female</td>
<td>NMD</td>
</tr>
<tr>
<td>Oscroft, 2010</td>
<td>Observational Retrospective in United Kingdom, 01/2000 to 12/2003</td>
<td>Moderate ROB</td>
<td>Inclusion: COPD diagnosis, smoking history &gt;20 pack years, ventilatory failure with a daytime PaCO2 &gt; 7 kPa with a pH &gt; 7.35 or nocturnal transcutaneous PaCO2 &gt; 9 kPa, hospital admission immediately prior to referral with clinical diagnosis of exacerbation of COPD Exclusion: Age&gt;80 years, other respiratory disease, BMI&gt;35, significant OSA, tracheostomy, impaired left ventricular function.</td>
<td>BPAP ST started after AECOPD</td>
<td>PSV ST AECOPD – Not FDA approved NIPPY I, 2 or 3; B &amp; D Electromedical (Stratford, United Kingdom)</td>
<td>31 Patients aged 66±6, 49% female</td>
<td>COPD</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>BPAP ST started in stable patient without exacerbation</td>
<td>PSV ST Stable – Not FDA approved NIPPY I, 2 or 3; B &amp; D Electromedical (Stratford, United Kingdom)</td>
<td>16 Patients aged 63±7</td>
<td></td>
</tr>
<tr>
<td>Oscroft, 2014</td>
<td>RCT in United Kingdom, 09/2007 to 12/2011</td>
<td>High ROB</td>
<td>Inclusion: COPD diagnosis, smoking history &gt;20 pack years, ventilatory failure with a daytime PaCO2 &gt; 7 kPa with a pH &gt; 7.35 or nocturnal transcutaneous PaCO2 &gt; 9 kPa Exclusion: Age&gt;80 years, other respiratory disease, BMI&gt;40, significant OSA.</td>
<td>BPAP IVAPS</td>
<td>BPAP IVAPS - FDA approved Intelligent volume assured pressure support (iVAPS); ResMed (Bella Vista, Australia)</td>
<td>20 Patients aged 67.6±7.9, 55% female</td>
<td>COPD</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>BPAP ST</td>
<td>BPAP ST – Not FDA approved NIPPY 3; B and D Electromedical (Stratford, United Kingdom)</td>
<td>20 Patients aged 67.4±8.2, 50% female</td>
<td></td>
</tr>
<tr>
<td>Author, Year</td>
<td>Study Country, Study Design, Study Period</td>
<td>Risk of Bias</td>
<td>Inclusion / Exclusion Criteria</td>
<td>Intervention and comparisons (Groups)</td>
<td>Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.</td>
<td>Patient Characteristics</td>
<td>Disease</td>
</tr>
<tr>
<td>--------------</td>
<td>------------------------------------------</td>
<td>--------------</td>
<td>-------------------------------</td>
<td>--------------------------------------</td>
<td>---------------------------------------------------------------</td>
<td>------------------------</td>
<td>---------</td>
</tr>
<tr>
<td>Paone, 2014&lt;sup&gt;47&lt;/sup&gt;</td>
<td>Observational Prospective in Italy, 3/2007 and 1/2010</td>
<td>Low ROB</td>
<td>Inclusion: Acute RF needing NIPPV, clinical stability with symptoms of nocturnal Hypoventilation, FEV1 &lt; 50% predicted, &lt;20% improvement in FEV1 following bronchodilator and a ratio FEV1/FVC &lt; 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&gt;40, systemic steroids therapy.</td>
<td>BPAP ST (PSV ST) + Home oxygen</td>
<td>BPAP - FDA approved Synchrony; Philips Respironics (Andover MA, USA) or Neftis; Linde (Munich Germany)</td>
<td>48 Patients, 56.2% female</td>
<td>COPD</td>
</tr>
<tr>
<td>Pinto, 1995&lt;sup&gt;48&lt;/sup&gt;</td>
<td>Observational Prospective in Portugal</td>
<td>High ROB</td>
<td>Inclusion: Consecutive ALS patients with bulbar features Exclusion: tracheotomised, refusal of attempts to prolong survival.</td>
<td>BPAP NOS</td>
<td>BPAP</td>
<td>10 Patients aged 60.66, 45% female between both groups</td>
<td>NMD</td>
</tr>
<tr>
<td>Pinto, 2010&lt;sup&gt;49&lt;/sup&gt;</td>
<td>Observational Prospective in Portugal, 01/2003 to 09/2006</td>
<td>High ROB</td>
<td>Inclusion: No signs/symptoms of respiratory insufficiency, age 18-75 years Exclusion: Gastrostomy, cognitive impairment, other significant disorders.</td>
<td>BPAP ST + weekly telemonitoring + standard care</td>
<td>BPAP Telemonitor - FDA approved Goodknight 425ST bi-level device; Tyco Healthcare Group LP (California, USA)</td>
<td>20 Patients aged 62±12.90, 31.6% female</td>
<td>NMD</td>
</tr>
<tr>
<td>Author, Year</td>
<td>Study Country, Study Design, Study Period</td>
<td>Risk of Bias</td>
<td>Inclusion / Exclusion Criteria</td>
<td>Intervention and comparisons (Groups)</td>
<td>Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.</td>
<td>Patient Characteristics</td>
<td>Disease</td>
</tr>
<tr>
<td>--------------</td>
<td>----------------------------------------</td>
<td>-------------</td>
<td>--------------------------------</td>
<td>-------------------------------------</td>
<td>------------------------------------------------------------------</td>
<td>--------------------------------</td>
<td>---------</td>
</tr>
<tr>
<td>Piper, 2008&lt;sup&gt;50&lt;/sup&gt;</td>
<td>RCT in Australia</td>
<td>Low ROB</td>
<td>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.</td>
<td>BPAP S</td>
<td>BPAP</td>
<td>18 Patients aged 47±13, 50% female</td>
<td>OHS</td>
</tr>
<tr>
<td>Salturk, 2015&lt;sup&gt;11&lt;/sup&gt;</td>
<td>Observational Retrospective in Turkey, 01/2011 to 01/2012</td>
<td>Low ROB</td>
<td>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.</td>
<td>BPAP ST COPD BPAP ST OHS BPAP ST Kyphoscoliosis BPAP ST Diffuse Parenchymal Lung Disease</td>
<td>BPAP COPD BPAP OHS BPAP</td>
<td>37 Patients aged 65±10, 8.1% female 34 Patients aged 65±8, 50% female 20 Patients aged 46±10, 45% female 14 Patients aged 62±12, 21.4% female</td>
<td>COPD OHS TRD Other</td>
</tr>
<tr>
<td>Sancho, 2014&lt;sup&gt;12&lt;/sup&gt;</td>
<td>Observational Retrospective in Spain/France, 03/2003 to 12/2007</td>
<td>Low ROB</td>
<td>Inclusion: Indication for NIPPV from presence of hypoventilation symptoms Exclusion: Presence of HMV (volume assist control ventilation)</td>
<td>HMV (volume assist control ventilation)</td>
<td>HMV - FDA approved PV 501; Breas Medical (Molndal, Sweden) or Legendair; Airox (Pau, France)</td>
<td>62 Patients aged 62.21±8.81, 54.8% female</td>
<td>NMD</td>
</tr>
<tr>
<td>Author, Year</td>
<td>Study Country, Study Design, Study Period</td>
<td>Risk of Bias</td>
<td>Inclusion / Exclusion Criteria</td>
<td>Intervention and comparisons (Groups)</td>
<td>Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.</td>
<td>Patient Characteristics</td>
<td>Disease</td>
</tr>
<tr>
<td>-------------------</td>
<td>------------------------------------------</td>
<td>--------------</td>
<td>-----------------------------------------------------------------------------------------------</td>
<td>--------------------------------------</td>
<td>---------------------------------------------------------------------</td>
<td>------------------------</td>
<td>---------</td>
</tr>
<tr>
<td>Sancho, 2017</td>
<td>Observational Prospective in Spain, 01/1/2013 to 12/31/2015</td>
<td>High ROB</td>
<td>previous pulmonary/airway disease, rapidly progressing disease with survival expectancy &lt;1 month, severe frontotemporal dementia, NIPPV tolerance &lt;4 consecutive hour/night.</td>
<td>BPAP ST</td>
<td>BPAP - FDA approved VPAP-III or VPAP-IV plus automatic ventilatory signal analysis (Reslink); Resmed (Sydney, Australia)</td>
<td>82 Patients aged 63.80±110.65, 24.4% female</td>
<td>NMD</td>
</tr>
<tr>
<td>Sanjuan-López, 2014</td>
<td>Observational Retrospective in Spain, 01/01/2000 to 12/31/2010</td>
<td>High ROB</td>
<td>Inclusion: ALS (Escorial criteria), hospital admission Exclusion: lung disease, &lt;1 year life expectancy, NIV use &lt;4 consecutive hours/night, slow disease progression (&gt;3 yrs), severe frontotemporal dementia</td>
<td>HMV (volume assist control ventilation) in no/mild bulbar</td>
<td>HMV – FDA approved Vivo 50; Breas Medical (Molndal, Sweden) Trilogy 100; Philips Respironics (Madrid, Spain)</td>
<td>105 patients aged 64.05±9.11, 53% female</td>
<td>NMD</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>No device in no/mild bulbar</td>
<td>No PAP</td>
<td>15 patients aged 64.05±9.11, 53% female</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>No device in moderate/severe bulbar</td>
<td>No PAP</td>
<td>6 patients aged 66.05±10.27, 70% female</td>
<td></td>
</tr>
</tbody>
</table>

D-18
<table>
<thead>
<tr>
<th>Author, Year</th>
<th>Study Country, Study Design, Study Period</th>
<th>Risk of Bias</th>
<th>Inclusion / Exclusion Criteria</th>
<th>Intervention and comparisons (Groups)</th>
<th>Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.</th>
<th>Patient Characteristics</th>
<th>Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Schonhofer, 2001&lt;sup&gt;13&lt;/sup&gt;</td>
<td>Observational Prospective in Germany</td>
<td>High ROB</td>
<td>Inclusion: Chronic respiratory failure from thoracic disease &amp; hypercapnic, clinically stable, no significant difference in blood gas analysis parameters Exclusion: Rapidly progressive NMD, OHS, COPD, acute RF, severe acidosis</td>
<td>Mixed: HMV (volume assist control ventilation) with change to BPAP ST if not tolerated</td>
<td>HMV - FDA approved Drager EV 800; Drager (Lubeck, Germany) or PLV 100 (Respironics; Murrysville, PA) (either in volume controlled ventilation). If either could not be tolerated by patient, then BP-T (in pressure controlled ventilation); Respironics (Murrysville, USA) was used</td>
<td>Standard care without HMV No HMV</td>
<td>10 Patients aged 53.5±8.2, 50% female</td>
</tr>
<tr>
<td>Sin, 2007&lt;sup&gt;16&lt;/sup&gt;</td>
<td>RCT in Canada, Moderate ROB</td>
<td>Inclusion: Diagnosis of COPD, age≥40 years, &gt;10 pack year smoking history Exclusion: Comorbidities making survival &lt;6 months unlikely, clinical history of left ventricular heart failure, apnea-hypopnea index &gt;20</td>
<td>BPAP NOS + standard care</td>
<td>BPAP - FDA approved VPAP II, ResMed (Sydney, Australia)</td>
<td>Sham BPAP (CPAP 4) Sham Device - FDA approved S7Elite; ResMed (Sydney, Australia)</td>
<td>11 Patients aged 64.1±10.6, 64% female</td>
<td>COPD</td>
</tr>
<tr>
<td>Sivori, 2007&lt;sup&gt;17&lt;/sup&gt;</td>
<td>Observational Prospective in Argentina, 12/1999 to 12/2004</td>
<td>Moderate ROB</td>
<td>Inclusion: Diagnosis of ALS.</td>
<td>BPAP NOS + riluzole</td>
<td>BPAP + riluzole</td>
<td>18 Patients aged 53±15.46, 44.4% female</td>
<td>NMD</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>BPAP NOS</td>
<td>BPAP</td>
<td>11 Patients aged 56.4±15.5, 36.4% female</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>no BPAP, no riluzole</td>
<td>No PAP</td>
<td>42 Patients aged 52.3±11.4, 31% female</td>
<td></td>
</tr>
<tr>
<td>Author, Year</td>
<td>Study Country, Study Design, Study Period</td>
<td>Risk of Bias</td>
<td>Inclusion / Exclusion Criteria</td>
<td>Intervention and comparisons (Groups)</td>
<td>Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.</td>
<td>Patient Characteristics</td>
<td>Disease</td>
</tr>
<tr>
<td>--------------</td>
<td>------------------------------------------</td>
<td>--------------</td>
<td>-------------------------------</td>
<td>--------------------------------------</td>
<td>---------------------------------------------------------------</td>
<td>------------------------</td>
<td>---------</td>
</tr>
<tr>
<td>Struik, 2014&lt;sup&gt;58&lt;/sup&gt;</td>
<td>RCT in the Netherlands, 12/01/2007 to 07/01/2012</td>
<td>Moderate ROB</td>
<td>Inclusion: COPD (GOLD III/IV), &gt;48 hours independence from ventilator support for acute RF, hypercapnia (PaCO2 &gt;6.0 kPa) daytime at rest</td>
<td>BPAP ST</td>
<td>BPAP – FDA approved BiPAP Synchrony; Respironics, Murrysville, USA</td>
<td>101 Patients aged 63.92±8.6, 59% female</td>
<td>COPD</td>
</tr>
<tr>
<td>Tsolaki, 2008&lt;sup&gt;39&lt;/sup&gt;</td>
<td>Observational Prospective in Greece, 09/2005 to 12/2006</td>
<td>High ROB</td>
<td>Inclusion: Age ≤75 years, smoking history &gt;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.</td>
<td>BPAP ST</td>
<td>BPAP - FDA approved VPAP III ST; ResMed (Sydney, Australia)</td>
<td>24 Patients aged 65.2±8.9, 29.2% female</td>
<td>COPD</td>
</tr>
<tr>
<td>Vasquez, 2017&lt;sup&gt;40&lt;/sup&gt;</td>
<td>Observational Retrospective in USA, 01/1/2009 to 10/31/2014</td>
<td>Moderate ROB</td>
<td>Inclusion: At least 2 COPD claims, age ≥40 years, continuous enrollment 12 month prior &amp; 6 months after claim</td>
<td>BPAP NOS</td>
<td>BPAP</td>
<td>9,156 Patients, 35.8% female</td>
<td>COPD</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>CPAP NOS</td>
<td>CPAP</td>
<td>39,385 Patients, 45.1% female</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>NIPPV on HMV NOS</td>
<td>NIPPV on HMV</td>
<td>315 Patients, 48.9% female</td>
<td></td>
</tr>
<tr>
<td>Vitacca, 2017&lt;sup&gt;41&lt;/sup&gt;</td>
<td>Observational Retrospective in Italy, 2008-2013</td>
<td>Moderate ROB</td>
<td>Inclusion: ALS NOS admitted to hospital, NIPPV use Exclusion: dementia confirmed by Mini-Mental State Examination score &lt;20, refusal of NIPPV</td>
<td>HMV/BPAP mix started in FVC&lt;80% (early) HMV/BPAP mix started in FVC&lt;80% (late)</td>
<td>HMV/BPAP mix</td>
<td>65 patients aged 62.62±11.34, 30.77% female</td>
<td>NMD</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>129 patients aged 64.66±11.33, 48.06% female</td>
<td></td>
</tr>
<tr>
<td>Author, Year</td>
<td>Study Country, Study Design, Study Period</td>
<td>Risk of Bias</td>
<td>Inclusion / Exclusion Criteria</td>
<td>Intervention and comparisons (Groups)</td>
<td>Device used (HMV, CPAP, BPAP) manufacturer, brand name, model no.</td>
<td>Patient Characteristics</td>
<td>Disease</td>
</tr>
<tr>
<td>--------------</td>
<td>------------------------------------------</td>
<td>-------------</td>
<td>--------------------------------</td>
<td>-------------------------------------</td>
<td>------------------------------------------------------------</td>
<td>------------------------</td>
<td>---------</td>
</tr>
<tr>
<td>Windisch, 2006&lt;sup&gt;22&lt;/sup&gt;</td>
<td>Observational in Germany</td>
<td>Moderate ROB</td>
<td>Inclusion: Stable disease hospitalized for establishing NIPPV, matched controls Exclusion: Acute RF, signs of respiratory infection, intubated or tracheotomised previously in life, established on other ventilatory support prior to admission</td>
<td>HMV (pressure controlled ventilation)</td>
<td>HMV - FDA approved PV401; Breas Medical AB (Moelnycke, Sweden)</td>
<td>6 Patients aged 55.2±10.0, 16.7% female</td>
<td>COPD</td>
</tr>
<tr>
<td>Zhou, 2017&lt;sup&gt;63&lt;/sup&gt;</td>
<td>RCT in China, 10/01/2015 to 05/31/2016</td>
<td>High ROB</td>
<td>Inclusion: Clinically stable, stage III/IV flow limitation &amp; chronic hypercapnic, age &gt; 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.</td>
<td>BPAP ST</td>
<td>BPAP – Not FDA approved Flexo ST 30 NIV; Curative Co. (SuZhou, China)</td>
<td>57 Patients aged 66.91±7.1, 36.8% female</td>
<td>COPD</td>
</tr>
</tbody>
</table>

Note: ± denotes standard deviation.

Appendix E. Risk of Bias

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

<table>
<thead>
<tr>
<th>Author, Year</th>
<th>Sequence Generation</th>
<th>Allocation Concealment</th>
<th>Blinding of Participants, Personnel</th>
<th>Blinding of Outcome Assessors</th>
<th>Incomplete Outcome Data</th>
<th>Selective Outcome Reporting</th>
<th>Other Sources of Bias</th>
<th>Overall RoB</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bertella, 2017</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>High ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
</tr>
<tr>
<td>Bhatt, 2013</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>High ROB</td>
<td>Unclear</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>High ROB</td>
<td>High ROB</td>
</tr>
<tr>
<td>Borel, 2011</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>High ROB</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>High ROB</td>
<td>High ROB</td>
<td>High ROB</td>
</tr>
<tr>
<td>Bourke, 2006</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>High ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>Low ROB</td>
<td>High ROB</td>
</tr>
<tr>
<td>Casanova, 2000</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>High ROB</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>Unclear</td>
<td>Unclear</td>
<td>High ROB</td>
</tr>
<tr>
<td>Cheung, 2010</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>High ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>High ROB</td>
</tr>
<tr>
<td>Chiang, 2003</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>High ROB</td>
<td>Unclear</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>Low ROB</td>
<td>Moderate ROB</td>
</tr>
<tr>
<td>Clini, 2002</td>
<td>Unclear</td>
<td>Low ROB</td>
<td>High ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>High ROB</td>
<td>High ROB</td>
</tr>
<tr>
<td>De Backer, 2011</td>
<td>Unclear</td>
<td>Unclear</td>
<td>Unclear</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>Low ROB</td>
<td>Moderate ROB</td>
<td>Moderate ROB</td>
</tr>
<tr>
<td>Dreher, 2010</td>
<td>Unclear</td>
<td>Unclear</td>
<td>High ROB</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>High ROB</td>
<td>High ROB</td>
<td>High ROB</td>
</tr>
<tr>
<td>Duiverman, 2008, 2011</td>
<td>Unclear</td>
<td>Low ROB</td>
<td>High ROB</td>
<td>Unclear</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Moderate ROB</td>
<td>Moderate ROB</td>
</tr>
<tr>
<td>Duiverman, 2017</td>
<td>Unclear</td>
<td>Unclear</td>
<td>High ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>High ROB</td>
<td>High ROB</td>
<td>High ROB</td>
</tr>
<tr>
<td>Funk, 2010</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>High ROB</td>
<td>High ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Moderate ROB</td>
<td>Moderate ROB</td>
</tr>
<tr>
<td>Garrod, 2000</td>
<td>Unclear</td>
<td>Low ROB</td>
<td>High ROB</td>
<td>Unclear</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>Unclear</td>
<td>High ROB</td>
</tr>
<tr>
<td>Gay, 1996</td>
<td>Unclear</td>
<td>Unclear</td>
<td>High ROB</td>
<td>High ROB</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>Unclear</td>
<td>High ROB</td>
</tr>
<tr>
<td>Hazenberg, 2014</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>High ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Moderate ROB</td>
<td>Moderate ROB</td>
</tr>
<tr>
<td>Howard, 2016</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>High ROB</td>
<td>Moderate ROB</td>
</tr>
<tr>
<td>Köhnlein, 2014</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>High ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>High ROB</td>
<td>High ROB</td>
</tr>
<tr>
<td>Marquez-Martín, 2014</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>High ROB</td>
<td>Unclear</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>Low ROB</td>
<td>Moderate ROB</td>
</tr>
<tr>
<td>Masa, 2015</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>High ROB</td>
<td>Unclear</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>Low ROB</td>
<td>Moderate ROB</td>
</tr>
<tr>
<td>McEvoy, 2009</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>High ROB</td>
<td>Unclear</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Moderate ROB</td>
<td>Moderate ROB</td>
</tr>
<tr>
<td>Murphy, 2012</td>
<td>Unclear</td>
<td>Low ROB</td>
<td>High ROB</td>
<td>Unclear</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Moderate ROB</td>
</tr>
<tr>
<td>Murphy, 2017</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>High ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>High ROB</td>
<td>High ROB</td>
</tr>
<tr>
<td>Oscroft, 2014</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>High ROB</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>High ROB</td>
<td>High ROB</td>
<td>High ROB</td>
</tr>
<tr>
<td>Piper, 2008</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>Unclear</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
</tr>
<tr>
<td>Author, Year</td>
<td>Representativeness of the Study Population</td>
<td>Ascertainment of Exposure</td>
<td>Assessment of Outcome</td>
<td>Adequate of Followup</td>
<td>Conflict of Interest</td>
<td>Overall RoB</td>
<td></td>
<td></td>
</tr>
<tr>
<td>--------------------</td>
<td>------------------------------------------</td>
<td>----------------------------</td>
<td>----------------------</td>
<td>----------------------</td>
<td>---------------------</td>
<td>-------------</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sin, 2007^6</td>
<td>High ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>Moderate ROB</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Struik, 2014^5</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>Moderate ROB</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Zhou, 2017^1</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>High ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>High ROB</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

