

Subject: Noninvasive Positive Pressure Ventilation in the Home Setting for Chronic Respiratory Failure		Original Effective Date: 5/23/2016
Policy Number: MCP-275	Revision Date(s): 12/13/2017, 7/10/18	
Review Date: 6/22/17		
MCPC Approval Date: 12/13/17, 7/16/18		

DISCLAIMER

This Molina Clinical Policy (MCP) is intended to facilitate the Utilization Management process. It expresses Molina's determination as to whether certain services or supplies are medically necessary, experimental, investigational, or cosmetic for purposes of determining appropriateness of payment. The conclusion that a particular service or supply is medically necessary does not constitute a representation or warranty that this service or supply is covered (i.e., will be paid for by Molina) for a particular member. The member's benefit plan determines coverage. Each benefit plan defines which services are covered, which are excluded, and which are subject to dollar caps or other limits. Members and their providers will need to consult the member's benefit plan to determine if there are any exclusion(s) or other benefit limitations applicable to this service or supply. If there is a discrepancy between this policy and a member's plan of benefits, the benefits plan will govern. In addition, coverage may be mandated by applicable legal requirements of a State, the Federal government or CMS for Medicare and Medicaid members. CMS's Coverage Database can be found on the CMS website. The coverage directive(s) and criteria from an existing National Coverage Determination (NCD) or Local Coverage Determination (LCD) will supersede the contents of this Molina Clinical Policy (MCP) document and provide the directive for all Medicare members.¹

DESCRIPTION OF PROCEDURE/SERVICE/PHARMACEUTICAL

Noninvasive positive pressure ventilation (NIPPV) is the use of assisted ventilation without an artificial airway (eg, tracheostomy tube or endotracheal tube) that is applied as positive pressure to the airway opening. The use of NIPPV has increased in the acute care hospital setting, the long-term care hospital and the home for the management of patients with chronic respiratory failure requiring prolonged mechanical ventilation. Home NIPPV has been investigated for use in patients with obstructive lung disease, neuromuscular disease, obesity hypoventilation syndrome (OHS), and restrictive chest wall disease.

The FDA has approved several portable home ventilators (e.g. Trilogy™, Newport®, Vela®, IVent, Puritan™, and LTV®) to provide continuous or intermittent positive pressure ventilation through invasive or non-invasive interface for use in those individuals who require mechanical ventilation. On February 11, 2014 Phillips Respironics initiated a voluntary recall to address a potentially defective component on Trilogy Ventilators Models 100, 200, and 202.⁴²⁻⁴³

RECOMMENDATION ³⁻⁹⁻¹⁰⁻³⁸⁻⁴⁰

This policy does not address the use of other respiratory assist devices including BiPAP or CPAP and NIPPV devices with bilevel positive airway pressure (bilevel PAP, BIPAP) or a bilevel PAP device with a backup rate. Please see McKesson InterQual or Milliman MCG for criteria. These devices are considered first line treatment and should have been tried and failed before following the criteria outlined below.

Home use of a non-invasive positive pressure ventilator (NIPPV) used with noninvasive interface, (e.g., mask, chest shell) that includes all of the following: [Trilogy™, Newport®, Vela®, IVent, Puritan™, and LTV®] for the treatment of chronic respiratory failure may be considered medically necessary when ALL of the following criteria are met: [ALL]

- Diagnosis of chronic respiratory failure due to progressive neuromuscular disease (e.g., muscular dystrophies, poliomyelitis, multiple sclerosis, spinal cord injuries, spinal muscular atrophy, diaphragmatic paralysis, myasthenia gravis, amyotrophic lateral sclerosis); and
- Mechanical ventilation required due to respiratory insufficiency with ANY ONE of the following: [ONE]
 - Arterial O₂ saturation < 88% for 5 consecutive minutes during nocturnal oximetry; or
 - Arterial PCO₂ ≥ 45 mm Hg (6.0 kPa), or
 - Maximal inspiratory pressures < 60 cm/H₂O or
 - Forced Vital Capacity (FVC) < 50% predicted

OR

On a case by case basis with medical director level of review the following conditions may be considered medically necessary:

- Diagnosis of chronic obstructive pulmonary disease (COPD) and all of the following:
 - Chronic hypercapnia with PaCO₂ of 50 mm Hg (6.7 kPa) to < 52 mm Hg (6.9 kPa) and one or more of the following: ⁴⁰ [ONE]
 - Arterial oxygen saturation less than or equal to 88% for 5 consecutive minutes during nocturnal oximetry while on at least 2 liters of oxygen per minute; or
 - Invasive or noninvasive ventilation for acute exacerbation required during 2 or more hospitalizations per year; or
 - Palliative care in patient with end-stage disease and advance directive stating no desire for intubation; and
- Diagnosis of obesity hypoventilation syndrome and all of the following: ⁴⁰ [ALL]
 - BMI greater than 30
 - Daytime hypercapnia with PaCO₂ greater than 45 mm Hg (6.0 kPa) without other etiology (eg, kyphoscoliosis, lung parenchymal disease, myopathy, severe hypothyroidism)
 - Sleep-disordered breathing or hypoventilation on polysomnography, as indicated by 1 or more of the following:
 - Apnea-hypopnea index of 5 or greater
 - Increase in PaCO₂ during sleep by more than 10 mm Hg (1.3 kPa) above value while awake
 - Significant oxygen desaturation (eg, less than 90%) not explained by obstructive apneas or hypopneas
 - TSH level does not demonstrate hypothyroidism; and
- Failure to improve arterial oxygen saturations and/or hypercapnia on CPAP or BiPAP devices; and
- Diagnosis and prescription for the device must be made by a physician specialist in the disease or a pulmonologist; and
- The NIPPV device is FDA approved for the clinical indications; and
- None of the following conditions are present:
 - alteration in level of consciousness (i.e. alert and oriented)
 - anatomic abnormality that precludes mask fitting (facial or neurological surgery, trauma, or deformity)
 - high risk for aspiration (excessive secretions, impaired cough or inability of mechanically assisted cough to clear secretions)
 - swallowing disorder

- inability to cooperate/protect airway
- upper airway obstruction

COVERAGE EXCLUSIONS

Home non-invasive positive pressure ventilators are excluded and considered not medically necessary for ALL of the following conditions:

- ❑ For the treatment of obstructive sleep apnea as the clinical outcomes have not been shown to be superior to other standard treatments (e.g., CPAP, BiPAP); and
- ❑ For the treatment of other conditions/diseases including but not limited to cystic fibrosis due to insufficient evidence in the peer reviewed medical publications

SUMMARY OF MEDICAL EVIDENCE¹⁰⁻³⁸

The peer reviewed published literature is sufficient to confirm improved outcomes of NIPPV in patients with progressive neuromuscular disease (e.g., muscular dystrophies, poliomyelitis, multiple sclerosis, spinal cord diseases, diaphragmatic paralysis, myasthenia gravis, amyotrophic lateral sclerosis). There are meta-analysis, systematic reviews and randomised controlled trials that have determined positive clinical effects of non-invasive ventilation and validate clinical net health outcomes of home NIPPV.

The peer reviewed published medical literature is sufficient and demonstrates at least a moderate net benefit of NIPPV in patients with chronic COPD. A multicenter randomized controlled trial of 195 patients with stable chronic obstructive pulmonary disease who were classified as having hypercapnic Global Initiative for Chronic Obstructive Lung Disease stage IV disease found, at 12-month follow-up, that the use of noninvasive positive pressure ventilation was associated with lower mortality as compared with usual care (12% and 33%, respectively).²⁷ A randomized controlled trial of 116 chronic obstructive pulmonary disease assigned patients with persistent hypercapnia and hypoxemia to home oxygen with or without noninvasive positive pressure ventilation for a 12-month period. Patients using noninvasive ventilation experienced longer time to readmission or death (ie, median 4.3 months vs 1.4 months in the oxygen alone arm).³⁰ A randomized trial of nocturnal NIV (known as the Assisted Ventilation in Chronic Airflow Limitation study or AVCAL) was performed in 144 patients with severe COPD and moderate hypercapnia (PaCO₂ >46 mmHg). The effects on survival, lung function, and quality of life of NIV plus long-term oxygen therapy (LTOT) were compared with LTOT alone. NIV plus LTOT improved sleep quality and initial nocturnal hypercapnia, with fair compliance to NIV therapy (mean nightly use [standard deviation (SD)], 4.5 [3.2] hours/night). After a mean follow-up of 2.2 years, the NIV group had an improved survival out to 36 months, but thereafter the survival curves converged. The forced expiratory volume in one second (FEV₁) and PaCO₂ at 6 and 12 months were not different between the groups. Despite improved initial survival and sleep quality, quality of life was actually lower with NIV.²⁸