ROB: Risk of Bias
<table>
<thead>
<tr>
<th>Author, Year</th>
<th>Representativeness of the Study Population</th>
<th>Ascertainment of Exposure</th>
<th>Assessment of Outcome</th>
<th>Adequate of Followup</th>
<th>Conflict of Interest</th>
<th>Overall RoB</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pinto, 2010</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>High ROB</td>
<td>High ROB</td>
</tr>
<tr>
<td>Salturk, 2015</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
</tr>
<tr>
<td>Sancho, 2014</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
</tr>
<tr>
<td>Sancho, 2017</td>
<td>High ROB</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>Unclear</td>
<td>Low ROB</td>
<td>High ROB</td>
</tr>
<tr>
<td>Sanjuan-López, 2014</td>
<td>Low ROB</td>
<td>High ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>High ROB</td>
</tr>
<tr>
<td>Schonhofer, 2001</td>
<td>High ROB</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>Unclear</td>
<td>Unclear</td>
<td>High ROB</td>
</tr>
<tr>
<td>Sivori, 2007</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>High ROB</td>
<td>Unclear</td>
<td>Low ROB</td>
<td>Moderate ROB</td>
</tr>
<tr>
<td>Tsolaki, 2008</td>
<td>High ROB</td>
<td>Low ROB</td>
<td>High ROB</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>High ROB</td>
</tr>
<tr>
<td>Vasquez, 2017</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>Unclear</td>
<td>Unclear</td>
<td>Moderate ROB</td>
</tr>
<tr>
<td>Vitacca, 2017</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>Unclear</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Moderate ROB</td>
</tr>
<tr>
<td>Windisch, 2006</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Low ROB</td>
<td>Unclear</td>
<td>Unclear</td>
<td>Moderate ROB</td>
</tr>
</tbody>
</table>

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>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Device/mode</th>
<th>Patient characteristics to start or continue device</th>
<th>Laboratory characteristics to start or continue device</th>
<th>Device titration</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Murphy, 2017</strong>&lt;sup&gt;13&lt;/sup&gt; RCT</td>
<td>BPAP ST</td>
<td>-COPD (FEV1 &lt; 50%) -NIPPV during hospital admission</td>
<td>-PaCO2 &gt;53 mmHg -PaO2 &lt;55 mmHg or PaO2 &lt; 60 mmHg with polycythemia, pulmonary hypertension or cor pulmonale -ST 90&lt;30% -pH &gt;7.30 (daytime, room air)</td>
<td>“High pressure ventilation strategy” titrated during polysomnography</td>
</tr>
<tr>
<td><strong>Oscroft, 2014</strong>&lt;sup&gt;16&lt;/sup&gt; RCT</td>
<td>BPAP IVAPS versus BPAP ST</td>
<td>-COPD (FEV1 &lt; 50%) -Mixed stable disease or following AECOPD</td>
<td>-PaCO2 &gt;7 kPa (53 mmHg) -pH &gt;7.35 or PtcCO2 &gt;9 kPa (68 mmHg) (daytime)</td>
<td>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 5cmH2O. (Titration took on average 5.2 [SD 2.8] days)</td>
</tr>
<tr>
<td><strong>Paone, 2014</strong>&lt;sup&gt;17&lt;/sup&gt; Prospective Observational</td>
<td>BPAP ST</td>
<td>-COPD (FEV1 &lt; 50%) -NIPPV during hospital admission</td>
<td>-PaCO2 &gt; 50 mmHg (after awakening from a night without NIPPV)</td>
<td>Maximum tolerated IPAP to target tidal volume of 6 mL/kg (measured body weight). EPAP set at 2-8 cmH2O. Backup rate set at 12 breaths/min.</td>
</tr>
<tr>
<td><strong>Galli, 2014</strong>&lt;sup&gt;18&lt;/sup&gt; Retrospective Observational</td>
<td>BPAP NOS</td>
<td>-COPD (ICD-9) -NIPPV during hospital admission</td>
<td>-PaCO2 &gt; 45 mmHg</td>
<td></td>
</tr>
</tbody>
</table>

Table F.1. COPD - New initiation of home device
<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Device/mode</th>
<th>Patient characteristics to start or continue device</th>
<th>Laboratory characteristics to start or continue device</th>
<th>Device titration</th>
</tr>
</thead>
</table>
| Bhatt, 2013<sup>7</sup> 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<sup>11,22</sup>, 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<sup>15</sup> Retrospective 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<sup>11</sup> 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. |
| Casanova, 2000<sup>9</sup> 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<sup>18</sup> RCT | BPAP S | -COPD (FEV1 <50%)  
- Stable (no AECOPD in prior 4 weeks)  
- exercise intolerance due to dyspnea | Maximum tolerated IPAP and EPAP. (Titrated over 1 week). | |
<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Device(mode)</th>
<th>Patient characteristics to start or continue device</th>
<th>Laboratory characteristics to start or continue device</th>
<th>Device titration</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clin, 1998Prospective Observational</td>
<td>BPAP ST</td>
<td>-COPD (FEV1&lt;50%)&lt;br&gt;-Stable (no AECOPD in prior 4 weeks)&lt;br&gt;-LTOT ≥12 months&lt;br&gt;-≥1 ICU admission due to AECOPD in prior 2 years</td>
<td>-PaCO2 &gt;6 kPa (45 mmHg)&lt;br&gt;-pH &gt;7.35&lt;br&gt;-PaO2 &lt;8 kPa (60 mmHg) (daytime, room air, rest)</td>
<td>Minimal IPAP to achieve an expiratory tidal volume &gt; 8ml/kg. EPAP was in order not to overcome the intrinsic PEEP. Backup rate set at 10 breaths/min.</td>
</tr>
<tr>
<td>Clin, 1996Prospective Observational</td>
<td>BPAP ST</td>
<td>-COPD (FEV1 30-49%)&lt;br&gt;-Stable (no AECOPD in prior 4 weeks)&lt;br&gt;-LTOT ≥18 months&lt;br&gt;-≥1 hospital admission due to AECOPD in prior 18 months</td>
<td>-PaCO2 &gt;6.7 kPa (50 mmHg)</td>
<td>Minimal IPAP to achieve an expiratory tidal volume &gt; 8ml/kg. Rate set at 10 breaths/min (Titration over 15 days in hospital).</td>
</tr>
<tr>
<td>Zhou, 2017RCT</td>
<td>BPAP ST</td>
<td>-COPD (FEV1&lt;50%)&lt;br&gt;-Stable (no AECOPD in prior 4 weeks)&lt;br&gt;-Hypercapnia (daytime, rest)</td>
<td>-PaCO2 &gt;45 mmHg&lt;br&gt;-PaO2 &lt; 60 mmHg</td>
<td>Maximum tolerated IPAP (≥ 10 cmH2O). EPAP set at 4 cmH2O. Backup rate set at 16 breaths/min.</td>
</tr>
<tr>
<td>Marquez-Martin, 2014RCT</td>
<td>BPAP ST</td>
<td>-COPD (FEV1&lt;50%)&lt;br&gt;-Stable (no AECOPD in prior 3 months)&lt;br&gt;-Hypercapnia (daytime, rest) NOS</td>
<td>-PaCO2 ≥ 7 kPa (53 mmHg)&lt;br&gt;-pH ≥ 7.35 (daytime, rest)</td>
<td>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.</td>
</tr>
<tr>
<td>Köhnlein, 2014RCT</td>
<td>BPAP ST</td>
<td>-COPD (FEV1&lt;30%)&lt;br&gt;-Stable (no AECOPD in prior 4 weeks)&lt;br&gt;-Hypercapnia (daytime, rest)</td>
<td>-PaCO2 &gt;45 mmHg on day 5-12 of hospitalization</td>
<td>Targeted to reduce baseline PaCO2 by ≥ 20% or achieve PaCO2 &lt;6.5 kPa (49 mmHg).</td>
</tr>
<tr>
<td>De Backer, 2011RCT</td>
<td>BPAP NOS</td>
<td>-COPD (FEV1&lt;50%)&lt;br&gt;-AECOPD requiring hospitalization</td>
<td>-PaCO2 &gt;45 mmHg</td>
<td>Targeted SaO2 &gt;90% during 90% of time and reduction in PaCO2 ≥ 5% in 1 hour.</td>
</tr>
<tr>
<td>Dreher, 2010RCT</td>
<td>HMV (pressure assist/control) versus HMV (PSV ST)</td>
<td>-COPD (Gold stage IV)&lt;br&gt;-Stable (no current AECOPD).</td>
<td>-PaCO2 &gt;45 mmHg (daytime) and PaCO2 &gt;50 mmHg (nocturnal)</td>
<td>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.</td>
</tr>
<tr>
<td>McEvoy, 2009RCT</td>
<td>BPAP S</td>
<td>-COPD (FEV1&lt;50% or &lt;1.5L)&lt;br&gt;-Stable disease&lt;br&gt;-LTOT for ≥3 months</td>
<td>-PaCO2 &gt;46 mmHg (at least twice in prior 6 months during stability)</td>
<td>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.)</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Device/mode</td>
<td>Patient characteristics to start or continue device</td>
<td>Laboratory characteristics to start or continue device</td>
<td>Device titration</td>
</tr>
<tr>
<td>---------------------------</td>
<td>-------------</td>
<td>------------------------------------------------------</td>
<td>------------------------------------------------------</td>
<td>-----------------</td>
</tr>
<tr>
<td>Tsolaki, 2008&lt;sup&gt;29&lt;/sup&gt; Prospective Observational</td>
<td>BPAP ST</td>
<td>-COPD (FEV1 &lt;50%) -Stable (no AECOPD in prior 4 weeks)</td>
<td>-PaCO2&gt;50 mmHg -PaO2&lt;60 mmHg (room air)</td>
<td>IPAP and EPAP to target patient comfort, decreased accessory muscle use, lower respiratory rate, and decrease in PaCO2 &gt;5% after 1 hour. (Titration in hospital).</td>
</tr>
<tr>
<td>Windisch, 2006&lt;sup&gt;42&lt;/sup&gt; Observational</td>
<td>HMV with pressure controlled ventilation (PCV) mode</td>
<td>-COPD NOS -Stable (no worsening symptoms in prior 2 weeks, respiratory rate &lt;30 breaths/minute, no signs of current respiratory infection, no changes in symptoms or medications in prior 3 months) -NIPPV in hospital admission</td>
<td>-pH≥7.35</td>
<td>Maximum tolerated IPAP to target a maximum decrease in PaCO2</td>
</tr>
<tr>
<td>Gay, 1996&lt;sup&gt;19&lt;/sup&gt; RCT</td>
<td>BPAP ST versus sham CPAP lowest setting</td>
<td>-COPD (FEV1 &lt; 40%) -Stable disease</td>
<td>-PaCO2 &gt; 45 mmHg (daytime, rest)</td>
<td>IPAP set to 10 cmH2O. EPAP set to lowest possible. Backup rate to target patient comfort.</td>
</tr>
<tr>
<td>Gad, 2014&lt;sup&gt;36&lt;/sup&gt; Prospective Observational</td>
<td>BPAP ST</td>
<td>-COPD (FEV1 &lt; 50%) -Stable (no AECOPD in prior 4 weeks) PaCO2&gt;50 mmHg</td>
<td>-PaCO2 &gt;50 mmHg -pH &gt; 7.35 (daytime)</td>
<td>Maximum tolerated IPAP (targeting 15-20 cmH2O). EPAP 3-6 cmH2O. (Titration occurred in hospital over 2-3 day period.)</td>
</tr>
<tr>
<td>Sin, 2007&lt;sup&gt;35&lt;/sup&gt; RCT</td>
<td>BPAP NOS versus sham CPAP 4 cmH2O</td>
<td>-COPD (FEV1 NOS) -Stable disease</td>
<td>-PaCO2 &gt;52.5mmHg -pH&lt;7.35 (recurrent acidosis)</td>
<td>Maximum tolerated IPAP (maximum of 20 cmH2O). EPAP set at 4 cmH2O.</td>
</tr>
<tr>
<td>Heinemann, 2011&lt;sup&gt;11&lt;/sup&gt;</td>
<td>BPAP (pressure controlled ventilation)</td>
<td>-COPD (FEV1 NOS) -invasive mechanical ventilation for AECOPD, pneumonia, or postoperative respiratory failure -prolonged weaning from invasive mechanical ventilation</td>
<td>-PaCO2&gt;55mmHg or -pH&lt;7.35 (recurrent acidosis)</td>
<td>Maximum tolerated IPAP to achieve maximum reduction in PaCO2.</td>
</tr>
<tr>
<td>Budweiser, 2007&lt;sup&gt;7&lt;/sup&gt;</td>
<td>BPAP (pressure controlled ventilation)</td>
<td>-COPD (FEV1&lt;50%) -Stable and unstable disease</td>
<td>-PaCO2&gt;55mmHg -pH&lt;7.35 (recurrent acidosis)</td>
<td>Maximum tolerated IPAP with goal decrease in PaCO2 &gt;5% after 1 hour; and nocturnal SaO2&gt;90% for 90% of time. (Titration in hospital).</td>
</tr>
<tr>
<td>Clini, 2002&lt;sup&gt;14&lt;/sup&gt;</td>
<td>BPAP ST</td>
<td>-COPD (FEV1 NOS) -Stable (no AECOPD in prior 4 weeks)</td>
<td>-PaCO2 &gt;6.6 kPa (50 mmHg) -pH&gt;7.35 (daytime, room air)</td>
<td>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.</td>
</tr>
<tr>
<td>Struik, 2014&lt;sup&gt;38&lt;/sup&gt;</td>
<td>BPAP ST</td>
<td>-COPD (FEV1 &lt;50%) -NIPPV or invasive mechanical ventilation in hospital admission</td>
<td>-PaCO2 &gt;6 kPa (45 mmHg)</td>
<td>Maximum tolerated IPAP to achieve normal PaCO2. Backup respiratory rate</td>
</tr>
<tr>
<td>Durao, 2018&lt;sup&gt;34&lt;/sup&gt;</td>
<td>HMV/BPAP mix</td>
<td>-COPD (NOS) -AECOPD</td>
<td></td>
<td>Maximum tolerated IPAP to achieve maximum reduction in PaCO2. Backup respiratory rate</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Device/mode</td>
<td>Patient characteristics to start or continue device</td>
<td>Laboratory characteristics to start or continue device</td>
<td>Device titration</td>
</tr>
<tr>
<td>---------------------------</td>
<td>-------------</td>
<td>-----------------------------------------------------</td>
<td>-----------------------------------------------------</td>
<td>-----------------</td>
</tr>
<tr>
<td>Salturk, 2015&lt;sup&gt;51&lt;/sup&gt; Retrospective Observational</td>
<td>BPAP ST</td>
<td>-COPD (FEV1 not specified) -post ICU admission -home NIPPV ≥ 4 hours/day</td>
<td></td>
<td>IPAP titrated to achieve “desired tidal volume” (maximum 30 mbar)</td>
</tr>
<tr>
<td>Blankenburg, 2017&lt;sup&gt;4&lt;/sup&gt;</td>
<td>HMV (pressure controlled ventilation or pressure support ventilation)</td>
<td>-COPD (FEV1 NOS) -Stable (no AECOPD in prior 2 weeks)</td>
<td>-PaCO2&gt;7.0kPa (53mmHg) -pH&gt;7.35</td>
<td>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.</td>
</tr>
<tr>
<td>Duiverman, 2017&lt;sup&gt;23&lt;/sup&gt;</td>
<td>HMV /BPAP mix (pressure controlled ventilation versus pressure support ventilation)</td>
<td>-COPD (FEV1 NOS) -Stable (no AECOPD in prior 4 weeks) -≥ 2 AECOPD with acute hypercapnic respiratory failure (pH&lt;7.35) per year</td>
<td>-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)</td>
<td>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 breaths/minute.</td>
</tr>
<tr>
<td>Duiverman, 2017&lt;sup&gt;23&lt;/sup&gt;</td>
<td>HMV/BPAP mix</td>
<td>-COPD (NOS) -Stable (no current AECOPD)</td>
<td>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 (&gt;12 hours/day) or intolerant to IPAP &gt;25 cmH2O)</td>
<td></td>
</tr>
<tr>
<td>Blankenburg, 2017&lt;sup&gt;4&lt;/sup&gt;</td>
<td>HMV (pressure controlled ventilation or pressure support ventilation)</td>
<td>-COPD (FEV1 NOS) -Stable (no AECOPD in prior 2 weeks)</td>
<td>-PaCO2&gt;7.0kPa (53mmHg) -pH&gt;7.35</td>
<td>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.</td>
</tr>
<tr>
<td>Salturk, 2015&lt;sup&gt;51&lt;/sup&gt;</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Device/mode or</th>
<th>Patient characteristics to start or continue device</th>
<th>Laboratory characteristics to start or continue device</th>
<th>Device titration</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hitzl, 2009&lt;sup&gt;17&lt;/sup&gt; Prospective Observational</td>
<td>HMV (pressure cycled assist control mode)</td>
<td>- Stable (no current AECOPD)</td>
<td>PaCO2 &gt; 45 mmHg (stable, measured immediately after awakening from a night without mechanical ventilation)</td>
<td>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.</td>
</tr>
<tr>
<td>Funk, 2010&lt;sup&gt;25&lt;/sup&gt; RCT</td>
<td>BPAP NOS for 6 months</td>
<td>- COPD &quot;standard criteria&quot; NOS</td>
<td>- PaCO2 &gt; 45 mmHg (stable, measured immediately after awakening from a night without mechanical ventilation)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>BPAP NOS more than 6 months</td>
<td>- AECOPD requiring NIPPV or invasive ventilation</td>
<td>- chronic nocturnal NIPPV use at home for ≥ 6 months</td>
<td></td>
</tr>
<tr>
<td></td>
<td>BPAP NOS versus CPAP NOS versus HMV NOS</td>
<td>- COPD (ICD-9)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Vásquez, 2017&lt;sup&gt;60&lt;/sup&gt; Retrospective cohort</td>
<td>BPAP NOS versus CPAP NOS versus HMV NOS</td>
<td>- COPD (ICD-9)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>


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

<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Device/mode</th>
<th>Patient characteristics to start or continue device</th>
<th>Laboratory characteristics to start or continue device</th>
<th>Device titration</th>
</tr>
</thead>
<tbody>
<tr>
<td>Salturk, 2015&lt;sup&gt;11&lt;/sup&gt; Retrospective Observational</td>
<td>BPAP ST</td>
<td>Kyphoscoliosis NOS</td>
<td>IPAP titrated to achieve “desired tidal volume” (maximum 30 mbar)</td>
<td></td>
</tr>
<tr>
<td>Domenéech-Clar, 2003&lt;sup&gt;19&lt;/sup&gt; Prospective Observational</td>
<td>BPAP NOS</td>
<td>- Kyphoscoliosis or fibrothorax or thoracoplasty</td>
<td>- PaCO2 &gt; 45 mmHg or - FVC &lt; 40% or - MIP &lt; 60 cm H2O or - nocturnal SaO2 &lt; 88% for ≥ 5 consecutive minutes</td>
<td>IPAP increased (minimum 10 cmH2O) to target a normal PaCO2 or a decrease of at least 10 mmHg. (Titration occurred in hospital)</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Device/mode</td>
<td>Patient characteristics to start or continue device</td>
<td>Laboratory characteristics to start or continue device</td>
<td>Device titration</td>
</tr>
<tr>
<td>----------------------------</td>
<td>-------------</td>
<td>-----------------------------------------------------</td>
<td>------------------------------------------------------</td>
<td>-----------------</td>
</tr>
<tr>
<td>Nauffal, 2002&lt;sup&gt;44&lt;/sup&gt; Prospective Observational</td>
<td>BPAP NOS</td>
<td>-Kyphoscoliosis NOS -stable (no infection in past 3 months) -symptoms (fatigue, dyspnea, morning headache)</td>
<td>-PaCO2 &gt;45 mmHg or -FVC &lt;50% or -MIP &lt;60 cm H2O or -SaO2 &lt; 88% for ≥ 5 consecutive minutes by nocturnal oximetry</td>
<td>IPAP and EPAP titrated to maximize change of arterial blood gases. (Titration occurred in hospital)</td>
</tr>
<tr>
<td>Masa, 2000&lt;sup&gt;37&lt;/sup&gt; Prospective Observational</td>
<td>HMV (volume controlled ventilation with change to pressure controlled ventilation if volume could not be tolerated)</td>
<td>-Kyphoscoliosis (scoliosis angle [Cobb] &gt;90 degrees -FEV1/FVC ≥65% -Apnea-hypopnea index ≤ 20 events/hour</td>
<td>-PaCO2 &gt;47 mmHg for at least 3 months</td>
<td>Ventilator parameters adjusted to target maximum reduction in PaCO2 as well as patient tolerance, air leakage, and nocturnal saturation &gt;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)</td>
</tr>
<tr>
<td>Schonhofer, 2001&lt;sup&gt;15&lt;/sup&gt; Prospective Observational</td>
<td>HMV (volume controlled ventilation with change to BPAP ST if volume could not be tolerated)</td>
<td>-TRD (post-TB or scoliosis NOS) -Stable disease (stable PaCO2, no hospital admission in prior 1 month)</td>
<td>-absence of severe acidosis -PaCO2 45-55 mmHg</td>
<td></td>
</tr>
</tbody>
</table>


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

<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Device/mode</th>
<th>Patient characteristics to start or continue device</th>
<th>Laboratory characteristics to start or continue device</th>
<th>Device titration</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hitzl, 2009&lt;sup&gt;13&lt;/sup&gt; Prospective Observational</td>
<td>HMV (pressure cycled assist control mode) in restrictive thoracic disease</td>
<td>-TRD NOS -HMV initiated ≥3 months</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Buyse, 2003&lt;sup&gt;3&lt;/sup&gt; Retrospective Observational</td>
<td>HMV (volume or pressure cycled ventilator NOS) + oxygen</td>
<td>-Kyphoscoliosis NOS -NIPPV use NOS</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

HMV: Home Mechanical Ventilation, NIPPV: Noninvasive positive pressure Ventilation, NOS: Not otherwise Specified, TRD: Thoracic Restrictive Disorder
<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Device/mode</th>
<th>Patient characteristics to start or continue device</th>
<th>Laboratory characteristics to start or continue device</th>
<th>Device titration</th>
</tr>
</thead>
</table>
| Sanjuan-López, 2014<sup>41</sup> Retrospective Observational | HMV (PSV or ST) started after outpatient pulmonary evaluation | -ALS (El Escorial criteria)  
-chronic respiratory failure by pulmonologist | | Increase in IPAP to target symptom relief. Monitored with daytime and nocturnal oximetry and blood gases. |
| | HMV (PSV or ST) started in an emergency situation without prior outpatient pulmonary evaluation | | | |
| Pinto, 1995<sup>48</sup> Prospective Observational | BPAP NOS | -ALS (El Escorial criteria)  
-bulbar features | | |
| Doménech-Clar, 2003<sup>19</sup> Prospective Observational | BPAP NOS | -NMD NOS  
-stable (no infection in past 3 months)  
symptoms (fatigue, dyspnea, morning headache) | -PaCO2 >45 mmHg  
-FVC <50%  
-MIP <60 cm H2O  
-nocturnal SaO2 < 88% for ≥ 5 consecutive minutes | IPAP increased (minimum 10 cmH2O) to target a normal PaCO2 or a decrease of at least 10 mmHg. (Titration occurred in hospital) |
| Nauffal, 2002<sup>14</sup> Prospective Observational | BPAP NOS | -NMD NOS  
-stable (no infection in past 3 months)  
symptoms (fatigue, dyspnea, morning headache) | -PaCO2 >45 mmHg  
-FVC <50%  
-MIP <60 cm H2O  
-SaO2 < 88% for ≥ 5 consecutive minutes by nocturnal oximetry | IPAP and EPAP titrated to maximize change of arterial blood gases. (Titration occurred in hospital) |
<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Device/mode</th>
<th>Patient characteristics to start or continue device</th>
<th>Laboratory characteristics to start or continue device</th>
<th>Device titration</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sancho, 2014 &amp; Retrospective Observational</td>
<td>HMV (volume cycled) versus BPAP ST</td>
<td>-ALS NOS -symptoms (fatigue, dyspnea, orthopnea, morning headache)</td>
<td>-PaCO2 &gt;45 mmHg or -FVC &lt;50% or -MIP &lt;60 cm H2O or -SaO2 &lt; 88% for ≥ 5 consecutive minutes by nocturnal oximetry</td>
<td>Titration occurred in the hospital.</td>
</tr>
<tr>
<td>Sivori, 2007 &amp; Prospective Observational</td>
<td>BPAP NOS</td>
<td>-ALS (El Escorial criteria) -symptomatic ventilatory impairment (dyspnea, morning headache, fatigue)</td>
<td>-PaCO2 &gt;45 mmHg or -FVC &lt;50% or -MIP &lt;60 cm H2O or -nocturnal SaO2 &lt; 88% for ≥ 5 consecutive minutes</td>
<td>IPAP adjusted to maintain SpO2 &gt;92% (ranged 13-25 cmH2O). EPAP set from 5-9 cmH2O.</td>
</tr>
<tr>
<td>Coco, 2006 &amp; Prospective Observational</td>
<td>BPAP ST</td>
<td>-ALS (El Escorial criteria) -symptomatic ventilatory impairment (dyspnea, morning headache, fatigue)</td>
<td>-PaCO2 &gt;45 mmHg or -FVC &lt;50% or -MIP &lt;60 cm H2O or -nocturnal SaO2 &lt; 88% for ≥ 5 consecutive minutes</td>
<td>Maximum tolerated IPAP and EPAP to target patient comfort, leaks, normal PaO2, PaCO2, SpO2, and symptom relief. IPAP started at 8-12 cmH2O and EPAP started at 3-4c cmH2O.</td>
</tr>
<tr>
<td>Bourke, 2006 &amp; RCT</td>
<td>BPAP ST</td>
<td>-ALS NOS</td>
<td>-Orthopnea with Pimax &lt;60% or -symptomatic daytime hypercapnia</td>
<td>IPAP and EPAP adjusted to optimize daytime arterial blood gases, nocturnal oximetry breathing room air, and increased use/duration of device.</td>
</tr>
<tr>
<td>Vitacca, 2017 &amp; HMV/BPAP mix started in FVC≥80% (early)</td>
<td>-ALS NOS -FVC≥80%</td>
<td>-ALS NOS -FVC&lt;80%</td>
<td>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.</td>
<td></td>
</tr>
<tr>
<td>Sancho, 2017 &amp; HMV (volume assist control ventilation)</td>
<td>-ALS (El Escorial criteria) -symptomatic ventilatory impairment (dyspnea, orthopnea, fatigue, morning headache, daytime hypersomnolence, decreased cognitive function)</td>
<td>-PaCO2 &gt;45 mmHg and -FVC &lt;50% and -nocturnal SaO2 &lt; 90% for ≥ 5% of time</td>
<td>Ventilator adjusted to target PaCO2&lt;45mmHg, nocturnal SaO2 &lt; 90% for &lt;5% of time, optimize comfort, prevent air leaks.</td>
<td></td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Device/mode</td>
<td>Patient characteristics to start or continue device</td>
<td>Laboratory characteristics to start or continue device</td>
<td>Device titration</td>
</tr>
<tr>
<td>---------------------------</td>
<td>-------------</td>
<td>----------------------------------------------------</td>
<td>----------------------------------------------------</td>
<td>-----------------</td>
</tr>
</tbody>
</table>
| Bertella, 2017*           | BPAP volume assured pressure support ventilation | -ALS (definite via El Esocirial 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. |


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

<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Device/mode</th>
<th>Patient characteristics to start or continue device</th>
<th>Laboratory characteristics to start or continue device</th>
<th>Device titration</th>
</tr>
</thead>
</table>
| Pinto, 2010* Prospective 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* Retrospective 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

<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Device/mode</th>
<th>Patient characteristics to start or continue device</th>
<th>Laboratory characteristics to start or continue device</th>
<th>Device titration</th>
</tr>
</thead>
</table>
| Howard, 2016\(^{14}\) 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\(^{11}\) Retrospective 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\(^{37}\) Prospective 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\(^{10}\) Prospective 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 (FEV1/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. |
| Masa, 2015\(^{18, 19}\) 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. |
<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Device/mode</th>
<th>Patient characteristics to start or continue device</th>
<th>Laboratory characteristics to start or continue device</th>
<th>Device titration</th>
</tr>
</thead>
<tbody>
<tr>
<td>Borel, 2011&lt;sup&gt;5&lt;/sup&gt; RCT</td>
<td>BPAP ST</td>
<td>-OHS (BMI &gt;30; daytime PaCO2 ≥ 45 mmHg, other causes of hypoventilation ruled out including airway obstruction, scoliosis, cardiac failure, progressive NMD)</td>
<td>-PaCO2 ≥ 45 mmHg (daytime)</td>
<td>(Titration occurred in hospital over 3-4 nights)</td>
</tr>
<tr>
<td>Murphy, 2012&lt;sup&gt;2&lt;/sup&gt; RCT</td>
<td>BPAP (volume assured pressure support ventilation) versus BPAP ST</td>
<td>-OHS (BMI&gt;40, daytime chronic PaCO2 &gt;6 kPa, pH &gt;7.35), absence of other identifiable hypoventilation cause, FEV1/FVC &gt;70%, FVC &lt;70% -Stable disease</td>
<td>-PaCO2 &gt;6 kPa (45 mmHg) -pH &gt;7.35 (daytime)</td>
<td>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)</td>
</tr>
<tr>
<td>Piper, 2008&lt;sup&gt;50&lt;/sup&gt; RCT</td>
<td>BPAP S versus CPAP</td>
<td>-OHS (BMI≥30, PaCO2 ≥ 45 mmHg [awake, stable], absence of another cause for hypercapnia, FEV1/FVC ≥ 70%)</td>
<td>-PaCO2 ≥45 mmHg -pH ≥7.34 (daytime, stable)</td>
<td>Excluded during CPAP titration study: -SaO2 &lt;80% for 10 minutes in absence of apnea -TcCO2 during REM ≥10mmHg -increase in afternoon to morning PaCO2 ≥10mmHg in patients with awake PaCO2 &gt;55 mmHg</td>
</tr>
<tr>
<td>Blankenburg, 2017&lt;sup&gt;4&lt;/sup&gt;</td>
<td>HMV (pressure controlled ventilation or pressure support ventilation)</td>
<td>-OHS (BMI&gt;30, PaCO2 &gt;6.7kPa[50mmHg], symptoms of hypercapnia NOS), absence of another cause for hypercapnia</td>
<td>-PaCO2&gt;7.0kPa (53mmHg) -pH&gt;7.35</td>
<td>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.</td>
</tr>
</tbody>
</table>

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

<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Disease</th>
<th>Device/mode</th>
<th>Patient characteristics to start or continue device</th>
<th>Laboratory characteristics to start or continue device</th>
<th>Device titration</th>
</tr>
</thead>
<tbody>
<tr>
<td>Salturk, 2015¹</td>
<td>Diffuse parenchymal lung disease</td>
<td>BPAP ST</td>
<td>-Diffuse parenchymal lung disease (sequela of TB or bronchiectasis with hypoxemia and hypercapnia)</td>
<td></td>
<td>IPAP titrated to achieve “desired tidal volume” (maximum 30 mbar)</td>
</tr>
</tbody>
</table>

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

<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Disease</th>
<th>Device/mode</th>
<th>Patient characteristics to start or continue device</th>
<th>Laboratory characteristics to start or continue device</th>
<th>Device titration</th>
</tr>
</thead>
<tbody>
<tr>
<td>Benhamou, 1997¹</td>
<td>Diffuse bronchiectasis</td>
<td>HMV (volume cycled)</td>
<td>-Diffuse bronchiectasis</td>
<td></td>
<td>Target PaO2 &gt; 9kPa (67 mmHg) without deterioration in PaCO2.</td>
</tr>
</tbody>
</table>

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

<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Disease</th>
<th>Device/mode</th>
<th>Patient characteristics to start or continue device</th>
<th>Laboratory characteristics to start or continue device</th>
<th>Device titration</th>
</tr>
</thead>
<tbody>
<tr>
<td>Windisch, 2006⁶²</td>
<td>TRD, OHS</td>
<td>HMV with pressure controlled ventilation (PCV) mode</td>
<td>-COPD NOS</td>
<td>-pH≥7.35</td>
<td>Maximum tolerated IPAP to target a maximum decrease in PaCO2</td>
</tr>
<tr>
<td>Hazenberg, 2014³¹</td>
<td>NMD, TRD</td>
<td>HMV (pressure or volume)</td>
<td>-NMD or thoracic cage disorder</td>
<td>-PaCO2 &gt;6.0 kPa (&gt;45 mmHg) (daytime)</td>
<td>Maximum tolerated IPAP to target a target tidal volume of 8-10 ml/kg and a</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Disease</td>
<td>Device/mode</td>
<td>Patient characteristics to start or continue device</td>
<td>Laboratory characteristics to start or continue device</td>
<td>Device titration</td>
</tr>
<tr>
<td>---------------------------</td>
<td>---------</td>
<td>-------------</td>
<td>---------------------------------------------------</td>
<td>---------------------------------------------------</td>
<td>-----------------</td>
</tr>
<tr>
<td>RCT</td>
<td>control started at home</td>
<td>HMV (pressure or volume control) started in the hospital</td>
<td>-NMD or thoracic cage disorder -Stable disease without acute respiratory failure</td>
<td>-PaCO2 &gt; 6.0 kPa (&gt;45 mmHg) (daytime)</td>
<td>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)</td>
</tr>
<tr>
<td>Munoz, 2005&lt;sup&gt;43&lt;/sup&gt; Retrospective Observational</td>
<td>NMD, TRD</td>
<td>HMV volume assist/control mode versus HMV volume control mode</td>
<td>-Hospital admission with chronic hypercapnic respiratory failure to NMD (ALS excluded) or kyphoscoliosis or post TB sequelae</td>
<td>-PaCO2 &gt; 45 mmHg (daytime, stable)</td>
<td>The tidal volume, respiratory frequency, and the I/E ratio were adjusted individually according to tolerance, air leaks, and ventilatory response.</td>
</tr>
<tr>
<td>Chiang, 2003&lt;sup&gt;42&lt;/sup&gt; RCT</td>
<td>COPD, Other</td>
<td>BPAP NOS</td>
<td>-COPD or asthma or bronchiectasis -Hospital readmission due to respiratory cause -Daytime sleepiness or morning headache</td>
<td>-PaCO2 &gt; 50 mmHg (daytime rest) -SpO2 &lt; 88% for more than 5 consecutive minutes while on usual oxygen during polysomnography</td>
<td>IPAP and EPAP and volumes set to target optimal daytime PaCO2</td>
</tr>
</tbody>
</table>

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

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

<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Disease</th>
<th>Inclusion/Exclusion Criteria</th>
<th>Device And Settings Used (Group)</th>
<th>Duration Of Device Use In Home Setting</th>
<th>Conclusion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vasquez, 2017&lt;sup&gt;60&lt;/sup&gt; Retrospective Observational</td>
<td>COPD</td>
<td>Inclusion: COPD (ICD-9); age ≥40 years</td>
<td>1) BPAP NOS &lt;br&gt; 2) CPAP NOS &lt;br&gt; 3) HMV NOS</td>
<td>Longest duration: 6 months</td>
<td>The HMV group had significantly more reduction of mortality than those with CPAP (p&lt;0.001) or BPAP (p&lt;0.001), and more reduction on COPD-related hospitalization than the CPAP group (p=0.01).</td>
</tr>
<tr>
<td>Murphy, 2017&lt;sup&gt;43&lt;/sup&gt;, RCT</td>
<td>COPD</td>
<td>Inclusion: COPD (FEV1 &lt; 50%, FEV1/FVC ratio &lt;60%, smoking history &gt;20 pack)</td>
<td>1) BPAP ST + home oxygen</td>
<td>Longest duration: 12 months</td>
<td>The BPAP ST group had significantly fewer AECOPD than the home oxygen alone group (rate ratio, 0.66;</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>---------------------------</td>
<td>---------</td>
<td>-----------------------------</td>
<td>----------------------------------</td>
<td>----------------------------------------</td>
<td>------------</td>
</tr>
<tr>
<td>Oscroft, 2014, RCT</td>
<td>COPD</td>
<td>Inclusion: COPD (FEV1 &lt; 50%, FEV1/FVC ratio &lt; 70%, TLC &gt; 80%, smoking history &gt; 20 pack years); daytime PaCO2 &gt; 7 kPa and pH &gt; 7.35</td>
<td>1) BPAP volume assured pressure support ventilation</td>
<td>3 months</td>
<td>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).</td>
</tr>
<tr>
<td>Autor, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>Autor, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>Autor, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>Autor, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>Autor, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>Autor, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>Autor, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>Autor, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>Autor, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>Autor, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
</tbody>
</table>