The peer reviewed published medical literature is insufficient to confirm improved outcomes of NIPPV in patients with cystic fibrosis and obstructive sleep apnea. Further investigation is needed with larger populations to determine appropriate candidates for NIPPV and validate long-term, predictable outcomes. Studies thus far have yielded mixed results due to methodological issues and problems with compliance. There is a need for long-term randomised controlled trials which are adequately designed to determine the clinical effects of non-invasive ventilation and explore potential interventions that can improve compliance, decrease adverse events associated with non-compliance, and validate clinical net health outcomes of home NIPPV.

Professional Society Guidelines:⁴⁻⁹

Global Initiative for Chronic Obstructive Lung Disease (GOLD) Guidelines:⁶

- The 2016 Guideline entitled “Global Strategy for the Diagnosis, Management and Prevention of COPD, Global Initiative for Chronic Obstructive Lung Disease (GOLD) indicates that Non-invasive ventilation (NIV) is increasingly used in patients with stable very severe COPD. Randomized controlled trials provide contradictory results regarding the clinical benefits of long-term NIV in patients with COPD and chronic hypercapnia, especially in terms of health status and survival. Thus, there is insufficient evidence to formulate recommendations. The combination of NIV with long-term oxygen therapy may be of some use in a selected subset of patients, particularly in those with pronounced daytime hypercapnia. It may improve survival but does not improve quality of life. However, in patients with both COPD and obstructive sleep apnea there are clear benefits from continuous positive airway pressure (CPAP) in both survival and risk of hospital admission.
- The 2017 GOLD Guidelines indicate that NIV may improve hospitalization-free survival in select patients after recent hospitalization, particularly in those with pronounced daytime hypercapnia (PaCO₂ ≥ 52mmHg). Some trials show improvement and others show no improvement and conclude that there is conflicting evidence whether NIPPV improves survival in COPD.
- The 2018 GOLD Guidelines state that whether to use NPPV chronically at home to treat COPD patients with acute on chronic respiratory failure following hospitalization remains undetermined and the outcome may be affected by persistent hypercapnia. ⁶

National Institute for Health and Clinical Excellence (NICE) Quality statement on Non-invasive ventilation (2016) indicates that noninvasive ventilation can be considered for the treatment of patients with chronic obstructive pulmonary disease and chronic hypercapnia who have required invasive or noninvasive ventilatory assistance during an exacerbation or who are hypercapnic or acidotic while on long-term oxygen therapy. ⁷

CODING INFORMATION: THE CODES LISTED IN THIS POLICY ARE FOR REFERENCE PURPOSES ONLY. LISTING OF A SERVICE OR DEVICE CODE IN THIS POLICY DOES NOT IMPLY THAT THE SERVICE DESCRIBED BY THIS CODE IS COVERED OR NON-COVERED. COVERAGE IS DETERMINED BY THE BENEFIT DOCUMENT. THIS LIST OF CODES MAY NOT BE ALL INCLUSIVE.

CPT	Description
	N/A

HCPCS	Description
E0466	Home ventilator, any type, used with noninvasive interface, (e.g., mask, chest shell)

ICD-10	Description: [For dates of service on or after 10/01/2015]
J44-J44.9	Chronic obstructive pulmonary disease
J96.10 – J96.12	Chronic respiratory failure
E66.2	Morbid (severe) obesity with alveolar hypoventilation

REFERENCES

Government Agency

1. Centers for Medicare & Medicaid Services (CMS). Medicare Coverage Database. National coverage determination (NCD) Search. Accessed at: <http://www.cms.gov/medicare-coverage-database/>
2. CMS Decision Memo for Noninvasive Positive Pressure RADs for COPD (CAG-00052N). June 29, 2001. Accessed at: <https://www.cms.gov/medicare-coverage-database/details/nca-decision-memo.aspx?NCAId=56&ver=5&viewAMA=Y&bc=AAAAAAAAEAAA&>
3. Williams JW, Jr., Cox CE, Hargett CW, et al. Noninvasive positive-pressure ventilation (NPPV) for acute respiratory failure. Comparative Effectiveness Review 68. Prepared by the Duke Evidence-based Practice Center

Professional Society Guidelines

4. American Thoracic Society; European Respiratory Society (ERS/ATS):
 - Official ERS/ATS clinical practice guidelines: noninvasive ventilation for acute respiratory failure. (2017). Accessed at: <https://www.thoracic.org/statements/resources/cc/niv-guidelines.pdf>
 - Management of COPD exacerbations: a European Respiratory Society/American Thoracic Society guideline. (2017) Accessed at: <http://www.thoracic.org/statements/copd.php>
5. British Thoracic Society. (2012). Guidelines for respiratory management of children with neuromuscular weakness. Accessed at: http://www.brit-thoracic.org.uk/Portals/0/Guidelines/Resp%20Mgt%20Children/thoraxjnl_67_Suppl_1_completeissue.pdf.
6. Global Strategy for the Diagnosis, Management and Prevention of COPD: Accessed at: <https://goldcopd.org/>.
 - Global Initiative for Chronic Obstructive Lung Disease (GOLD) 2016.
 - Global Initiative for Chronic Obstructive Lung Disease (GOLD) 2017.
 - Global Initiative for Chronic Obstructive Lung Disease (GOLD) 2018.
7. National Institute for Health and Clinical Excellence. Quality statement 7: Non-invasive ventilation. (2016). Accessed at: <https://www.nice.org.uk/guidance/qs10/chapter/quality-statement-7-noninvasive-ventilation>
8. National Institute for Health and Clinical Excellence. Nice Guideline. Accessed at: <https://www.nice.org.uk/guidance?action=find>
 - Management of chronic obstructive pulmonary disease in adults in primary and secondary care. (2010) CG101.
 - Motor neuron diseases (2010): The use of noninvasive ventilation in the treatment of motor neuron diseases.
9. Ontario Health Technology Assessment Series. (2012). Chronic obstructive pulmonary disease evidentiary framework. Accessed at: http://www.hqontario.ca/en/mas/tech/pdfs/2012/rev_COPD_Framework_March.pdf.

Peer Reviewed Publications

10. Annane D, Orlikowski D, Chevret S, Chevrolet JC, Raphael JC. Nocturnal mechanical ventilation for chronic hypoventilation in patients with neuromuscular and chest wall disorders. *Cochrane Database Syst Rev*. 2007(4):Cd001941.
11. Carron M, Freo U, BaHammam AS, et al. Complications of non-invasive ventilation techniques: a comprehensive qualitative review of randomized trials. *Br J Anaesth*. 2013 Jun;110(6):896 – 914.
12. Ekkernkamp E, Storre JH, Windisch W, Dreher M. Impact of intelligent volume-assured pressure support on sleep quality in stable hypercapnic chronic obstructive pulmonary disease patients: a randomized, crossover study. *Respiration*. 2014;88(4):270-6. doi: 10.1159/000364946. Epub 2014 Aug 22.
13. Gupta D, Nath A, Agarwal R, Behera D. A prospective randomized controlled trial on the efficacy of noninvasive ventilation in severe acute asthma. *Respir Care*. 2010;55(5):536-543.
14. Hess DR. The Growing Role of Noninvasive Ventilation in Patients Requiring Prolonged Mechanical Ventilation. *Respiratory Care* June 1, 2012 vol. 57 no. 6 900-920. Accessed at: <http://rc.rcjournal.com/content/57/6/900.full>
15. Keenan SP, Mehta S. Noninvasive ventilation for patients presenting with acute respiratory failure: The randomized controlled trials. *Respir Care*. 2009;54(1):116-126.
16. Meneses J, Bhandari V, Alves JG, Herrmann D. Noninvasive ventilation for respiratory distress syndrome: A randomized controlled trial. *Pediatrics*. 2011;127(2):300-307.
17. Moran F1, Bradley JM, Piper AJ. Non-invasive ventilation for cystic fibrosis. *Cochrane Database Syst Rev*. 2013 Apr 30;4:CD002769. doi: 10.1002/14651858.CD002769.pub4.
18. Radunovic A, Annane D, Rafiq MK, Mustfa N. Mechanical ventilation for amyotrophic lateral sclerosis/motor neuron disease. *Cochrane Database Syst Rev*. 2013;3:CD004427.