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.
<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Disease</th>
<th>Inclusion/Exclusion Criteria</th>
<th>Device And Settings Used (Group)</th>
<th>Duration Of Device Use In Home Setting</th>
<th>Conclusion</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>COPD</td>
<td>or PtCO2 &gt;9 kPa Exclusion: Age&gt;80 years, other respiratory disease, BMI&gt;40, significant OSA.</td>
<td>2) BPAP ST</td>
<td>3 months</td>
<td>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.</td>
</tr>
<tr>
<td>Paone, 2014[1], Observational study</td>
<td>COPD</td>
<td>Inclusion: COPD (FEV1 &lt;50%, FEV1/FVC ratio &lt;70%, &lt;20% improvement bronchodilator response); NIPPV during hospital stay; PaCO2 &gt; 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&gt;40, systemic corticosteroids.</td>
<td>1) BPAP ST + Home oxygen 2) Home oxygen</td>
<td>24 months</td>
<td>The BPAP ST + home oxygen group had significantly less hospital admissions (Rate Ratio= 0.50; 95% CI: 0.35 to 0.71; p&lt;0.01). There was no significant difference on mortality (27.1% vs. 22.2%; p=0.59).</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>---------------------------</td>
<td>---------</td>
<td>-----------------------------</td>
<td>---------------------------------</td>
<td>---------------------------------------</td>
<td>------------</td>
</tr>
<tr>
<td>Galli, 2014&lt;sup&gt;27&lt;/sup&gt;, Observational study</td>
<td>COPD</td>
<td>Inclusion: AECOPD (ICD-9); PaCO2 &gt; 45 mmHg; NIPPV during hospital stay Exclusion: discharged to hospice.</td>
<td>1) BPAP NOS</td>
<td>Longest duration: 6 months</td>
<td>BPAP was associated with significantly fewer hospital readmissions (p &lt; 0.0001) and ICU readmissions. There was no significant difference on mortality at 6-month followup (10% vs. 19%, p=0.13).</td>
</tr>
<tr>
<td>Bhatt, 2013&lt;sup&gt;3&lt;/sup&gt;, RCT</td>
<td>COPD</td>
<td>Inclusion: COPD (FEV/FVC &lt; 70%, smoking &gt;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&lt;35 years, diseases limiting life expectancy &lt;2 years, active malignancies in previous 2 years, process precluding a nasal mask.</td>
<td>1) BPAP NOS</td>
<td>Longest duration: 6 months</td>
<td>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.</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>----------------------------</td>
<td>---------</td>
<td>-----------------------------</td>
<td>----------------------------------</td>
<td>---------------------------------------</td>
<td>------------</td>
</tr>
<tr>
<td>Duiverman, 201121,22, RCT</td>
<td>COPD</td>
<td>Inclusion: COPD (FEV1 &lt;50%, FEV1/FVC &lt; 70%, GOLD stage III/IV); age 40-76 years; no exacerbation in past 4 weeks; daytime PaCO2 &gt;6.0 kPa</td>
<td>1) BPAP ST + Pulmonary rehabilitation</td>
<td>Longest duration: 24 months</td>
<td>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.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Exclusion: cardiac/neuromuscular disease limiting exercise tolerance, pulmonary rehabilitation in past 18 months, prior NIPPV, apnea-hypopnea index ≥10h.</td>
<td>2) Pulmonary rehabilitation alone</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Oscroft, 201015, Observational study</td>
<td>COPD</td>
<td>Inclusion: COPD (FEV1 &lt;50%, FEV1/FVC ratio &lt;70%, smoking history &gt;20 pack years); AECOPD requiring hospital admission; daytime PaCO2 &gt;7.5 kPa with pH 7.35-7.45 or daytime PaCO2 &gt;6.5 kPa with pH</td>
<td>1) BPAP ST started in AECOPD</td>
<td>28.6 months, 95% CI 10.9-46.8 months, Median 52.4 months</td>
<td>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).</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>-----------------------------</td>
<td>---------</td>
<td>-----------------------------</td>
<td>----------------------------------</td>
<td>----------------------------------------</td>
<td>------------</td>
</tr>
<tr>
<td>Cheung, 2010[^1], RCT</td>
<td>COPD</td>
<td>7.35-7.45 with PtcCO₂ &gt;9 kPa Exclusion: Age&gt;80 years, other respiratory disease, BMI&gt;35, significant OSA, tracheostomy, impaired left ventricular function.</td>
<td>2) BPAP ST started in stable COPD</td>
<td>Longest duration: 12 months</td>
<td>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).</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>1) CPAP</td>
<td>2) BPAP ST</td>
<td></td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>----------------------------</td>
<td>---------</td>
<td>------------------------------</td>
<td>----------------------------------</td>
<td>----------------------------------------</td>
<td>------------</td>
</tr>
<tr>
<td>McEvoy, 2009, RCT</td>
<td>COPD</td>
<td>Inclusion: COPD (FEV1&lt;50% or &lt;1.5L, bronchodilator response &lt;20%, FEV1/FVC ratio &lt;60%); PaCO2 &lt;46 mmHg at least twice in the prior 6 months during clinical stability; LTOT for ≥3 month; Age&lt;80 years Exclusion: current smokers, significant comorbidities (malignancies, left ventricular HF, unstable angina) likely affecting 2 year survival, severe psychiatric disorder impairing ability to comply to NIPPV, BMI&gt;40, evidence of sleep apnea.</td>
<td>1) BPAP S + Oxygen</td>
<td>Longest duration: 12 months</td>
<td>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.</td>
</tr>
<tr>
<td>Casanova9, 2000, RCT</td>
<td>COPD</td>
<td>Inclusion: COPD (FEV1 &lt;45%, FEV1/FVC &lt;70%, smoking &gt;20 pack years, TLC ≥80%); stable disease (no AECOPD in past 3 months); age 45-75</td>
<td>1) BPAP S + Standard care</td>
<td>Longest duration: 12 months</td>
<td>There were no significant differences on mortality, the number of acute exacerbations, hospital admissions, intubations, dyspnea (Medical Research</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>----------------------------</td>
<td>---------</td>
<td>-----------------------------</td>
<td>---------------------------------</td>
<td>----------------------------------------</td>
<td>------------</td>
</tr>
</tbody>
</table>
| Garrod, 2000<sup>10</sup>, RCT | COPD    | Inclusion: COPD (FEV1 <50%, bronchodilator response <15%); exercise intolerance due to dyspnea, no prior NIPPV  
Exclusion: unstable angina, intermittent claudication, other mobility-limiting conditions. | 1) BPAP S + Pulmonary rehabilitation  
2) Pulmonary rehabilitation | 2) Standard care | The BPAP S plus pulmonary rehabilitation had significantly better outcomes on quality of life (Chronic Respiratory Disease Questionnaire, 12.3; 95% CI: 1.19 to 23.4; p=0.03), and shuttle walk test (72 meters, 95% CI: 12.9 to 131 meters). There was no difference on activities of daily living, and dyspnea. |
| Clini, 1998<sup>14</sup>, Observational study | 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 |
<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Disease</th>
<th>Inclusion/Exclusion Criteria</th>
<th>Device And Settings Used (Group)</th>
<th>Duration Of Device Use In Home Setting</th>
<th>Conclusion</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>COPD</td>
<td>PaCO2 &gt;6 kPa (daytime room air); ≥1 ICU admission due to AECOPD in prior 2 years. Exclusion: other organ failure, cancer, suspected OSA.</td>
<td>2) Oxygen</td>
<td>(p&lt;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).</td>
<td></td>
</tr>
<tr>
<td>Clin, 1996, Observational study</td>
<td>COPD</td>
<td>Inclusion: COPD, LTOT ≥18 mo.; chronic PaCO2 &gt;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.</td>
<td>1) BPAP ST + Home care + Oxygen</td>
<td>Longest duration: 18 months</td>
<td>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).</td>
</tr>
<tr>
<td>Zhou, 2017, RCT</td>
<td>COPD</td>
<td>Inclusion: COPD (Gold Stage III/IV); chronic hypercapnia (measured during daytime at rest with no oxygen or NIPPV); age≥40 years. Exclusion:</td>
<td>1) BPAP ST</td>
<td>3 months</td>
<td>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</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>----------------------------</td>
<td>---------</td>
<td>------------------------------</td>
<td>---------------------------------</td>
<td>--------------------------------------</td>
<td>------------</td>
</tr>
<tr>
<td></td>
<td></td>
<td>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.</td>
<td>2) Standard care</td>
<td>standard care group. No significant difference was found on mortality, and quality of life (Severe Respiratory Insufficiency Questionnaire).</td>
<td></td>
</tr>
<tr>
<td>Marquez-Martin, 2014&lt;sup&gt;30&lt;/sup&gt;, RCT</td>
<td>COPD</td>
<td>Inclusion: COPD (FEV1 &lt;50%); PaO2 &lt; 60 mmHg (chronic); PaCO2 &gt; 45 mmHg (chronic).</td>
<td>1) BPAP ST</td>
<td>3 months</td>
<td>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</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>---------------------------</td>
<td>---------</td>
<td>-----------------------------</td>
<td>----------------------------------</td>
<td>---------------------------------------</td>
<td>------------</td>
</tr>
<tr>
<td>Köhnlein, 2014&lt;sup&gt;35&lt;/sup&gt;, RCT</td>
<td>COPD</td>
<td>Inclusion: COPD (GOLD IV); clinically stable (no AECOPD in prior 4 weeks); PaCO2 ≥ 7</td>
<td>1) BPAP ST + Standard care</td>
<td>12 months</td>
<td>The BPAP group was found to have significantly less mortality rate at 1...</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>2) Exercise program</td>
<td></td>
<td>combined group (p&lt;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).</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>---------------------------</td>
<td>---------</td>
<td>-----------------------------</td>
<td>---------------------------------</td>
<td>---------------------------------------</td>
<td>------------</td>
</tr>
<tr>
<td>De Backer, 2011&lt;sup&gt;18&lt;/sup&gt;, RCT</td>
<td>COPD</td>
<td>Inclusion: COPD (FEV1&lt;50%, FEV1/FVC &lt;70%), AECOPD requiring hospitalization, PaCO2 &gt;45 mmHg, stopped smoking</td>
<td>1) BPAP NOS</td>
<td>At least 6 months</td>
<td>The 6-minute walk distance increased significantly in the BPAP group (232 ± 151 m to 282 ± 146 m, p = 0.01), while there was no significant difference found on 6-minute walk distance test.</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>2) Standard care</td>
<td></td>
<td>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.</td>
</tr>
</tbody>
</table>

Inclusion/Exclusion Criteria:
- 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.
<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Disease</th>
<th>Inclusion/Exclusion Criteria</th>
<th>Device And Settings Used (Group)</th>
<th>Duration Of Device Use In Home Setting</th>
<th>Conclusion</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>COPD</td>
<td>Exclusion: COPD; AECOPD requiring NIPPV or invasive ventilation; chronic nocturnal NIPPV use at home for ≥ 6 months; clinically stable, PaCO2 &gt; 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.</td>
<td>2) BPAP NOS for more than 6 months</td>
<td>Longest duration: 12 months</td>
<td>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).</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>----------------------------</td>
<td>---------</td>
<td>-----------------------------</td>
<td>---------------------------------</td>
<td>----------------------------------------</td>
<td>------------</td>
</tr>
<tr>
<td>Dreher, 2010&lt;sup&gt;10&lt;/sup&gt;, RCT</td>
<td>COPD</td>
<td>Inclusion: COPD (Gold stage IV); daytime PaCO2 &gt; 45 mmHg; nocturnal PaCO2 &gt; 50 mmHg Exclusion: Acute RF, invasive ventilation via tracheostomy, weaned from invasive ventilation, intubated during prior 3 months, other ventilatory support prior to study.</td>
<td>1) HMV (pressure controlled ventilation) (time period 1) 2) HMV (pressure support ventilation)(time period 1) 2) Pulmonary rehabilitation alone</td>
<td>1.5 months</td>
<td>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.</td>
</tr>
<tr>
<td>Tsolaki, 2008&lt;sup&gt;39&lt;/sup&gt;, Observational study</td>
<td>COPD</td>
<td>Inclusion: COPD (FEV1 &lt;50%, FEV/FVC &lt;70%); smoking &gt;20 pack</td>
<td>1) BPAP ST</td>
<td>12 months</td>
<td>Compared to standard care, the BPAP group was found to have significantly better</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>-----------------------------</td>
<td>---------</td>
<td>------------------------------</td>
<td>----------------------------------</td>
<td>---------------------------------------</td>
<td>------------</td>
</tr>
<tr>
<td>Chiang, 2003&lt;sup&gt;12&lt;/sup&gt;, RCT</td>
<td>COPD, other</td>
<td>Inclusion: COPD or asthma or bronchiectasis; hospital readmission due to respiratory cause; daytime sleepiness or morning</td>
<td>1) BPAP NOS</td>
<td>6 months</td>
<td>Compared to the standard care group, the BPAP group had significantly better outcomes on 6-minute walk.</td>
</tr>
</tbody>
</table>

Inclusion/Exclusion Criteria:
- **1)** BPAP NOS
- 6 months

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.

Device And Settings Used (Group):
- 2) Standard care

Conclusion:
- 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.
<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Disease</th>
<th>Inclusion/Exclusion Criteria</th>
<th>Device And Settings Used (Group)</th>
<th>Duration Of Device Use In Home Setting</th>
<th>Conclusion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gay, 1996&lt;sup&gt;ii&lt;/sup&gt;, RCT</td>
<td>COPD</td>
<td>Inclusion: COPD (FEV1 &lt; 40%); PaCO2 &gt;45 mmHg (daytime, rest); Age&lt;80 years, BMI≤30</td>
<td>1) BPAP ST</td>
<td>3 months</td>
<td>No difference was found on 6-minute walk distance test, total sleep time, sleep efficiency,</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>2) Standard care</td>
<td></td>
<td>distance test group (101.2 meters vs. -33.8 meters, p&lt;0.05), number of hospitalization, and total hospital stay. No significant difference was found on resting Borg score and Borg score at end of 6-minute walk distance test.</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>---------------------------</td>
<td>---------</td>
<td>------------------------------</td>
<td>----------------------------------</td>
<td>----------------------------------------</td>
<td>------------</td>
</tr>
<tr>
<td>Gad, 2014&lt;sup&gt;30&lt;/sup&gt;, Observational study</td>
<td>COPD</td>
<td>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.</td>
<td>2) Sham BPAP ST/no device</td>
<td></td>
<td>REM sleep, and multiple sleep latency tests.</td>
</tr>
<tr>
<td>Sin, 2007&lt;sup&gt;5&lt;/sup&gt;, RCT</td>
<td>COPD</td>
<td>Inclusion: COPD (FEV1 &lt; 50%, FEV1/FVC &lt;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.</td>
<td>1) BPAP ST + Exercise program 2) Exercise program</td>
<td>3 months</td>
<td>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).</td>
</tr>
</tbody>
</table>
| | | | | | After 3 months, the changes in 6-minute walk distance test was
<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Disease</th>
<th>Inclusion/Exclusion Criteria</th>
<th>Device And Settings Used (Group)</th>
<th>Duration Of Device Use In Home Setting</th>
<th>Conclusion</th>
</tr>
</thead>
</table>
| Heinemann, 2011<sup>12</sup>, Observational study | COPD | 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. | 1) HMV pressure controlled ventilation
2) Sham BPAP/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). |
<p>| Budweiser, 2007&lt;sup&gt;7&lt;/sup&gt;, Observational study | COPD | Inclusion: severe COPD (Global Initiative of Chronic Obstructive Lung Disease (GOLD) IV, FEV1/VC &lt; 70% and FEV1&lt; 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&lt;0.05). |</p>
<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Disease</th>
<th>Inclusion/Exclusion Criteria</th>
<th>Device And Settings Used (Group)</th>
<th>Duration Of Device Use In Home Setting</th>
<th>Conclusion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clinl, 2002, RCT</td>
<td>COPD</td>
<td>Inclusion: severe COPD (American Thoracic Society criteria), CVF, stable clinical condition (arterial pH&gt;7.35, free from exacerbation in the 4 weeks), age≤75 years, LTOT at least 6 months, MRC dyspnea score≥2, FEV1&lt;1.5 L, FEV1/FVC&lt;60%, total lung capacity ≥90% predicted, PaCO2&gt;6.6 kPa, PaO2&lt;7.8 kPa</td>
<td>1) BPAP ST plus LTOT</td>
<td>24 months</td>
<td>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)</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>---------------------------</td>
<td>---------</td>
<td>------------------------------</td>
<td>----------------------------------</td>
<td>--------------------------------------</td>
<td>------------</td>
</tr>
<tr>
<td>Struijk, 2014&lt;sup&gt;SM&lt;/sup&gt;, RCT</td>
<td>COPD</td>
<td>Inclusion: Severe COPD (GOLD stage 3 and 4), &gt;48 hours independence from ventilatory support (invasive or non-invasive) for</td>
<td>1) BPAP ST</td>
<td>12 months</td>
<td>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.</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>----------------------------</td>
<td>---------</td>
<td>------------------------------</td>
<td>----------------------------------</td>
<td>---------------------------------------</td>
<td>------------</td>
</tr>
<tr>
<td>Durao, 2018(^1)</td>
<td>COPD</td>
<td>Inclusion: COPD NOS</td>
<td>1) HMV/BPAP mix started in AECOPD</td>
<td>&gt;1 year</td>
<td>There were no difference on number of hospital admission for respiratory causes (changes before</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Exclusion: No clinical assessment in prior 6 months, OSA with a history</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>2) Standard care</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>ARF, prolonged hypercapnia(PaCO(_2) &gt;6.0 kPa) during daytime at rest without oxygen or ventilatory support</td>
<td>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).</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>----------------------------</td>
<td>---------</td>
<td>-----------------------------</td>
<td>---------------------------------</td>
<td>--------------------------------------</td>
<td>------------</td>
</tr>
<tr>
<td>Duiverman, 2017&lt;sup&gt;23&lt;/sup&gt;</td>
<td>COPD</td>
<td>Inclusion: COPD (GOLD III or IV), ≥2 AECOPD with acute hypercapnic respiratory failure (pH&lt;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&gt;7.35).</td>
<td>1) HMV/BPAP mix (pressure controlled ventilation) (high intensity)</td>
<td>1.5 months</td>
<td>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).</td>
</tr>
<tr>
<td></td>
<td></td>
<td>2) HMV (pressure controlled ventilation) (low intensity)</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Note: ± denotes standard deviation.

<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Disease</th>
<th>Inclusion/Exclusion Criteria</th>
<th>Device And Settings Used (Group) (add n per arm)</th>
<th>Duration Of Device Use In Home Setting</th>
<th>Conclusion</th>
</tr>
</thead>
</table>
| Buyse, 2003<sup>3</sup>, Observational study | TRD     | Inclusion: Kyphoscoliosis with respiratory insufficiency who started LTOT and/or NIPPV. Exclusion: Rapidly progressive NMD, OHS, COPD, acute RF, severe acidosis. | 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<sup>13</sup>, Observational study | TRD     | Inclusion: TRD (post-TB or scoliosis); PaCO2 45-55 mmHg; stable PaCO2 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. |

<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Disease</th>
<th>Inclusion/Exclusion Criteria</th>
<th>Device And Settings Used (Group)</th>
<th>Duration Of Device Use In Home Setting</th>
<th>Conclusion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sanjuan-López, 2014&lt;sup&gt;44&lt;/sup&gt;, Observational study</td>
<td>NMD</td>
<td>Inclusion: ALS; hospital admission; chronic RF by pulmonologist Exclusion: Neuromuscular processes other than ALS, treatment in social welfare palliative center.</td>
<td>1) HMV (pressure support ventilation mode or BPAP ST mode) started after outpatient pulmonary evaluation 2) HMV (pressure support ventilation mode or BPAP ST mode) started in an emergency situation without prior outpatient pulmonary evaluation</td>
<td>23.3 months (95% CI, 16.7–28.8) 26.7 months</td>
<td>Patients received HMV after pulmonary evaluation have longer length of survival than those without pulmonary evaluation (mean survival: 12.3 months vs. 2.8 months, p&lt;0.004).</td>
</tr>
<tr>
<td>Pinto, 2010&lt;sup&gt;49&lt;/sup&gt;, Observational study</td>
<td>NMD</td>
<td>Inclusion: ALS; home BPAP use; FVC ≥75%; PaO₂ ≥80 mmHg; PaCO₂ ≤ 45 mmHg; age 18-75 years Exclusion: Gastrostomy, cognitive impairment, other significant disorders.</td>
<td>1) BPAP ST + Weekly telemonitoring + Standard care 2) BPAP ST + Standard care</td>
<td>36 months</td>
<td>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).</td>
</tr>
<tr>
<td>Gonzalez-Bermejo, 2013&lt;sup&gt;30&lt;/sup&gt;, Observational study</td>
<td>NMD</td>
<td>Inclusion: ALS on home BPAP with 4 hour/night minimal adherence Exclusion: Use of other ventilator types, without integrated SpO₂</td>
<td>1) BPAP ST &quot;correctly ventilated patients&quot; 2) BPAP ST &quot;insufficiently ventilated patients&quot;</td>
<td>12 months 12 months</td>
<td>The &quot;correctly ventilated&quot; patients had significantly higher survival than those &quot;insufficiently ventilated&quot; patients (OR= 0.25; 95% CI: 0.10 to 0.64).</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>----------------------------</td>
<td>---------</td>
<td>-----------------------------</td>
<td>---------------------------------</td>
<td>---------------------------------------</td>
<td>------------</td>
</tr>
</tbody>
</table>
| Sancho, 2014*<sup>12</sup>, Observational study | 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: 10.25 to 19.75), p=0.53) |
| Sivori, 2007*<sup>7</sup>, Observational study | 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). |
<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Disease</th>
<th>Inclusion/Exclusion Criteria</th>
<th>Device And Settings Used (Group)</th>
<th>Duration Of Device Use In Home Setting</th>
<th>Conclusion</th>
</tr>
</thead>
</table>
| Coco, 2006<sup>16</sup>, Observational study | NMD | Inclusion: ALS; symptomatic ventilatory impairment (dyspnea, morning headache, hypersomnia, fatigue) plus 1) PaCO2 $\geq$ 45 mmHg or 2) nocturnal oxygen saturation by pulse oximeter $\leq$ 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 $\geq$ 4 hours/day)  
2) BPAP ST < 4 hours/day | Longest Duration: 30 months | The group with $\geq$4 hours/day 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 |
<p>| Bourke, 2006&lt;sup&gt;6&lt;/sup&gt;, RCT | NMD | Inclusion: ALS; orthopnea with Pimax &lt;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 |</p>
<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Disease</th>
<th>Inclusion/Exclusion Criteria</th>
<th>Device And Settings Used (Group)</th>
<th>Duration Of Device Use In Home Setting</th>
<th>Conclusion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pinto, 1995&lt;sup&gt;48&lt;/sup&gt;, Observational study</td>
<td>NMD</td>
<td>Inclusion: ALS; bulbar features Exclusion: Tracheotomised, refusal of attempts to prolong survival.</td>
<td>1) BPAP NOS 2) No BPAP NOS</td>
<td>Longest Duration: 42 months</td>
<td>With a 3-year followup, patients treated with BPAP were found to have significantly higher overall survival than patients with palliative management (p=0.004).</td>
</tr>
<tr>
<td>Vitacca, 2017&lt;sup&gt;61&lt;/sup&gt; Observational study</td>
<td>NMD</td>
<td>Inclusion: ALS NOS admitted to hospital, NIPPV use Exclusion: dementia confirmed by Mini-Mental State Examination score &lt;20, refusal of NIPPV</td>
<td>1) HMV/BPAP mix started in FVC ≥ 80% (early) 2) HMV/BPAP mix started in FVC &lt;80% (late)</td>
<td>36 months</td>
<td>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 &lt;80% (late).</td>
</tr>
<tr>
<td>Sancho, 2018&lt;sup&gt;53&lt;/sup&gt;, Observational study</td>
<td>NMD</td>
<td>Inclusion: ALS (Escorial criteria), hospital admission Exclusion: lung disease, &lt;1 year life expectancy, NIV use &lt;4 consecutive hours/night, slow disease progression (&gt;3)</td>
<td>1) HMV (volume assist control ventilation) 2) No device</td>
<td>Longest Duration: 36 months</td>
<td>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:</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>---------------------------</td>
<td>---------</td>
<td>-----------------------------</td>
<td>----------------------------------</td>
<td>----------------------------------------</td>
<td>------------</td>
</tr>
<tr>
<td>Bertella, 2017&lt;sup&gt;2&lt;/sup&gt;, RCT</td>
<td>NMD</td>
<td>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 &gt;40 km.</td>
<td>1) BPAP volume assured pressure support ventilation outpatient initiation 2) BPAP volume assured pressure support ventilation inpatient initiation</td>
<td>3 months</td>
<td>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.</td>
</tr>
</tbody>
</table>