19. Ram FSF, Wellington SR, Rowe B, Wedzicha JA. Non-invasive positive pressure ventilation for treatment of respiratory failure due to severe acute exacerbations of asthma. *Cochrane Database Syst Rev*. 2005;(3):CD004360.
20. Walkey AJ, Wiener RS. Use of noninvasive ventilation in patients with acute respiratory failure, 2000-2009: A population-based study. *Ann Am Thorac Soc*. 2013;10(1):10-17.
21. Ward S, Chatwin M, Heather S, Simonds AK. Randomised controlled trial of non-invasive ventilation (NIV) for nocturnal hypoventilation in neuromuscular and chest wall disease patients with daytime normocapnia. *Thorax*. 2005 Dec;60(12):1019-24.

COPD Publications

22. Bhatt SP, Peterson MW, Wilson JS, Durairaj L. Noninvasive positive pressure ventilation in subjects with stable COPD: a randomized trial. *Int J Chron Obstruct Pulmon Dis*. 2013;8:581-9. doi: 10.2147/COPD.S53619. Epub 2013 Nov 22.
23. Cochrane Database of Systematic Reviews: Osadnik CR, Tee VS, Carson-Chahhoud KV, Picot J, Wedzicha JA, Smith BJ. Non-invasive ventilation for the management of acute hypercapnic respiratory failure due to exacerbation of chronic obstructive pulmonary disease. *Cochrane Database of Systematic Reviews* 2017, Issue 7. Art. No.: CD004104. DOI: 10.1002/14651858.CD004104.pub4.
24. Dreher M, Storre JH, Schmoor C, Windisch W. High-intensity versus low-intensity non-invasive ventilation in patients with stable hypercapnic COPD: a randomised crossover trial. *Thorax* 2010; 65:303.
25. Duiverman ML, Wempe JB, Bladder G et al. Two-year home-based nocturnal noninvasive ventilation added to rehabilitation in chronic obstructive pulmonary disease patients: a randomized controlled trial. *Respir Res*. 2011 Aug 23;12:112. doi: 10.1186/1465-9921-12-112.
26. Hajian B1, De Backer J2, Sneyers C3, Ferreira F2, Barboza KC2, Leemans G1, Vos W2, De Backer W1. Pathophysiological mechanism of long-term noninvasive ventilation in stable hypercapnic patients with COPD using functional respiratory imaging. *Int J Chron Obstruct Pulmon Dis*. 2017 Jul 28;12:2197-2205.
27. Köhnlein T, Windisch W, Köhler D, et al. Non-invasive positive pressure ventilation for the treatment of severe stable chronic obstructive pulmonary disease: a prospective, multicentre, randomised, controlled clinical trial. *Lancet Respir Med* 2014; 2:698.
28. McEvoy RD, Pierce RJ, Hillman D, et al. Nocturnal non-invasive nasal ventilation in stable hypercapnic COPD: a randomised controlled trial. *Thorax* 2009; 64:561.
29. Murphy PB, Brignall K, Moxham J, et al. High pressure versus high intensity noninvasive ventilation in stable hypercapnic chronic obstructive pulmonary disease: a randomized crossover trial. *Int J Chron Obstruct Pulmon Dis* 2012; 7:811.
30. Murphy PB, Rehal S, Arbane G, et al. Effect of Home Noninvasive Ventilation With Oxygen Therapy vs Oxygen Therapy Alone on Hospital Readmission or Death After an Acute COPD Exacerbation: A Randomized Clinical Trial. *JAMA*. 2017;317(21):2177.
31. Ram FSF, Picot J, Lightowler J, Wedzicha JA. Non-invasive positive pressure ventilation for treatment of respiratory failure due to exacerbations of chronic obstructive pulmonary disease. *Cochrane Database Syst Rev*. 2004;(3):CD004014.
32. Shi JX, Xu J, Sun WK, et al. Effect of noninvasive, positive pressure ventilation on patients with severe, stable chronic obstructive pulmonary disease: A meta-analysis. *Chin Med J (Engl)*. 2013;126(1):140-146.
33. Struik FM, Lacasse Y, Goldstein RS, et al. Nocturnal noninvasive positive pressure ventilation in stable COPD: A systematic review and individual patient data meta-analysis. *Respir Med*. 2014;108(2):329-337.
34. Struik, F., Lacasse, Y., Goldstein, R., Kerstjens, H., & Wijkstra, P. (2013). Nocturnal non-invasive positive pressure ventilation for stable chronic obstructive pulmonary disease. *Cochrane Database of Systematic Rev*. 2013, Issue 6. Art. No.: CD002878. DOI: 10.1002/14651858.CD002878.pub2.
35. Struik FM, Sprooten RT, Kerstjens HA et al. Nocturnal non-invasive ventilation in COPD patients with prolonged hypercapnia after ventilatory support for acute respiratory failure: a randomised, controlled, parallel-group study. *Thorax*. 2014 Sep;69(9):826-34. doi: 10.1136/thoraxjnl-2014-205126. Epub 2014 Apr 29.
36. Tsai CL, Lee WY, Delclos GL, et al. Comparative effectiveness of noninvasive ventilation vs invasive mechanical ventilation in chronic obstructive pulmonary disease patients with acute respiratory failure. *J Hosp Med*. 2013;8(4):165-172.