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

<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Disease</th>
<th>Inclusion/Exclusion Criteria</th>
<th>Device And Settings Used (Group) (add n per arm)</th>
<th>Duration Of Device Use In Home Setting</th>
<th>Conclusion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Howard, 2016&lt;sup&gt;14&lt;/sup&gt;, RCT</td>
<td>OHS</td>
<td>Inclusion: OHS (BMI &gt;30, daytime PaCO2 &gt;45 mmHg) Exclusion: Other</td>
<td>1) BPAP ST 2) CPAP</td>
<td>3 months 3 months</td>
<td>No significant difference was found between groups on Epworth Sleepiness Scale scores (p=0.86),</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group) (add n per arm)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>---------------------------</td>
<td>---------</td>
<td>-----------------------------</td>
<td>-------------------------------------------------</td>
<td>---------------------------------------</td>
<td>-------------</td>
</tr>
<tr>
<td>Masa, 2015&lt;sup&gt;18&lt;/sup&gt;, RCT</td>
<td>OHS</td>
<td>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 &lt;70%), NMD, narcolepsy, restless legs syndrome, psychophysical, severe chronic debilitating illness, severe chronic nasal obstruction.</td>
<td>1) HMV/BPAP mix (all with bilevel pressure with assured volume) + lifestyle modification 2) CPAP + Lifestyle modification 3) Lifestyle modification</td>
<td>2 months 2 months 2 months</td>
<td>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 groups on quality of life and number of dropouts.</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group) (add n per arm)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>---------------------------</td>
<td>---------</td>
<td>----------------------------</td>
<td>-----------------------------------------------</td>
<td>--------------------------------------</td>
<td>------------</td>
</tr>
<tr>
<td>Borel, 2011*, RCT</td>
<td>OHS</td>
<td>Inclusion: OHS (BMI &gt;30; daytime PaCO2 ≥ 45 mmHg); age 20-75 years Exclusion: Declined or presented any significant airway obstruction, scoliosis, cardiac failure, progressive NMD.</td>
<td>1) BPAP ST 2) Lifestyle counseling</td>
<td>Longest Duration: 1 month</td>
<td>No significant difference were found on sleep quality measured by Epworth Sleepiness Scale (p=0.49).</td>
</tr>
<tr>
<td>Murphy, 2012**, RCT</td>
<td>OHS</td>
<td>Inclusion: OHS (BMI&gt;40, daytime chronic PaCO2 &gt;6 kPa, pH &gt;7.35), absence of other identifiable hypoventilation cause, FEV1/FVC &gt;70%, FVC &lt;70% Exclusion: Inability to provide written consent.</td>
<td>1) BPAP AVAPS 2) BPAP ST</td>
<td>Longest Duration: 3 months</td>
<td>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).</td>
</tr>
<tr>
<td>Piper, 2008***, RCT</td>
<td>OHS</td>
<td>Inclusion: OHS (BMI≥30, PaCO2 ≥ 45 mmHg (awake, stable), absence of another cause for hypercapnia,</td>
<td>1) CPAP</td>
<td>Longest Duration: 3 months</td>
<td>No significant difference was found between groups on Epworth Sleepiness Scale (p=0.59), SF-36 Physical</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group) (add n per arm)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>---------------------------</td>
<td>---------</td>
<td>-----------------------------</td>
<td>-------------------------------------------</td>
<td>---------------------------------------</td>
<td>------------</td>
</tr>
<tr>
<td></td>
<td>FEV1/FVC ≥ 70%</td>
<td>Exclusion: psychiatric illness, current home NIPPV use, PtcCO2 during REM ≥10mmHg, increase in afternoon to morning PaCO2 ≥10mmHg in patients with awake PaCO2 &gt;55 mmHg.</td>
<td>2) BPAP S</td>
<td></td>
<td>Component (p=0.22), and SF-36 mental Component (p=0.28).</td>
</tr>
</tbody>
</table>

### Table F.16. Other Respiratory Diseases – Effectiveness of Home Devices

<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Disease</th>
<th>Inclusion/Exclusion Criteria</th>
<th>Device And Settings Used (Group)</th>
<th>Duration Of Device Use In Home Setting</th>
<th>Conclusion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Benhamou, 1997, Observational study</td>
<td>Other</td>
<td>Inclusion: Bronchiectasis; home nasal mask ventilation; LTOT.</td>
<td>1) HMV (volume assist control ventilation) + Oxygen</td>
<td>Longest Duration: 89 months</td>
<td>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&gt;0.05).</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>2) Oxygen alone</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

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

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

<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Disease</th>
<th>Inclusion/Exclusion Criteria</th>
<th>Device And Settings Used (Group)</th>
<th>Duration Of Device Use In Home Setting</th>
<th>Conclusion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hazenberg, 2014, RCT</td>
<td>NMD, TRD</td>
<td>Inclusion: NMD or thoracic cage disorder; PaCO2 &gt;45 mmHg with respiratory symptoms Exclusion: COPD, not mask naïve, acute RF, age &lt; 18 years, invasive ventilation, nursing home resident.</td>
<td>1) HMV started at home pressure controlled ventilation with change to volume assist control ventilation if not tolerated</td>
<td>Longest Duration: 6 months</td>
<td>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).</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>2) HMV started in the hospital pressure controlled ventilation with change to volume assist control ventilation if not tolerated</td>
<td>Longest Duration: 6 months</td>
<td></td>
</tr>
<tr>
<td>Munoz, 2005, Observational study</td>
<td>NMD, TRD</td>
<td>Inclusion: Hospital admission with CHRF secondary to NMD (ALS excluded) or kyphoscoliosis or post TB sequelae; PaCO2 &gt; 45 mmHg; HMV</td>
<td>1) HMV volume assist control ventilation</td>
<td>Longest Duration: 12 months</td>
<td>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</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>2) HMV volume control</td>
<td>Longest Duration: 3 months</td>
<td></td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Disease</td>
<td>Inclusion/Exclusion Criteria</td>
<td>Device And Settings Used (Group)</td>
<td>Duration Of Device Use In Home Setting</td>
<td>Conclusion</td>
</tr>
<tr>
<td>-----------------------------</td>
<td>---------</td>
<td>------------------------------</td>
<td>---------------------------------</td>
<td>---------------------------------------</td>
<td>------------</td>
</tr>
</tbody>
</table>
| Chiang, 2003[2], RCT        | COPD, other | started in stable phase of disease  
Exclusion: BiPAP users, ALS. | 1) BPAP NOS  
2) No BPAP NOS | 6 months |  
mode vs. 0.04 per patient in HMV volume control mode, p=0.11.  
Adverse events were similar between the two groups.  
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. |

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

Table F.18. COPD – Equipment parameters

<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Device/mode</th>
<th>Model (Manufacturer; Location of Manufacturer)</th>
<th>Device Characteristics</th>
<th>Prescribed Usage (frequency and duration)</th>
<th>Actual Usage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vasquez, 2017 Retrospective cohort</td>
<td>BPAP NOS</td>
<td>NR</td>
<td>IPAP, EPAP</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td></td>
<td>CPAP NOS</td>
<td>NR</td>
<td>CPAP</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td></td>
<td>HMV NOS</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Murphy, 2017 RCT</td>
<td>BPAP ST</td>
<td>Harmony (Philips Respironics; USA) (FDA approved) or VPAP III STa (ResMed; Bella Vista, Australia) (FDA approved)</td>
<td>IPAP, EPAP, rate</td>
<td>≥ 6 hours nightly</td>
<td>-4.7 (2.5-5.6) hours/day (6 weeks) -7.6(3.6-8.4) hours/day (12 months). -IPAP: 24 (22-26) cm H2O -EPAP: 4 (4-5) cmH2O -Rate: 14 (14-16) breaths/minute</td>
</tr>
<tr>
<td>Salturk, 2015 Retrospective cohort</td>
<td>BPAP ST</td>
<td>NR</td>
<td>IPAP, EPAP, rate</td>
<td>NR</td>
<td>-6.7 ± 1.9 hours/day -IPAP: 24 ± 3 cm H2O -EPAP: 5.3 ± 0.7 cmH2O</td>
</tr>
<tr>
<td>Oscroft, 2014 RCT</td>
<td>BPAP volume assured pressure support ventilation</td>
<td>Intelligent volume assured pressure support (iVAPS) (ResMed; Bella Vista, Australia) (FDA approved)</td>
<td>IPAP, EPAP, rate, target minute ventilation</td>
<td>NR</td>
<td>-Target minute ventilation 8.4 [5.7-9.8] L/minute -EPAP: 4 (4-4) cmH2O -Rate: 15 (13.3-19.4) breaths/minute</td>
</tr>
<tr>
<td></td>
<td>BPAP ST</td>
<td>NIPPY 3 (B and D Electromedical; Stratford, United Kingdom) (Not FDA approved)</td>
<td>IPAP, EPAP, rate</td>
<td>NR</td>
<td>-IPAP: 28 (27.3-30) cmH2O -EPAP: 5 (5-5) cmH2O -Rate: 15.0 (15-15) breaths/minute</td>
</tr>
<tr>
<td>Paone, 2014 Prospective cohort</td>
<td>BPAP ST</td>
<td>Synchrony (Philips Respironics; Andover, MA) (FDA approved) or Neftis (Linde; Munich, Germany) (FDA approved)</td>
<td>IPAP, EPAP, rate</td>
<td>NR</td>
<td>-IPAP: 18.5 ± 2.66 cm H2O -EPAP: 3.9 ± 1 cm H2O -Rate: 12 breaths/minute</td>
</tr>
<tr>
<td>Galli, 2014 Retrospective cohort</td>
<td>BPAP NOS</td>
<td>NR</td>
<td>IPAP, EPAP</td>
<td>NR</td>
<td>-IPAP: 22.1 ± 6.2 cm H2O -EPAP: 5.9 ± 1.8 cm</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Device/mode</td>
<td>Model (Manufacturer; Location of Manufacturer)</td>
<td>Device Characteristics</td>
<td>Prescribed Usage (frequency and duration)</td>
<td>Actual Usage</td>
</tr>
<tr>
<td>---------------------------</td>
<td>-------------</td>
<td>-----------------------------------------------</td>
<td>------------------------</td>
<td>------------------------------------------</td>
<td>--------------</td>
</tr>
</tbody>
</table>
| Bhatt, 2013<sup>3</sup>  | BPAP NOS    | BiPAP Synchrony (Respironics Inc.; Murrysville, USA) (FDA approved) | IPAP, EPAP | ≥ 6 hours daily for 6 months | H2O | -IPAP: 15 cm H2O  
-EPAP: 5 cm H2O |
| Duiverman, 2011<sup>21, 22</sup> RCT | BPAP ST | BiPAP Synchrony (Respironics Inc.; Murrysville, USA) (FDA approved) | 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<sup>13</sup>  | BPAP ST | NIPPY 1, 2 or 3 (B & D Electromedical; Stratford, United Kingdom) (Not FDA approved) | 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<sup>11</sup> RCT | CPAP | NR | CPAP | >8 hours nightly for 12 months | NR |
|                        | BPAP ST | NR | IPAP, EPAP, rate | >8 hours nightly for 12 months | -7-9 hours/night  
-IPAP: 14.8 ± 1.1 cm H2O  
-EPAP: 5 ± 0 cm H2O |
| Hitzl, 2009<sup>13</sup> Prospective cohort | 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<sup>40</sup> RCT | BPAP S | VPAP S mode (ResMed; Sydney, Australia) (FDA approved) | 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 |
<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Device/mode</th>
<th>Model (Manufacturer; Location of Manufacturer)</th>
<th>Device Characteristics</th>
<th>Prescribed Usage (frequency and duration)</th>
<th>Actual Usage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Windisch, 2006&lt;sup&gt;6&lt;/sup&gt; Observational study</td>
<td>HMV (pressure controlled ventilation)</td>
<td>PV401 (Breas Medical AB; Moelnlycke, Sweden) (FDA approved)</td>
<td>Inspiratory pressure, PEEP, inspiratory time, rate</td>
<td>NR</td>
<td>-IPAP: 31.0 ± 6.6 mbar -Rate: 20.7 ± 2.1 breaths/minute -Inspiratory time 1.0 ± 0.1 seconds</td>
</tr>
<tr>
<td>Casanova, 2000&lt;sup&gt;1&lt;/sup&gt; RCT</td>
<td>BPAP S</td>
<td>DP-90 (Taema; Paris, France) (Not FDA approved)</td>
<td>IPAP, EPAP</td>
<td>NR</td>
<td>-6.2 hours/day (at 3 months) -5.9 hours/day (at 6 months) -IPAP: 12 ± 2 cm H2O</td>
</tr>
<tr>
<td>Garrod, 2000&lt;sup&gt;16&lt;/sup&gt; RCT</td>
<td>BPAP S</td>
<td>BiPAP ST 30 (Respironics Inc.; Murrysville, USA) (FDA approved)</td>
<td>IPAP, EPAP</td>
<td>≥ 8 hours daily</td>
<td>-IPAP: 16 (13-24) cm H2O -EPAP: 4 (4-6) cm H2O</td>
</tr>
<tr>
<td>Clini, 1998&lt;sup&gt;14&lt;/sup&gt; Prospective cohort</td>
<td>BPAP ST</td>
<td>BiPAP (Respironics Inc.; Murrysville, USA) (FDA approved)</td>
<td>IPAP, EPAP, rate</td>
<td>NR</td>
<td>-7.4 ± 1.3 hours/day -IPAP: 10-16 cm H2O -EPAP: 2-4 cm H2O</td>
</tr>
<tr>
<td>Clini, 1996&lt;sup&gt;13&lt;/sup&gt; Prospective cohort</td>
<td>BPAP ST</td>
<td>BiPAP (Respironics Inc.; Murrysville, USA) (FDA approved)</td>
<td>IPAP, EPAP, rate</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Zhou, 2017 RCT</td>
<td>BPAP ST</td>
<td>Flexo ST 30 NIV (Curative Co.; SuZhou, China) (Not FDA approved)</td>
<td>IPAP, EPAP, rate</td>
<td>NR</td>
<td>-5.6 ± 1.4 hours/day -IPAP: 17.8 ± 2.08 cm H2O -EPAP: 4.2 ± 0.1 cm H2O</td>
</tr>
<tr>
<td>Marquez-Martin, 2014&lt;sup&gt;16&lt;/sup&gt; RCT</td>
<td>BPAP ST</td>
<td>BiPAP (Respironics Inc.; Murrysville, USA) (FDA approved)</td>
<td>IPAP, EPAP, rate</td>
<td>NR</td>
<td>-7 (6.5-9) hours nightly -IPAP: 16 cm H2O (median) (both NIPPV groups) -EPAP: 4 cm H2O (median, both NIPPV groups)</td>
</tr>
<tr>
<td>Köhnlein, 2014&lt;sup&gt;15&lt;/sup&gt; RCT</td>
<td>BPAP ST</td>
<td>Models not reported. Manufacturers: ResMed (Martinsried, Germany), Weinmann (Hamburg, Germany, or Tyco Healthcare (Neubrug, Germany) (FDA approved)</td>
<td>IPAP, EPAP, rate</td>
<td>≥ 6 hours daily</td>
<td>-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,</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Device/mode</td>
<td>Model (Manufacturer; Location of Manufacturer)</td>
<td>Device Characteristics</td>
<td>Prescribed Usage (frequency and duration)</td>
<td>Actual Usage</td>
</tr>
<tr>
<td>---------------------------</td>
<td>-------------</td>
<td>-----------------------------------------------</td>
<td>------------------------</td>
<td>------------------------------------------</td>
<td>--------------</td>
</tr>
<tr>
<td>De Backer, 2011&lt;sup&gt;18&lt;/sup&gt; RCT</td>
<td>BPAP NOS</td>
<td>BiPAP Synchrony (Respironics Inc., Murrysville, USA) (FDA approved)</td>
<td>IPAP, EPAP</td>
<td>&gt; 5 hours daily</td>
<td>NR</td>
</tr>
<tr>
<td>Funk, 2010&lt;sup&gt;25&lt;/sup&gt; RCT</td>
<td>BPAP NOS</td>
<td>NR</td>
<td>IPAP, EPAP</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Dreher, 2010&lt;sup&gt;25&lt;/sup&gt; RCT</td>
<td>HMV (pressure controlled ventilation)</td>
<td>Breas Vivo 40 (Breas Medical AB; Molnlycke, Sweden) (FDA approved) or Smart Air (Airox; Pau Cedex, France) (FDA approved)</td>
<td>Inspiratory pressure, PEEP, inspiratory time, rate, inspiratory flow trigger</td>
<td>Entire night while sleeping and during daytime naps.</td>
<td>-IPAP: 28.6 ± 1.9 cm H2O -EPAP: 4.5 ± 0.7 cm H2O -Rate: 17.5 ± 0.7 breaths/minute</td>
</tr>
<tr>
<td></td>
<td>HMV (pressure support ventilation)</td>
<td>Breas Vivo 40 (Breas Medical AB; Molnlycke, Sweden) (FDA approved) or Smart Air (Airox; Pau Cedex, France) (FDA approved)</td>
<td>Inspiratory pressure, PEEP, inspiratory flow trigger, expiratory flow trigger</td>
<td>Use the entire night while sleeping and during daytime naps.</td>
<td>-IPAP: 14.6 ± 0.8 cm H2O -EPAP: 4 ± 0 cm H2O -Rate: 8.0 ± 0 breaths/minute</td>
</tr>
<tr>
<td>Tsolaki, 2008&lt;sup&gt;19&lt;/sup&gt; Prospective cohort</td>
<td>BPAP ST</td>
<td>VPAP III ST (ResMed; Sydney, Australia) (FDA approved)</td>
<td>IPAP, EPAP, rate</td>
<td>≥ 5 hours daily</td>
<td>-9 ± 2.2 hours/day -IPAP: 15.3 ± 2 cm H2O -EPAP: 5.4 ± 0.7 (4-8) cm H2O</td>
</tr>
<tr>
<td>Gay, 1996&lt;sup&gt;19&lt;/sup&gt; RCT</td>
<td>BPAP ST</td>
<td>BiPAP (Respironics Inc.; Murrysville, USA) (FDA approved)</td>
<td>IPAP, EPAP, rate</td>
<td>NR</td>
<td>-5.1 ± 3.8 hours/day</td>
</tr>
<tr>
<td>Gad, 2014&lt;sup&gt;26&lt;/sup&gt; Prospective cohort</td>
<td>BPAP ST</td>
<td>NR</td>
<td>IPAP, EPAP, rate</td>
<td>NR</td>
<td>-IPAP: 15.5 ± 4.2 cm H2O -EPAP: 4.0 ± 0 cm H2O -9 ± 2 hours/day</td>
</tr>
<tr>
<td>Sin, 2007&lt;sup&gt;56&lt;/sup&gt; RCT</td>
<td>BPAP NOS</td>
<td>VPAP II (ResMed; Sydney, Australia) (FDA approved)</td>
<td>IPAP, EPAP</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Heinemann,</td>
<td>BPAP</td>
<td>NR</td>
<td>IPAP, EPAP, rate,</td>
<td>NR</td>
<td>-IPAP: 22.7 ± 4.3 mbar</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Device/mode</td>
<td>Model (Manufacturer; Location of Manufacturer)</td>
<td>Device Characteristics</td>
<td>Prescribed Usage (frequency and duration)</td>
<td>Actual Usage</td>
</tr>
<tr>
<td>---------------------------</td>
<td>-------------</td>
<td>-----------------------------------------------</td>
<td>------------------------</td>
<td>------------------------------------------</td>
<td>--------------</td>
</tr>
<tr>
<td>2011&lt;sup&gt;12&lt;/sup&gt;</td>
<td>(pressure controlled ventilation)</td>
<td>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)</td>
<td>inspiratory time</td>
<td>-EPAP: 5.0 ± 1.3 mbar -Rate: 16.8 ± 3.0 breaths/minute</td>
<td></td>
</tr>
<tr>
<td>Budweiser, 2007&lt;sup&gt;7&lt;/sup&gt;</td>
<td>BPAP (pressure controlled ventilation)</td>
<td>IPAP, EPAP, rate, inspiratory time</td>
<td>NR</td>
<td>-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 breaths/minute</td>
<td></td>
</tr>
<tr>
<td>Clini, 2002&lt;sup&gt;13&lt;/sup&gt;</td>
<td>BPAP ST</td>
<td>BiPAP ST 30 (Respironics Inc.; Murrysville, USA) (FDA approved)</td>
<td>IPAP, EPAP, rate</td>
<td>NR</td>
<td>-9 ± 2 hours/day -IPAP: 14 ± 3 cm H2O -EPAP: 2 ± 1 cm H2O</td>
</tr>
<tr>
<td>Struijker, 2014&lt;sup&gt;38&lt;/sup&gt;</td>
<td>BPAP ST</td>
<td>BiPAP Synchrony (Respironics Inc.; Murrysville, USA) (FDA approved)</td>
<td>IPAP, EPAP, rate</td>
<td>NR</td>
<td>-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</td>
</tr>
<tr>
<td>Durao, 2018&lt;sup&gt;34&lt;/sup&gt;</td>
<td>HMV/BPAP mix. HMV mode: pressure controlled ventilation. BPAP modes: ST and volume assured pressure support ventilation</td>
<td>VPAP ST S9 (Resmed) VPAP ST STA (Resmed) BIPAP PR1 (Philips Respironics) BiPAP A30 (Philips Respironics) BiPAP A40 (Philips Respironics)</td>
<td>IPAP, EPAP, rate, inspiratory time, target tidal volume</td>
<td>NR</td>
<td>-8.7 ± 3.6 hours/day -IPAP: 23.7 ± 5.3 cm H2O -Rate: 15.2 ± 1.4 breaths/minute</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Device/mode</td>
<td>Model (Manufacturer; Location of Manufacturer)</td>
<td>Device Characteristics</td>
<td>Prescribed Usage (frequency and duration)</td>
<td>Actual Usage</td>
</tr>
<tr>
<td>---------------------------</td>
<td>-------------</td>
<td>------------------------------------------------</td>
<td>------------------------</td>
<td>------------------------------------------</td>
<td>--------------</td>
</tr>
<tr>
<td>Blankenburg, 2017&lt;sup&gt;4&lt;/sup&gt;</td>
<td>HMV (pressure controlled ventilation or pressure support ventilation)</td>
<td>VS III; ResMed (Saime SA, France) (FDA approved)</td>
<td>Inspiratory pressure, PEEP, inspiratory time, rate, inspiratory flow trigger</td>
<td>12 hours/day</td>
<td>-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</td>
</tr>
<tr>
<td>Blankenburg, 2017&lt;sup&gt;3&lt;/sup&gt;</td>
<td>HMV/BPAP mix (pressure controlled ventilation) (high intensity)</td>
<td>Breas Vivo 50 (Breas Medical AB; Molnlycke, Sweden) (FDA approved)</td>
<td>Inspiratory pressure, PEEP, inspiratory time, rate, inspiratory flow trigger</td>
<td>-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</td>
<td></td>
</tr>
<tr>
<td>Blankenburg, 2017&lt;sup&gt;4&lt;/sup&gt;</td>
<td>HMV/BPAP mix (pressure support ventilation) (low intensity)</td>
<td>Breas Vivo 50 (Breas Medical AB; Molnlycke, Sweden) (FDA approved) or Stellar 100; Resmed (Martinsried, Germany) (FDA approved)</td>
<td>Inspiratory pressure, PEEP, inspiratory flow trigger, expiratory flow trigger</td>
<td>-4.2 (0.04-7.5) hours/day -IPAP: 15.5 ± 1.1 cm H2O -EPAP: 5.2 ± 0.6 cm H2O -Rate: 11.6 ± 1.5 breaths/minute</td>
<td></td>
</tr>
</tbody>
</table>

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


Table F.19. Thoracic Restrictive Disorders – Equipment parameters
<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Device/mode</th>
<th>Model; Manufacturer (Location of manufacturer)</th>
<th>Device Characteristics</th>
<th>Prescribed Usage (frequency and duration)</th>
<th>Actual Usage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Salturk, 2015&lt;sup&gt;11&lt;/sup&gt; Retrospective cohort</td>
<td>BPAP ST</td>
<td>NR</td>
<td>IPAP, EPAP, rate</td>
<td>NR</td>
<td>5.9 ± 1.8 hours/day &lt;br&gt; -IPAP: 22 ± 5 cm H2O &lt;br&gt; -EPAP: 5.3 ± 0.6 cmH2O</td>
</tr>
<tr>
<td>Hitzl, 2009&lt;sup&gt;13&lt;/sup&gt; Prospective cohort</td>
<td>HMV (pressure controlled ventilation)</td>
<td>NR</td>
<td>inspiratory pressure, PEEP, inspiratory time, rate</td>
<td>NR</td>
<td>-IPAP: 20.9 ± 4.0 cm H2O &lt;br&gt; -EPAP: 4.2 ± 1.9 cm H2O &lt;br&gt; -Rate: 19.1 ± 3.8 breaths/minute</td>
</tr>
<tr>
<td>Doménech-Clar, 2003&lt;sup&gt;10&lt;/sup&gt; Prospective cohort</td>
<td>BPAP NOS</td>
<td>DP-90 (Taema; Paris, France) (Not FDA approved)</td>
<td>IPAP, EPAP</td>
<td>≥7 hours/night</td>
<td>-mean 6 hours/night</td>
</tr>
<tr>
<td>Buyse, 2003&lt;sup&gt;6&lt;/sup&gt; Retrospective cohort</td>
<td>HMV (volume cycled or pressure cycled)</td>
<td>Eole 3 (Saime; Savigny-Le-Temple, France) (FDA approved) or O'nyx (Nellcor Puritan Bennet; Villers-les-Nancy, France) (FDA approved)</td>
<td>NR</td>
<td>NR</td>
<td></td>
</tr>
<tr>
<td>Nauffal, 2002&lt;sup&gt;24&lt;/sup&gt; Prospective cohort</td>
<td>BPAP NOS</td>
<td>DP-90 (Taema; Paris, France) (Not FDA approved)</td>
<td>IPAP, EPAP</td>
<td>NR</td>
<td>-mean 7 hours/night</td>
</tr>
<tr>
<td>Schonhofer, 2001&lt;sup&gt;15&lt;/sup&gt; Prospective cohort</td>
<td>Mixed: HMV (volume assist control ventilation) with change to BPAP ST if not tolerated</td>
<td>HMV: Drager EV 800 (Drager; Lubeck, Germany) or PLV 100 (Respironics; Murrysville, USA) (FDA approved) &lt;br&gt; BPAP ST: BP-T (Respironics Inc.; Murrysville, USA) (FDA approved)</td>
<td>HMV: tidal volume, PEEP, rate &lt;br&gt; BPAP: IPAP, EPAP, rate</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Masa, 2000&lt;sup&gt;7&lt;/sup&gt; Prospective cohort</td>
<td>HMV (volume cycled or pressure cycled)</td>
<td>Monal DCC (Taema; Paris, France). (Not FDA approved) &lt;br&gt; If could not tolerate Monal DCC, then patients were switched to a Onyx Plus</td>
<td>NR</td>
<td>NR</td>
<td>-7.3 ± 0.7 hours/day</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Device/mode</td>
<td>Model; Manufacturer (Location of manufacturer)</td>
<td>Device Characteristics</td>
<td>Prescribed Usage (frequency and duration)</td>
<td>Actual Usage</td>
</tr>
<tr>
<td>----------------------------</td>
<td>-------------</td>
<td>-----------------------------------------------</td>
<td>------------------------</td>
<td>------------------------------------------</td>
<td>--------------</td>
</tr>
<tr>
<td></td>
<td></td>
<td>(Mallinckrodt SEFAM; Nancy, France). (FDA approved)</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Note: ± denotes standard deviation.

<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Device/mode</th>
<th>Model and Manufacturer</th>
<th>Device Characteristics</th>
<th>Prescribed Usage (frequency and duration)</th>
<th>Actual Usage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sanjuan-López, 2014&lt;sup&gt;44&lt;/sup&gt; Retrospective cohort</td>
<td>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</td>
<td>VS ultra and VS III (ResMed) (FDA approved)</td>
<td>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</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Pinto, 2010&lt;sup&gt;49&lt;/sup&gt; Prospective cohort</td>
<td>BPAP ST + weekly telemonitoring</td>
<td>Goodknight 425ST bi-level device (Tyco Healthcare Group LP; California) (FDA approved)</td>
<td>-IPAP, EPAP, rate -FlowSens technology (allows the physician &quot;to customize the inspiratory and expiratory settings for greater patient comfort and synchronicity&quot;) -Telemonitoring (wireless telemetry to remotely monitor settings and change ventilator settings and to detect alarms. &quot;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.&quot; Patients were instructed to activate the system once a week or when difficulties arose.</td>
<td>≥6 hours/day</td>
<td>NR</td>
</tr>
<tr>
<td></td>
<td>BPAP ST (no telemonitoring)</td>
<td>Goodknight 425ST bi-level device (Tyco Healthcare Group LP; California) (FDA approved)</td>
<td>-IPAP, EPAP, rate -FlowSens technology (allows the physician &quot;to customize the inspiratory and expiratory settings for greater patient comfort and synchronicity&quot;)</td>
<td>≥6 hours/day</td>
<td>NR</td>
</tr>
<tr>
<td>Doménech-Clar, 2003&lt;sup&gt;19&lt;/sup&gt; Prospective cohort</td>
<td>BPAP NOS</td>
<td>DP-90 (Taema; Paris, France) (Not FDA approved)</td>
<td>IPAP, EPAP</td>
<td>≥7 hours/night</td>
<td>-Mean 6 hours/night</td>
</tr>
<tr>
<td>Nauffal, 2002&lt;sup&gt;44&lt;/sup&gt; Prospective</td>
<td>BPAP NOS</td>
<td>DP-90 (Taema; Paris, France)</td>
<td>IPAP, EPAP</td>
<td>NR</td>
<td>-Mean 7 hours/night</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Device/mode</td>
<td>Model and Manufacturer</td>
<td>Device Characteristics</td>
<td>Prescribed Usage (frequency and duration)</td>
<td>Actual Usage</td>
</tr>
<tr>
<td>-----------------------------</td>
<td>-------------</td>
<td>------------------------</td>
<td>------------------------</td>
<td>-------------------------------------------</td>
<td>--------------</td>
</tr>
</tbody>
</table>
| Gonzalez-Bermejo, 2013<sup>30</sup> Retrospective cohort | BPAP ST | VPAP-III or VPAP-IV plus Reslink automatic ventilatory signal analysis (Resmed; Sydney, Australia) (FDA approved) | IPAP, EPAP, rate | As long as possible at night and during daytime as needed | -IPAP: 13 ± 2 cm H2O  
-EPAP: 5 ± 2 cm H2O  
-Rate: 12 ± 1 breaths/minute |
| Sancho, 2014<sup>24</sup> Retrospective cohort | HMV, volume assist control ventilation | PV 501 (Breas Medical; Molndal, Sweden) (FDA approved) or Legendair (Airox; Pau, France) (FDA approved) | 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) (FDA approved) | IPAP, EPAP, rate | -IPAP: 12.01 ± 2.38 cm H2O  
-EPAP: 4.43 ± 1.14 cm H2O  
-Tidal volume: 417.84 ± 136.62 ml  
-Rate: 11.66 ± 0.99 breaths/minute |
| Sivori, 2007<sup>27</sup> Prospective cohort | BPAP NOS | NR | IPAP, EPAP | NR | NR |
| Coco, 2006<sup>16</sup> Prospective cohort | BPAP ST | BiPAP (Respironics Inc.; Vitalaire, Italy) (FDA approved) | IPAP, EPAP, rate | Use ≥ 4 or < 4 hours/day | NR |
| Bourke, 2006<sup>6</sup> RCT | BPAP ST | VPAP STII (ResMed UK Ltd; Abingdon, United Kingdom) (FDA approved) | IPAP, EPAP, rate | NR | -Mean 9.3 hours/day (good bulbar)  
-Mean 3.8 hours/day (poor bulbar)  
-Mean IPAP 15 |
<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Device/mode</th>
<th>Model and Manufacturer</th>
<th>Device Characteristics</th>
<th>Prescribed Usage (frequency and duration)</th>
<th>Actual Usage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pinto, 1995 Prospective cohort</td>
<td>BPAP NOS</td>
<td>NR</td>
<td>IPAP, EPAP</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Vitacca, 2017</td>
<td>HMV/BPAP mix using the following modes: ST, AVAPS, Bi-level, volume cycled, pressure controlled ventilation</td>
<td>NR</td>
<td>NR</td>
<td>≥4 hours/day and ≥120 hours/month</td>
<td>-IPAP: 15.33 ± 3.62 cm H2O -EPAP: 5.34 ± 1.77 cm H2O -Tidal volume: 7.06 ± 1.47 ml/kg -Rate: 12.67 ± 1.46 breaths/minute</td>
</tr>
<tr>
<td>Sancho, 2017</td>
<td>HMV (volume assist control ventilation)</td>
<td>Vivo 50; Breas Medical (Molndal, Sweden) (FDA approved) Trilogy 100; Philips Respironics (Madrid, Spain) (FDA approved)</td>
<td>Tidal volume, PEEP, rate</td>
<td>≥4 hours/day</td>
<td>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</td>
</tr>
<tr>
<td>Bertella, 2017</td>
<td>BPAP volume assured pressure support ventilation</td>
<td>Trend II ST 30; Hoffrichter (Schwerin, Germany) (not FDA approved) BiPAP Synchrony II, Philips Respironics (Murrysville, PA,</td>
<td>IPAP, EPAP, rate, target minute ventilation</td>
<td>≥4 hours /day</td>
<td>Inpatient: 6.97 ± 1.05 hours/day Outpatient: 7.68 ± 0.67 hours/day</td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Device/mode</td>
<td>Model and Manufacturer</td>
<td>Device Characteristics</td>
<td>Prescribed Usage (frequency and duration)</td>
<td>Actual Usage</td>
</tr>
<tr>
<td>----------------------------</td>
<td>-------------</td>
<td>------------------------</td>
<td>------------------------</td>
<td>-------------------------------------------</td>
<td>--------------</td>
</tr>
<tr>
<td></td>
<td>UPA) (FDA approved)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

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

<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Device/model</th>
<th>Model and Manufacturer</th>
<th>Device Characteristics</th>
<th>Prescribed Usage (frequency and duration)</th>
<th>Actual Usage</th>
</tr>
</thead>
</table>
| Howard, 2016
RCT             | BPAP ST      | NR                     | IPAP, EPAP, rate      | NR                                       | -5.3 (2.63) hours/night |
|                   |              |                        |                       | IPAP: 19.3 ± 2.8 cm H2O                  | -5.0 (2.4) hours/night |
|                   |              |                        |                       | EPAP: 11.9 ± 2.3 cm H2O                  |                           |
|                   |              |                        |                       | Rate: 15.0 ± 2.7 breaths/minute           |                           |
| Salturk, 2015
Retrospective cohort | BPAP ST      | NR                     | IPAP, EPAP, rate      | NR                                       | -6.4 ± 2.4 hours/day |
|                   |              |                        |                       | IPAP: 23 ± 3 cm H2O                      | -5.7 ± 1.3 hours/night |
|                   |              |                        |                       | EPAP: 5.8 ± 0.8 cm H2O                   |                           |
| Masa, 2000
Prospective cohort | HMV (volume cycled or pressure cycled) | Monal DCC (Taema; Paris, France). (Not FDA approved) | NR | -7.2 ± 0.8 hours/day |
|                   |              |                        |                       | If could not tolerate Monal DCC, then patients were switched to a Onyx Plus (Mallincrodt SEFAM; Nancy, France). (FDA approved) | |
| Castillejo, 2014
Prospective cohort | BPAP ST      | Harmony BiPAP (Respironics; Louisville, USA) (FDA approved) | IPAP, EPAP, rate | -5.7 ± 1.3 hours/night |
| Masa, 2015
RCT             | Mixed: HMV and BPAP mix (all with bilevel pressure with assured volume) | Breas Vivo 40 (General Electric; England) (FDA approved) or BiPAP AVAPS (Philips-Respironics; Netherlands) (FDA approved) or Trilogy 100 (Philips-Respironics; Netherlands) (FDA approved) or VS Ultra (ResMed; Australia) | IPAP, EPAP, rate, target minute ventilation | -IPAP: at Initiation 20 ± 3.3 cm H2O; at 2 months 20 ± 3 cm H2O |
<p>|                   |              |                        |                       | -EPAP: at Initiation 7.7 ± 1.8 cm H2O     | -5.7 ± 1.8 cm H2O |
|                   |              |                        |                       | -Rate: at Initiation 14 ± 3 breaths/minute; at 2 months 14 ± 3.1 breaths/minute | |</p>
<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Device/mode</th>
<th>Model and Manufacturer</th>
<th>Device Characteristics</th>
<th>Prescribed Usage (frequency and duration)</th>
<th>Actual Usage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Borel, 2011&lt;sup&gt;5&lt;/sup&gt; RCT</td>
<td>CPAP</td>
<td>Monal T50 (Air Liquide; France), or Puritan Bennett 560 (Puritan Bennett; USA) (FDA approved)</td>
<td>CPAP</td>
<td>NR</td>
<td>-CPAP: at Initiation 11 ± 2.5 cm H2O; at 2 months 11 ± 2.6 cm H2O</td>
</tr>
<tr>
<td>Murphy, 2012&lt;sup&gt;12&lt;/sup&gt; RCT</td>
<td>BPAP ST</td>
<td>GoodKnight-425ST (Covidien) (FDA approved)</td>
<td>IPAP, EPAP, rate</td>
<td>NR</td>
<td>-5.6 ± 2.2 hours/night -IPAP: 18 ± 3 cm H2O -EPAP: 11 ± 2 cm H2O -Rate: 13 ± 2 breaths/minute</td>
</tr>
<tr>
<td>Piper, 2008&lt;sup&gt;50&lt;/sup&gt; RCT</td>
<td>BPAP ST</td>
<td>BiPAP synchrony (Respirronics Inc.; Murrysville, USA) (FDA approved)</td>
<td>IPAP, EPAP, rate, target minute ventilation</td>
<td>NR</td>
<td>-IPAP: 25 ± 3 cm H2O -EPAP: 10 ± 2 cm H2O -Rate: 14 ± 1 breaths/minute</td>
</tr>
<tr>
<td>Blankenburg, 2017&lt;sup&gt;4&lt;/sup&gt;</td>
<td>HMV</td>
<td>VS III; ResMed (Saime SA, France) (FDA approved)</td>
<td>Inspiratory pressure, PEEP, inspiratory time, rate, inspiratory flow trigger</td>
<td>12 hours/day</td>
<td>-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</td>
</tr>
</tbody>
</table>

Note: ± denotes standard deviation.

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

<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Device/mode</th>
<th>Model and Manufacturer</th>
<th>Device Characteristics</th>
<th>Prescribed Usage (frequency and duration)</th>
<th>Actual Usage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Salturk, 2015&lt;sup&gt;1&lt;/sup&gt; Retrospective cohort</td>
<td>BPAP ST</td>
<td>NR</td>
<td>IPAP, EPAP, rate</td>
<td>NR</td>
<td>-5.8 ± 1.4 hours/day -IPAP: 21 ± 5 cm H2O -EPAP: 5.5 ± 0.7 cmH2O</td>
</tr>
<tr>
<td>Benhamou, 1997&lt;sup&gt;1&lt;/sup&gt; Case-control study</td>
<td>HMV (volume assist control ventilation)</td>
<td>Monnal D (Taema; Antony, France) (Not FDA approved) or Eole 3 (Saime; Savigny-Le-Temple, France) (FDA approved)</td>
<td>tidal volume, PEEP, rate</td>
<td>NR</td>
<td>NR</td>
</tr>
</tbody>
</table>

Note: ± denotes standard deviation.


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

<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Diseases</th>
<th>Device/mode</th>
<th>Model and Manufacturer</th>
<th>Device Characteristics</th>
<th>Prescribed Usage (frequency and duration)</th>
<th>Actual Usage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hazenberg, 2014&lt;sup&gt;11&lt;/sup&gt; RCT</td>
<td>NMD, TRD</td>
<td>HMV (pressure controlled ventilation) with change to HMV (volume assist control ventilation) if not tolerated</td>
<td>Elisee 150 (ResMed; Paris, France) (FDA approved)</td>
<td>HMV (pressure controlled ventilation): inspiratory pressure, PEEP, rate, inspiratory time HMV (volume assist control ventilation): tidal volume, PEEP, rate</td>
<td>≥ 6 hours/night</td>
<td>-IPAP: 10 cm H2O (pressure mode) -EPAP: 4 cm H2O (pressure mode) -Tidal volume: 8-10 ml/kg (pressure mode)</td>
</tr>
</tbody>
</table>
| Crespo, 2009<sup>17</sup> Retrospective cohort | COPD, TRD, NMD, OHS, Other | HMV (pressure or volume controlled NOS) | NR | NR | NR | Age ≥ 75 years old -IPAP: 14-20 cm H2O -EPAP: 3-8 cmH2O -Tidal volume: 500-
<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Diseases</th>
<th>Device/mode</th>
<th>Model and Manufacturer</th>
<th>Device Characteristics</th>
<th>Prescribed Usage (frequency and duration)</th>
<th>Actual Usage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Munoz, 2005&lt;sup&gt;11&lt;/sup&gt; Retrospective cohort</td>
<td>NMD, TRD</td>
<td>HMV (volume assist control ventilation)</td>
<td>NR</td>
<td>tidal volume, PEEP, rate</td>
<td>NR</td>
<td>tidal volume: 9.5 ± 0.7 ml/kg&lt;br&gt;-Rate: 16.8 ± 2.7 breaths/minute</td>
</tr>
<tr>
<td></td>
<td></td>
<td>HMV (volume control)</td>
<td>NR</td>
<td>tidal volume, PEEP, rate</td>
<td>NR</td>
<td>Tidal volume: 8.61 ± 1.6 ml/kg&lt;br&gt;-Rate: 16.7 ± 2.7 breaths/minute</td>
</tr>
<tr>
<td>Chiang, 2003&lt;sup&gt;12&lt;/sup&gt; RCT</td>
<td>COPD, Other</td>
<td>BPAP NOS</td>
<td>NR</td>
<td>IPAP, EPAP</td>
<td>NR</td>
<td>-IPAP: 11.8 ± 0.6 cm H2O&lt;br&gt;-EPAP: 4.5 ± 0.4 cm H2O</td>
</tr>
<tr>
<td>Windisch, 2006&lt;sup&gt;12&lt;/sup&gt; Observational study</td>
<td>HMV (pressure controlled ventilation)</td>
<td>PV401(Breas Medical AB; Moelnlycke, Sweden) (FDA approved)</td>
<td>inspiratory pressure, PEEP, inspiratory time, rate</td>
<td>NR</td>
<td>-IPAP: 23.2 ± 2.8 mbar&lt;br&gt;-Rate: 20.5 ± 1.9 breaths/minute&lt;br&gt;-Inspiratory time 1.2 ± 0.1 seconds</td>
<td></td>
</tr>
</tbody>
</table>

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>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Device/Mode</th>
<th>Respiratory Services delivered in the home (including by whom and how frequently)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Murphy, 2017(^{11}) RCT</td>
<td>BPAP ST</td>
<td>-Smoking cessation NOS &lt;br&gt;-COPD education NOS</td>
</tr>
<tr>
<td>Oscroft, 2014(^{16}) RCT</td>
<td>BPAP iVAPS versus BPAP ST</td>
<td>-24 hour hotline NOS</td>
</tr>
<tr>
<td>Bhatt, 2013(^{1}) RCT</td>
<td>BPAP NOS</td>
<td>-Daily phone call by respiratory therapist during first week &lt;br&gt;-One home visit by respiratory therapist during first week</td>
</tr>
<tr>
<td>Duiverman, 2011(^{21,22}) RCT</td>
<td>BPAP ST</td>
<td>-Supervision by specialized nurse NOS</td>
</tr>
<tr>
<td>Oscroft, 2010(^{10}) Retrospective Observational</td>
<td>BPAP ST started after AECOPD versus BPAP ST started in stable patient without exacerbation</td>
<td>-24 hour hotline NOS</td>
</tr>
<tr>
<td>Crespo, 2009(^{17}) Retrospective Observational</td>
<td>HMV (pressure or volume NOS)</td>
<td>-Emergency phone number</td>
</tr>
<tr>
<td>Cheung, 2010(^{11}) RCT</td>
<td>BPAP ST versus CPAP</td>
<td>-Nurse hotline NOS</td>
</tr>
<tr>
<td>McEvoy, 2009(^{40}) RCT</td>
<td>BPAP S</td>
<td>-Telephone calls answered by nurses as needed</td>
</tr>
<tr>
<td>Casanova, 2000(^{7}) RCT</td>
<td>BPAP S</td>
<td>-“Close contact was maintained” for first 3 weeks</td>
</tr>
<tr>
<td>Garrod, 2000(^{38}) RCT</td>
<td>BPAP S</td>
<td>-Phone call every 2 weeks to encourage use</td>
</tr>
<tr>
<td>Clini, 1996(^{13}) Prospective Observational</td>
<td>BPAP ST</td>
<td>-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).</td>
</tr>
<tr>
<td>Köhnlein, 2014(^{35}) RCT</td>
<td>BPAP ST</td>
<td>-24 hour hotline staffed by health-care providers and specialized nurses</td>
</tr>
<tr>
<td>Gay, 1996(^{32}) RCT</td>
<td>BPAP ST</td>
<td>-Regular phone calls to ensure compliance.</td>
</tr>
<tr>
<td>Durao, 2018(^{34}) RCT</td>
<td>HMV/BPAP mix</td>
<td>-Smoking cessation NOS</td>
</tr>
</tbody>
</table>

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

Table F.25. Thoracic Restrictive Disorders – Respiratory services

<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Device/Mode</th>
<th>Respiratory Services delivered in the home (including by whom and how frequently)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Doménech-Clar, 2003&lt;sup&gt;19&lt;/sup&gt; Prospective Observational</td>
<td>BPAP NOS</td>
<td>-Telephone helpline (24 hours)</td>
</tr>
</tbody>
</table>

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

<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Device/Mode</th>
<th>Respiratory Services delivered in the home (including by whom and how frequently)</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sanjuan-López, 2014&lt;sup&gt;14&lt;/sup&gt; Retrospective Observational</td>
<td>HMV (PSV or ST)</td>
<td>-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 &lt; 270 l/minute despite assisted cough physiotherapy.</td>
<td></td>
</tr>
<tr>
<td>Pinto, 2010&lt;sup&gt;19&lt;/sup&gt; Prospective Observational</td>
<td>BPAP ST + weekly telemonitoring versus BPAP ST without weekly telemonitoring</td>
<td>-Telephone helpline</td>
<td>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).</td>
</tr>
<tr>
<td>Doménech-Clar, 2003&lt;sup&gt;19&lt;/sup&gt; Prospective Observational</td>
<td>BPAP NOS</td>
<td>-Telephone helpline (24 hours)</td>
<td></td>
</tr>
<tr>
<td>Nauffal, 2002&lt;sup&gt;14&lt;/sup&gt; Prospective</td>
<td>BPAP NOS</td>
<td>-Telephone helpline (24 hours)</td>
<td></td>
</tr>
<tr>
<td>Author, Year, Study Design</td>
<td>Device/Mode</td>
<td>Respiratory Services delivered in the home (including by whom and how frequently)</td>
<td>Outcome</td>
</tr>
<tr>
<td>-----------------------------</td>
<td>-------------</td>
<td>---------------------------------------------------------------------------------</td>
<td>---------</td>
</tr>
<tr>
<td>Gonzalez-Bermejo, 2013&lt;sup&gt;50&lt;/sup&gt; Retrospective Observational</td>
<td>BPAP ST</td>
<td>-Instruction on assisted cough techniques including mechanical insufflation-exsufflation by a respiratory physiotherapist</td>
<td></td>
</tr>
<tr>
<td>Sancho, 2014&lt;sup&gt;52&lt;/sup&gt; Retrospective Observational</td>
<td>HMV (volume cycled) versus BPAP ST</td>
<td>-Guideline based multidisciplinary care, management of cough impairment when necessary, nutritional support, and medical treatment with riluzole.</td>
<td></td>
</tr>
<tr>
<td>Coco, 2006&lt;sup&gt;16&lt;/sup&gt; Prospective Observational</td>
<td>BPAP ST</td>
<td>-Suction devices for secretion clearance -All patients were also taught assisted cough techniques by an experienced respiratory physiotherapist, including mechanical insufflators-exsufflators.</td>
<td></td>
</tr>
<tr>
<td>Bourke, 2006&lt;sup&gt;6&lt;/sup&gt; RCT</td>
<td>BPAP ST</td>
<td>-Multidisciplinary clinical team review, education about assisted cough techniques, posture, bed raisers, adjustable beds, palliative care, hospice as needed.</td>
<td></td>
</tr>
<tr>
<td>Sancho, 2017&lt;sup&gt;53&lt;/sup&gt;</td>
<td>HMV (volume assist control ventilation)</td>
<td>-Guideline based multidisciplinary care, management of cough impairment when necessary, nutritional support, and medical treatment with riluzole.</td>
<td></td>
</tr>
</tbody>
</table>

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

<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Device/Mode</th>
<th>Respiratory Services delivered in the home (including by whom and how frequently)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Masa, 2015&lt;sup&gt;18&lt;/sup&gt; RCT</td>
<td>Mixed: HMV and BPAP mix (all with bilevel pressure with assured volume)</td>
<td>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).</td>
</tr>
<tr>
<td>Borel, 2011&lt;sup&gt;5&lt;/sup&gt; RCT</td>
<td>BPAP ST</td>
<td>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.</td>
</tr>
</tbody>
</table>

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


<table>
<thead>
<tr>
<th>Author, Year, Study Design</th>
<th>Diseases</th>
<th>Device/Mode</th>
<th>Respiratory Services delivered in the home (including by whom and how frequently)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Munoz, 2005&lt;sup&gt;41&lt;/sup&gt; Retrospective Observational</td>
<td>NMD/TRD</td>
<td>HMV volume assist/control mode versus HMV volume control mode</td>
<td>-Telephone helpline</td>
</tr>
<tr>
<td>Chiang, 2003&lt;sup&gt;12&lt;/sup&gt; RCT</td>
<td>COPD, other</td>
<td>BPAP NOS</td>
<td>-Telephone interviews by respiratory therapist every 2 weeks to assess compliance and ventilator usage.</td>
</tr>
</tbody>
</table>

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

## Appendix G. Guidelines

**Table G.1. Guidelines for all conditions**

<table>
<thead>
<tr>
<th>Organization</th>
<th>Topic</th>
<th>KQ</th>
<th>Statement</th>
</tr>
</thead>
</table>
| Agency for Clinical Innovation, Australia, Domiciliary Non-Invasive Ventilation in Adult Patients, 2012<sup>44</sup> | Device initiation criteria                                          | KQ1 | Generally NIV should be commenced when there is evidence of: Daytime hypercapnia, PaCO₂ ≥45mmHg and/or Evidence of nocturnal hypventilation (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, Domiciliary Non-Invasive Ventilation in Adult Patients, 2012<sup>44</sup> | 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 gogginess, 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<sup>15</sup> | 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. |
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.

---

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.

---

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)
<table>
<thead>
<tr>
<th>Organization</th>
<th>Topic</th>
<th>KQ</th>
<th>Statement</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rep, American Academy of Home Care Physicians, American College of Chest</td>
<td>Clinical Indications for Noninvasive Positive Pressure Ventilation in</td>
<td>KQ1 and</td>
<td>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. 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.</td>
</tr>
<tr>
<td>Physicians, American College of Physicians, American Sleep Disorders</td>
<td>Chronic Respiratory Failure due to Restrictive Lung Disease, COPD, and</td>
<td>KQ2</td>
<td></td>
</tr>
<tr>
<td>Association, American Thoracic Society, National Association for Medical</td>
<td>Nocturnal Hypoventilation—A Consensus Conference Report, American</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Direction of Respiratory Care, 199967</td>
<td>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, 199967</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Agency for Clinical Innovation, Australia, Domiciliary Non-Invasive</td>
<td>Device continuation, compliance, and outcomes</td>
<td>KQ1 and</td>
<td>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 &gt; 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.</td>
</tr>
<tr>
<td>Ventilation in Adult Patients, 201264</td>
<td></td>
<td>KQ2</td>
<td></td>
</tr>
<tr>
<td>Organization</td>
<td>Topic</td>
<td>KQ</td>
<td>Statement</td>
</tr>
<tr>
<td>----------------------------------------------------------------------------</td>
<td>----------------------------------------------------------------------</td>
<td>----------</td>
<td>----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------</td>
</tr>
<tr>
<td>German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010&lt;sup&gt;th&lt;/sup&gt;</td>
<td>Device continuation, compliance, and outcomes</td>
<td>KQ1 and KQ2</td>
<td>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.</td>
</tr>
<tr>
<td>Agency for Clinical Innovation, Australia, Domiciliary Non-Invasive Ventilation in Adult Patients, 2012&lt;sup&gt;nd&lt;/sup&gt;</td>
<td>Device characteristics and titration</td>
<td>KQ3</td>
<td>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&gt;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.</td>
</tr>
<tr>
<td>British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory</td>
<td>Device characteristics and titration</td>
<td>KQ3</td>
<td>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.</td>
</tr>
<tr>
<td>Organization</td>
<td>Topic</td>
<td>KQ</td>
<td>Statement</td>
</tr>
<tr>
<td>----------------------------------------------------------------------------</td>
<td>----------------------------------------------------------------------</td>
<td>-------</td>
<td>---------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------</td>
</tr>
<tr>
<td>Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[69]</td>
<td></td>
<td></td>
<td>NIV circuits must allow adequate clearance of exhaled air through an exhalation valve or an integral exhalation port on the mask. As patients recover from acute hypercapnic respiratory failure, ventilator requirements change and ventilator settings should be reviewed regularly.</td>
</tr>
<tr>
<td>German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010[68]</td>
<td>Device characteristics and titration</td>
<td>KQ3</td>
<td>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.</td>
</tr>
<tr>
<td>Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[65]</td>
<td>Respiratory services</td>
<td>KQ4</td>
<td>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 &lt;270 L/min. Manually assisted coughing is recommended alone or in addition to lung volume recruitment to increase peak cough flows to &gt;270 L/min. In the absence of contraindications, mechanical in-exsufflation should be recommended for patients unable to achieve peak cough flows &gt;270 L/min with lung volume recruitment and/or manually assisted coughing, particularly during respiratory infection.</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Organization</th>
<th>Topic</th>
<th>KQ</th>
<th>Statement</th>
</tr>
</thead>
<tbody>
<tr>
<td>German Society for Pneumology, Guidelines for Non-Invasive and Invasive</td>
<td>Device initiation criteria</td>
<td>KQ1</td>
<td>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:</td>
</tr>
<tr>
<td>Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010</td>
<td></td>
<td></td>
<td>- Chronic daytime hypercapnia with PaCO2 ≥ 50mmHg</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>- Nocturnal hypercapnia with PaCO2 ≥ 55mmHg</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>- Stable daytime hypercapnia with PaCO2 46–50mmHg and a rise in P tcCO2 to ≥ 10mmHg during sleep.</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>- 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</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>- Following an acute exacerbation needing ventilatory support, according to clinical estimation.</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>- Poor compliance with medication intake and/or LTOT are relative contraindications. Complete discontinuation of nicotine abuse should be aspired to.</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>- NIV is the primary treatment option for HMV of COPD patients with CRF.</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>- 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.</td>
</tr>
<tr>
<td>Agency for Clinical Innovation,</td>
<td>Device initiation criteria</td>
<td>KQ1</td>
<td>Nocturnal NIV is indicated in COPD with PaCO2 &gt; 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.</td>
</tr>
<tr>
<td>Australia, Domiciliary Non-Invasive Ventilation in Adult Patients, 2012</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Canadian Thoracic Society,</td>
<td>Device initiation criteria</td>
<td>KQ1</td>
<td>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 &gt;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.</td>
</tr>
<tr>
<td>Home Mechanical Ventilation Clinical Practice Guideline, 2011</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>8th International Conference on Management and Rehabilitation of Chronic</td>
<td>Device initiation criteria</td>
<td>KQ1</td>
<td>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.</td>
</tr>
<tr>
<td>Respiratory Failure, Pescara, Italy, 2015</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Clinical Indications for Noninvasive Positive Pressure Ventilation in Chronic</td>
<td>Device initiation criteria</td>
<td>KQ1</td>
<td>Indications for usage: symptoms (such as fatigue, dyspnea, morning headache, etc.) and physiologic criteria (one of the following): PaCO2 &gt; 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</td>
</tr>
<tr>
<td>Respiratory Failure due to Restrictive Lung Disease, COPD, and Nocturnal</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hypoventilation—A Consensus</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

G-6
<table>
<thead>
<tr>
<th>Organization</th>
<th>Topic</th>
<th>KQ</th>
<th>Statement</th>
</tr>
</thead>
</table>
| 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<sup>67</sup> | 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. |
<p>| British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016&lt;sup&gt;69&lt;/sup&gt; | 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. |
| Agency for Clinical Innovation, Australia, Domiciliary Non-Invasive Ventilation in Adult Patients, 2012&lt;sup&gt;64&lt;/sup&gt; | 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. |</p>
<table>
<thead>
<tr>
<th>Organization</th>
<th>Topic</th>
<th>KQ</th>
<th>Statement</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clearinghouse Clinical Practice Guideline for the Management of Chronic Obstructive Pulmonary Disease, 2014&lt;sup&gt;71&lt;/sup&gt;</td>
<td>Device initiation, monitoring, and candidate selection</td>
<td>KQ1</td>
<td>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.</td>
</tr>
<tr>
<td>United Kingdom National Institute for Health and Care Excellence (NICE), Chronic Obstructive Pulmonary Disease in Over 16s: Diagnosis and Management, 2010&lt;sup&gt;72&lt;/sup&gt;</td>
<td>Device initiation, monitoring, and candidate selection</td>
<td>KQ1</td>
<td>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.</td>
</tr>
<tr>
<td>Agency for Clinical Innovation, Australia, Domiciliary Non-Invasive Ventilation in Adult Patients, 2012&lt;sup&gt;64&lt;/sup&gt;</td>
<td>Device continuation, compliance, and outcomes</td>
<td>KQ1 and KQ2</td>
<td>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.</td>
</tr>
<tr>
<td>German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010&lt;sup&gt;68&lt;/sup&gt;</td>
<td>Device characteristics and titration</td>
<td>KQ3</td>
<td>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.</td>
</tr>
<tr>
<td>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&lt;sup&gt;67&lt;/sup&gt;</td>
<td>Device characteristics and titration</td>
<td>KQ3</td>
<td>Telemonitoring in ventilator dependent patients: 1) Home mechanical ventilators may be equipped with</td>
</tr>
<tr>
<td>8th International Conference</td>
<td>Respiratory</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Organization</td>
<td>Topic</td>
<td>KQ</td>
<td>Statement</td>
</tr>
<tr>
<td>--------------</td>
<td>-------</td>
<td>----</td>
<td>-----------</td>
</tr>
<tr>
<td>on Management and Rehabilitation of Chronic Respiratory Failure, Pescara, Italy, 2015*</td>
<td>services</td>
<td></td>
<td>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</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Organization</th>
<th>Topic</th>
<th>KQ</th>
<th>Statement</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Agency for Clinical Innovation, Australia, Domiciliary Non-Invasive Ventilation in Adult Patients, 2012</strong></td>
<td>Device initiation criteria</td>
<td>KQ1</td>
<td>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).</td>
</tr>
<tr>
<td><strong>British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016</strong></td>
<td>Device initiation criteria</td>
<td>KQ1</td>
<td>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 &lt;1 L and RR &gt;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.</td>
</tr>
<tr>
<td><strong>United Kingdom National Institute for Health and Care Excellence (NICE), Motor Neuron Disease: Assessment and Management, 2016</strong></td>
<td>Device initiation criteria</td>
<td>KQ1</td>
<td>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.</td>
</tr>
<tr>
<td>Organization</td>
<td>Topic</td>
<td>KQ</td>
<td>Statement</td>
</tr>
<tr>
<td>----------------------------------------------------------------------------</td>
<td>----------------------------------------------------------------------</td>
<td>------</td>
<td>-------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------</td>
</tr>
<tr>
<td>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</td>
<td>Device initiation criteria</td>
<td>KQ1</td>
<td>Indications for usage&lt;br&gt;Symptoms (such as fatigue, dyspnea, morning headache, etc.) and one of the following Physiologic criteria (one of the following&lt;br&gt;( \text{PaCO}_2 \geq 45 \text{ mm Hg} )&lt;br&gt;Nocturnal oximetry demonstrating oxygen saturation ( \leq 88% ) for 5 consecutive minutes&lt;br&gt;For progressive neuromuscular disease, maximal inspiratory pressures ( &lt; 60 \text{ cm H}_2\text{O} ) or ( \text{FVC} &lt; 50% ) predicted</td>
</tr>
<tr>
<td>Agency for Clinical Innovation, Australia, Domiciliary Non-Invasive Ventilation in Adult Patients, 2012</td>
<td>Device initiation criteria</td>
<td>KQ1</td>
<td>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).</td>
</tr>
<tr>
<td>German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010</td>
<td>Device initiation criteria</td>
<td>KQ1</td>
<td>NIV of NMD patients with clinical signs of CRF is indicated by the following (at least 1 criterion should be fulfilled):&lt;br&gt;Chronic daytime hypercapnia with ( \text{PaCO}_2 \geq 45\text{mmHg} )&lt;br&gt;Nocturnal hypercapnia with ( \text{PaCO}_2 \geq 50\text{mmHg} )&lt;br&gt;Daytime normocapnia with a rise in ( \text{PTcCO}_2 ) of ( \geq 10\text{mmHg} ) during the night&lt;br&gt;A rapid, significant reduction in VC&lt;br&gt;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.&lt;br&gt;NIV is also indicated prior to elective vertebral column correction surgery when VC &lt; 60% target value and ( \text{FEV1} &lt; 40% ) target value, respectively, or during pregnancy with restricted lung function, as well as palliative care of dyspnea.&lt;br&gt;Patients with NMD should undergo clinical assessment and assessment of VC at 3–12 month-intervals.&lt;br&gt;Polygraphy and PTcCO2-measurement are indicated when VC is ( &lt; 70% ).&lt;br&gt;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.&lt;br&gt;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.</td>
</tr>
<tr>
<td>Organization</td>
<td>Topic</td>
<td>KQ</td>
<td>Statement</td>
</tr>
<tr>
<td>-----------------------------------------------------------------------------</td>
<td>----------------------------------------------------------------------</td>
<td>--------</td>
<td>----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------</td>
</tr>
<tr>
<td>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</td>
<td>Device initiation Criteria (ALS)</td>
<td>KQ1</td>
<td>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.</td>
</tr>
<tr>
<td>Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011</td>
<td>Device initiation Criteria (ALS)</td>
<td>KQ1</td>
<td>NIV should be offered to patients with any one of the following: Orthopnea Daytime hypercapnia Symptomatic sleep disordered breathing FVC &lt;50% predicted SNP &lt;40 cmH2O or PImax&lt;40 cmH2O</td>
</tr>
<tr>
<td>American Thoracic Society, Respiratory Care of the Patient with Duchenne Muscular Dystrophy, 2004</td>
<td>Device initiation criteria (Duchenne Muscular Dystrophy)</td>
<td>KQ1</td>
<td>Consider daytime ventilation when measured waking Pco2 exceeds 50 mm Hg or when hemoglobin saturation remains &lt; 92% while awake.</td>
</tr>
<tr>
<td>Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011</td>
<td>Device initiation criteria (Duchenne Muscular Dystrophy)</td>
<td></td>
<td>Offer nocturnal NIV to patients with diurnal hypercapnia (daytime arterial PCO2 &gt;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.</td>
</tr>
<tr>
<td>German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010</td>
<td>Device initiation, monitoring, and candidate selection</td>
<td>KQ1</td>
<td>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</td>
</tr>
<tr>
<td>British Thoracic Society, Guidelines for Home Oxygen Use in Adults, 2015</td>
<td>Device initiation, monitoring, and candidate selection</td>
<td>KQ1</td>
<td>Non-invasive ventilation (NIV) should be the treatment of choice for patients with NMD or chest wall disease causing type 2 respiratory failure.</td>
</tr>
<tr>
<td>Organization</td>
<td>Topic</td>
<td>KQ</td>
<td>Statement</td>
</tr>
<tr>
<td>----------------------------------------------------------------------------</td>
<td>----------------------------------------------------------------------</td>
<td>-----</td>
<td>----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------</td>
</tr>
</tbody>
</table>
| British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016<sup>69</sup> | 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. |
| United Kingdom National Institute for Health and Care Excellence (NICE), Motor Neuron Disease: Assessment and Management, 2016<sup>73</sup> | 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:  
ox 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. |

G-13
<table>
<thead>
<tr>
<th>Organization</th>
<th>Topic</th>
<th>KQ</th>
<th>Statement</th>
</tr>
</thead>
</table>
| 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 | 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. |
<p>| British Thoracic Society, Guidelines for Home Oxygen Use in Adults, 2015   | 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.                                                                                                                                                                           |</p>
<table>
<thead>
<tr>
<th>Organization</th>
<th>Topic</th>
<th>KQ</th>
<th>Statement</th>
</tr>
</thead>
</table>
| Agency for Clinical Innovation, Australia, Domiciliary Non-Invasive Ventilation in Adult Patients, 2012* | 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.  
Slowly progressive NMD  
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.  
Rapidly progressive NMD  
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. |
| Agency for Clinical Innovation, Australia, Domiciliary Non-Invasive Ventilation in Adult Patients, 2012* | 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.  
Slowly progressive NMD  
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.  
Rapidly progressive NMD  
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. |
<table>
<thead>
<tr>
<th>Organization</th>
<th>Topic</th>
<th>KQ</th>
<th>Statement</th>
</tr>
</thead>
<tbody>
<tr>
<td>European Federation of Neurological Societies (EFNS)</td>
<td>Device initiation, monitoring, and candidate selection (ALS)</td>
<td>KQ1</td>
<td>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.</td>
</tr>
<tr>
<td>Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011</td>
<td>Device initiation, monitoring, and candidate selection (ALS)</td>
<td>KQ1</td>
<td>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.</td>
</tr>
<tr>
<td>Organization</td>
<td>Topic</td>
<td>KQ</td>
<td>Statement</td>
</tr>
<tr>
<td>----------------------------------------------------------------------------</td>
<td>------------------------------------------------------------</td>
<td>-----</td>
<td>--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------</td>
</tr>
<tr>
<td>American Thoracic Society, Respiratory Care of the Patient with Duchenne Muscular Dystrophy, 2004[^13]</td>
<td>Device initiation, monitoring, and candidate selection (Duchenne Muscular Dystrophy)</td>
<td>KQ1</td>
<td>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.</td>
</tr>
<tr>
<td>Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[^13]</td>
<td>Device initiation, monitoring, and candidate selection (Duchenne Muscular Dystrophy)</td>
<td>KQ1</td>
<td>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 &lt;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 &lt;40% predicted.</td>
</tr>
<tr>
<td>Organization</td>
<td>Topic</td>
<td>KQ</td>
<td>Statement</td>
</tr>
<tr>
<td>------------------------------------------------------------------------------</td>
<td>----------------------------------------------------------------------</td>
<td>------</td>
<td>------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------</td>
</tr>
<tr>
<td>Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011</td>
<td>Device initiation, monitoring, and candidate selection (Other myopathies)</td>
<td>KQ1</td>
<td>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 &lt;60%. Consider ABGs or nocturnal measure of CO2 in patients with VC &lt;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.</td>
</tr>
<tr>
<td>Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011</td>
<td>Device initiation, monitoring, and candidate selection (Myotonic dystrophy)</td>
<td>KQ1</td>
<td>Obtain six to 12 monthly clinical assessment of symptoms of daytime or nocturnal hypoventilation. Obtain yearly VC and consider daytime PaCO2 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.</td>
</tr>
<tr>
<td>Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011</td>
<td>Device initiation, monitoring, and candidate selection (Post-polio syndrome)</td>
<td>KQ1</td>
<td>Yearly assessment of VC is recommended from the time of presentation of post polio syndrome. If VC &gt;50% with symptoms of hypoventilation, perform measurements of daytime ABGs, overnight oximetry and consider polysomnography. When VC &lt;50%, perform ABG analysis and/or nocturnal oximetry yearly. With confirmation of the presence of chronic hypoventilation, offer NIV.</td>
</tr>
<tr>
<td>Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011</td>
<td>Device initiation, monitoring, and candidate selection (Spinal cord injury)</td>
<td>KQ1</td>
<td>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.</td>
</tr>
<tr>
<td>Agency for Clinical Innovation, Australia, Domiciliary Non-Invasive Ventilation in Adult Patients, 2012</td>
<td>Device initiation, monitoring, and candidate selection (Spinal cord injury)</td>
<td>KQ1</td>
<td>NIV is indicated when there is intractable or refractory sputum retention, atelectasis, respiratory tract infection or type-I respiratory failure (PaO2 &lt; 80 mmHg, SpO2 &lt;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.</td>
</tr>
<tr>
<td>Organization</td>
<td>Topic</td>
<td>KQ</td>
<td>Statement</td>
</tr>
<tr>
<td>----------------------------------------------------------------------------</td>
<td>----------------------------------------------------------------------</td>
<td>----</td>
<td>----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------</td>
</tr>
<tr>
<td>Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011</td>
<td>Device characteristics and titration (Duchenne Muscular Dystrophy)</td>
<td>KQ3</td>
<td>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.</td>
</tr>
<tr>
<td>British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016</td>
<td>Device characteristics and titration</td>
<td>KQ3</td>
<td>In patients with NMD or CWD, consider controlled ventilation as triggering may be ineffective.</td>
</tr>
<tr>
<td>Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011</td>
<td>Device characteristics and titration (ALS)</td>
<td>KQ3</td>
<td>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.</td>
</tr>
<tr>
<td>Agency for Clinical Innovation, Australia, Domiciliary Non-Invasive Ventilation in Adult Patients, 2012</td>
<td>Respiratory services</td>
<td>KQ4</td>
<td>Ability to generate PCF of at least 160 L/min is necessary for non-invasive management of pulmonary secretions. Baseline assisted PCF &lt;270 L/min are likely to decrease to &lt;160 L/min during chest infections, increasing the likelihood of pneumonia and respiratory failure. Patients with a baseline PCF &lt; 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 &lt; 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 &lt; 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 &gt;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.</td>
</tr>
<tr>
<td>German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010</td>
<td>Respiratory services</td>
<td>KQ4</td>
<td>A reduced cough impulse (peak cough flow: PCF &lt; 270 l/min) can lead to acute decompensations and increased incidence of aspiration pneumonia. Measures to eliminate secretions should therefore be taken when SaO2&lt; 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 &lt; 270 l/min) indicates the need for the initiation of secretion management.</td>
</tr>
<tr>
<td>Organization</td>
<td>Topic</td>
<td>KQ</td>
<td>Statement</td>
</tr>
<tr>
<td>--------------</td>
<td>-------</td>
<td>----</td>
<td>-----------</td>
</tr>
<tr>
<td>British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016</td>
<td>Respiratory services</td>
<td>KQ4</td>
<td>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.</td>
</tr>
<tr>
<td>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</td>
<td>Respiratory services</td>
<td>KQ4</td>
<td>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.</td>
</tr>
<tr>
<td>United Kingdom National Institute for Health and Care Excellence (NICE), Motor Neuron Disease: Assessment and Management, 2016</td>
<td>Respiratory services</td>
<td>KQ4</td>
<td>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.</td>
</tr>
<tr>
<td>Canadian Thoracic Society 2011</td>
<td>Respiratory services (ALS)</td>
<td>KQ4</td>
<td>Lung volume recruitment maneuvers should be introduced with declining VC. Methods to assist secretion clearance should be initiated when PCF is &lt;4.25 L/s or the Norris bulbar core is &lt;29.</td>
</tr>
<tr>
<td>Organization</td>
<td>Topic</td>
<td>KQ</td>
<td>Statement</td>
</tr>
<tr>
<td>--------------</td>
<td>-------</td>
<td>----</td>
<td>-----------</td>
</tr>
<tr>
<td>European Federation of Neurological Societies (EFNS) Guidelines on the Clinical Management of Amyotrophic Lateral Sclerosis, 2012&lt;sup&gt;16&lt;/sup&gt;</td>
<td>Respiratory services (ALS)</td>
<td>KQ4</td>
<td>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 (&gt;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.</td>
</tr>
<tr>
<td>American Thoracic Society, Respiratory Care of the Patient with Duchenne Muscular Dystrophy, 2004&lt;sup&gt;73&lt;/sup&gt;</td>
<td>Respiratory services (Duchenne Muscular Dystrophy)</td>
<td>KQ4</td>
<td>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.</td>
</tr>
<tr>
<td>Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011&lt;sup&gt;63&lt;/sup&gt;</td>
<td>Respiratory services (Duchenne Muscular Dystrophy)</td>
<td>KQ4</td>
<td>Lung volume recruitment maneuvers should be introduced with declining VC. Methods to assist secretion clearance should be initiated when PCF &lt;270 L/min.</td>
</tr>
<tr>
<td>Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011&lt;sup&gt;63&lt;/sup&gt;</td>
<td>Respiratory services (All NMD except ALS and Duchenne Muscular Dystrophy)</td>
<td>KQ4</td>
<td>Assess airway clearance ability with peak cough flows and implement cough-assistance strategies</td>
</tr>
</tbody>
</table>
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.
## Table G.4. Guidelines for Thoracic Restrictive Disorders

<table>
<thead>
<tr>
<th>Organization</th>
<th>Topic</th>
<th>KQ</th>
<th>Statement</th>
</tr>
</thead>
<tbody>
<tr>
<td>British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016&lt;sup&gt;69&lt;/sup&gt;</td>
<td>Device initiation criteria</td>
<td>KQ1</td>
<td>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 &lt;1 L and RR &gt;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.</td>
</tr>
<tr>
<td>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&lt;sup&gt;15&lt;/sup&gt;</td>
<td>Device initiation criteria</td>
<td>KQ1</td>
<td>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 &lt; 60 cmH2O or FVC &lt;50% predicted</td>
</tr>
<tr>
<td>Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011&lt;sup&gt;14&lt;/sup&gt;</td>
<td>Device initiation criteria</td>
<td>KQ1</td>
<td>Patients with kyphoscoliosis should undergo periodical spirometry testing, and if FVC is &lt;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.</td>
</tr>
<tr>
<td>Agency for Clinical Innovation, Australia, Domiciliary Non-Invasive Ventilation in Adult Patients, 2012&lt;sup&gt;64&lt;/sup&gt;</td>
<td>Device initiation criteria</td>
<td>KQ1</td>
<td>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.</td>
</tr>
<tr>
<td>Organization</td>
<td>Topic</td>
<td>KQ</td>
<td>Statement</td>
</tr>
<tr>
<td>----------------------------------------------------------------------------</td>
<td>--------------------------------------------</td>
<td>----</td>
<td>-------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------</td>
</tr>
<tr>
<td>German Society for Pneumology, Guidelines for Non-Invasive and Invasive</td>
<td>Device initiation criteria</td>
<td>KQ1</td>
<td>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 PaCO2 ≥ 45mmHg Nocturnal hypercapnia with PaCO2 ≥ 50mmHg Daytime normocapnia with a rise in PTcCO2 of ≥ 10mmHg during the night Patients without manifest hypercapnia but with severe, restrictive ventilatory dysfunction (VC &lt; 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.</td>
</tr>
<tr>
<td>Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>British Thoracic Society, Guidelines for Home Oxygen Use in Adults, 2015</td>
<td>Device initiation, monitoring, and candidate selection</td>
<td>KQ1</td>
<td>Non-invasive ventilation (NIV) should be the treatment of choice for patients with NMD or chest wall disease causing type 2 respiratory failure.</td>
</tr>
<tr>
<td>British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016</td>
<td>Device initiation, monitoring, and candidate selection</td>
<td>KQ1</td>
<td>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.</td>
</tr>
<tr>
<td>Organization</td>
<td>Topic</td>
<td>KQ</td>
<td>Statement</td>
</tr>
<tr>
<td>----------------------------------------------------------------------------</td>
<td>----------------------------------------------------------------------</td>
<td>----------</td>
<td>----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------</td>
</tr>
</tbody>
</table>
| 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<sup>67</sup> | 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<sup>66</sup> | 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<sup>69</sup> | Device characteristics and titration                                | KQ3      | In patients with NMD or CWD, consider controlled ventilation as triggering may be ineffective.                                                                                                             |
| Agency for Clinical Innovation, Australia, Domiciliary Non-Invasive Ventilation in Adult Patients, 2012<sup>64</sup> | 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<sup>68</sup> | 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. |
<table>
<thead>
<tr>
<th>Organization</th>
<th>Topic</th>
<th>KQ</th>
<th>Statement</th>
</tr>
</thead>
<tbody>
<tr>
<td>British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016*</td>
<td>Respiratory services</td>
<td>KQ4</td>
<td>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.</td>
</tr>
<tr>
<td>Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011*</td>
<td>Respiratory services</td>
<td>KQ4</td>
<td>Methods to assist secretion clearance should be initiated when peak cough flow is &lt;270 L/min</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Table G.5. Guidelines for Obesity Hypoventilation Syndrome</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Organization</strong></td>
</tr>
<tr>
<td>British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016&lt;sup&gt;69&lt;/sup&gt;</td>
</tr>
<tr>
<td>Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011&lt;sup&gt;63&lt;/sup&gt;</td>
</tr>
<tr>
<td>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&lt;sup&gt;27&lt;/sup&gt;</td>
</tr>
<tr>
<td>Agency for Clinical Innovation, Australia, Domiciliary Non-Invasive Ventilation in Adult Patients, 2012&lt;sup&gt;14&lt;/sup&gt;</td>
</tr>
<tr>
<td>German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for</td>
</tr>
</tbody>
</table>

G-27
<table>
<thead>
<tr>
<th>Organization</th>
<th>Topic</th>
<th>KQ</th>
<th>Statement</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Treatment of Chronic Respiratory Failure, 2010</strong></td>
<td></td>
<td></td>
<td>comparison to the awake state or Desaturations &lt; 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 &gt; 55mmHg and PaCO2 ≥ 10 mmHg, respectively, in comparison to normocapnia during the awake state, or desaturation &lt; 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.</td>
</tr>
<tr>
<td><strong>Agency for Clinical Innovation, Australia, Domiciliary Non-Invasive Ventilation in Adult Patients, 2012</strong></td>
<td>Device initiation, monitoring, and candidate selection</td>
<td>KQ1</td>
<td>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 &gt;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. Auto titrating 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.</td>
</tr>
<tr>
<td><strong>Agency for Clinical Innovation, Australia, Domiciliary Non-Invasive Ventilation in Adult Patients, 2012</strong></td>
<td>Device continuation, compliance, and outcomes</td>
<td>KQ1 and KQ2</td>
<td>Individuals initially using bilevel support should be reviewed again after 3 months on therapy and CPAP retreated, 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.</td>
</tr>
<tr>
<td><strong>German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010</strong></td>
<td>Device characteristics and titration</td>
<td>KQ3</td>
<td>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.</td>
</tr>
<tr>
<td>Organization</td>
<td>Topic</td>
<td>KQ</td>
<td>Statement</td>
</tr>
<tr>
<td>----------------------------------------------------------------------------</td>
<td>----------------------------------------------------------------------</td>
<td>----</td>
<td>----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------</td>
</tr>
<tr>
<td>Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice</td>
<td>Device characteristics and titration (Central hypoventilation syndrome)</td>
<td>KQ3</td>
<td>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.</td>
</tr>
<tr>
<td>Guideline, 2011</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016</td>
<td>Device characteristics and titration</td>
<td>KQ3</td>
<td>High inspiratory positive airway pressure (IPAP) and expiratory positive airway pressure (EPAP) settings are commonly required in patients with OHS (e.g., IPAP&gt;30, EPAP&gt;8). Volume control (or volume assured) modes of providing NIV may be more effective when high inflation pressures are required.</td>
</tr>
<tr>
<td>Agency for Clinical Innovation, Australia, Domiciliary Non-Invasive Ventilation in Adult Patients, 2012</td>
<td>Respiratory services</td>
<td>KQ4</td>
<td>All patients should be advised on appropriate dietary and lifestyle changes to promote weight loss and referred to appropriate programs where possible.</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Organization</th>
<th>Topic</th>
<th>KQ</th>
<th>Statement</th>
</tr>
</thead>
<tbody>
<tr>
<td>British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016&lt;sup&gt;69&lt;/sup&gt;</td>
<td>Device initiation, monitoring, and candidate selection (Asthma)</td>
<td>KQ1</td>
<td>Acute (or acute on chronic) episodes of hypercapnia may complicate chronic asthma. This condition closely resembles COPD and should be managed as such.</td>
</tr>
<tr>
<td>British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016&lt;sup&gt;69&lt;/sup&gt;</td>
<td>Device initiation, monitoring, and candidate selection (Bronchiectasis)</td>
<td>KQ1</td>
<td>In patients with non-CF bronchiectasis, NIV should be started in acute hypercapnic respiratory failure using the same criteria as in AECOPD (pH&lt;7.35 and pCO₂ &gt;6.5 kPa persist or develop despite optimal medical therapy).</td>
</tr>
<tr>
<td>British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016&lt;sup&gt;69&lt;/sup&gt;</td>
<td>Device initiation, monitoring, and candidate selection (Cystic fibrosis)</td>
<td>KQ1</td>
<td>In patients with cystic fibrosis, NIV is the treatment of choice when ventilatory support is needed.</td>
</tr>
<tr>
<td>British Thoracic Society, Guidelines for Home Oxygen Use in Adults, 2015&lt;sup&gt;66&lt;/sup&gt;</td>
<td>Device initiation, monitoring, and candidate selection (Cystic fibrosis)</td>
<td>KQ1</td>
<td>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.</td>
</tr>
<tr>
<td>Agency for Clinical Innovation, Australia, Domiciliary Non-Invasive Ventilation in Adult Patients, 2012&lt;sup&gt;64&lt;/sup&gt;</td>
<td>Device initiation, monitoring, and candidate selection (Cystic fibrosis)</td>
<td>KQ1</td>
<td>Individuals with awake SpO₂&lt;94% or spirometry (FEV₁&lt;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&gt;45mmHg and nocturnal gas exchange shows SpO₂&lt;90% for &gt;5% of TST and/or a rise in TcCO₂ / ETCO₂ from NREM to REM &gt;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.</td>
</tr>
<tr>
<td>Agency for Clinical Innovation,</td>
<td>Device</td>
<td>KQ1</td>
<td>Awake PaCO₂ &gt; 45 mmHg in the absence of lung and chest wall abnormalities, skeletal malformations and</td>
</tr>
<tr>
<td>Organization</td>
<td>Topic</td>
<td>KQ</td>
<td>Statement</td>
</tr>
<tr>
<td>--------------</td>
<td>-------</td>
<td>----</td>
<td>-----------</td>
</tr>
<tr>
<td>Australia, Domiciliary Non-Invasive Ventilation in Adult Patients, 2012&lt;sup&gt;64&lt;/sup&gt;</td>
<td>initiation, monitoring, and candidate selection (Hypercapnic central sleep apnea)</td>
<td>KQ</td>
<td>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 dyspnœa in response to pneumonia may mask severe respiratory compromise.</td>
</tr>
<tr>
<td>British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016&lt;sup&gt;69&lt;/sup&gt;</td>
<td>Respiratory services (Cystic fibrosis)</td>
<td>KQ4</td>
<td>In patients with cystic fibrosis, specialised physiotherapy is needed to aid sputum clearance.</td>
</tr>
</tbody>
</table>

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

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Design</th>
<th>SMD (95% CI)</th>
<th>Weight</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dulverman, 2011</td>
<td></td>
<td>RCT</td>
<td>0.23 (0.01, 0.49)</td>
<td>36.27</td>
</tr>
<tr>
<td>Garnod, 2000</td>
<td></td>
<td>RCT</td>
<td>0.03 (-0.06, 0.61)</td>
<td>10.16</td>
</tr>
<tr>
<td>Struk, 2014</td>
<td></td>
<td>RCT</td>
<td>-0.02 (-0.16, 0.12)</td>
<td>53.67</td>
</tr>
<tr>
<td>Overall (I² = 49.7%, p = 0.153)</td>
<td></td>
<td></td>
<td>0.08 (-0.12, 0.28)</td>
<td>100.00</td>
</tr>
</tbody>
</table>

NOTE: Weights are from random effects analysis

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

<table>
<thead>
<tr>
<th>Author, Year</th>
<th>Study Type</th>
<th>Rate (95% CI)</th>
<th>Weight</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bhatt, 2013</td>
<td>RCT</td>
<td>1.00 (0.06, 15.99)</td>
<td>0.28</td>
</tr>
<tr>
<td>Duhverman, 2011</td>
<td>RCT</td>
<td>1.00 (0.83, 1.21)</td>
<td>60.58</td>
</tr>
<tr>
<td>Struk, 2014</td>
<td>RCT</td>
<td>1.00 (0.76, 1.32)</td>
<td>28.21</td>
</tr>
<tr>
<td>Tsolaki, 2008</td>
<td>Observational</td>
<td>0.77 (0.50, 1.21)</td>
<td>10.94</td>
</tr>
<tr>
<td>Overall (I²-squared = 0.0%, p = 0.768)</td>
<td>0.97 (0.84, 1.13)</td>
<td>100.00</td>
<td></td>
</tr>
</tbody>
</table>

NOTE: Weights are from random effects analysis

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

<table>
<thead>
<tr>
<th>Author</th>
<th>Study Type</th>
<th>OR (95% CI)</th>
<th>Weight</th>
</tr>
</thead>
<tbody>
<tr>
<td>Galli, 2014</td>
<td>Observational</td>
<td>0.31 (0.11, 0.89)</td>
<td>73.20</td>
</tr>
<tr>
<td>Casanova, 2010</td>
<td>RCT</td>
<td>0.48 (0.44, 0.65)</td>
<td>13.43</td>
</tr>
<tr>
<td>Towaki, 2008</td>
<td>Observational</td>
<td>0.38 (0.03, 4.55)</td>
<td>13.37</td>
</tr>
<tr>
<td>Overall</td>
<td></td>
<td>0.34 (0.14, 0.83)</td>
<td>100.00</td>
</tr>
</tbody>
</table>

NOTE: Weights are from random effects analysis

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
Appendix I. References for Appendixes