37. Waseem S.M.A., Hossain M.M., Ahmad Z. Obesity, smoking and inflammation: Trilogy and vicious: COPD we are coming - A pilot study. *Biomedicine (India)*. 33 (2) (pp 270-273), 2013.
38. Zhou L, Li X, Guan L, et al. Home noninvasive positive pressure ventilation with built-in software in stable hypercapnic COPD: a short-term prospective, multicenter, randomized, controlled trial. *Int J Chron Obstruct Pulmon Dis*. 2017 Apr 27;12:1279-1286. doi: 10.2147/COPD.S127540.

Other Resources

39. Hayes Search & Summary. Noninvasive Positive Pressure Ventilation Using the Trilogy 100 (Respironics Inc.) for Intermittent Home Use in Adults with Severe Chronic Obstructive Pulmonary Disease. Winifred Hayes, Inc. Lansdale, PA. July, 2017.
40. Milliman Care Guidelines, MCG: Ambulatory Care > Durable Medical Equipment, Prosthetics, Orthotics, and Supplies (DMEPOS) >Home Ventilator (Invasive or Noninvasive Interface) (A-0893). 2018
41. UpToDate: [website]. Waltham, MA: Walters Kluwer Health; 2018.
 - Hill NS, Kramer NR. Practical aspects of nocturnal noninvasive ventilation in neuromuscular and chest wall disease. 2018
 - Hill NS, Kramer NR Types of noninvasive nocturnal ventilatory support in neuromuscular and chest wall disease. 2018
 - Gay P. Nocturnal ventilatory support in COPD. 2018.
 - Martin T. Noninvasive positive airway pressure therapy of the obesity hypoventilation syndrome. 2018.
 - Martin T. Treatment and prognosis of the obesity hypoventilation syndrome. 2018
42. U. S. Food and Drug Administration. (2014, February).Phillips Respironics initiates recall of Trilogy ventilator. Accessed at: <http://www.fda.gov/Safety/Recalls?ucm386490.htm>.
43. U. S. Food and Drug Administration. Center for Devices and Radiological Health. 510(k) Premarket Notification Database. Enter the 510(k) information for the device. K083526, K082966, K093416, K111146, K111610. Accessed at: <http://www.accessdata.fda.gov/scripts/cdrh/cfdocs/cfPMN/pmn.cfm>.

Peer Review: Policy reviewed by AMR practicing physician board certified in Internal Medicine, Pulmonary Disease. 10/10/17 and 5/21/18

Review/Revision History:

12/17: The clinical criteria have not changed. References and professional society guidelines were updated.
6/18: Policy was reviewed and the clinical criteria has changed. Added medically necessary criteria for the diagnosis of COPD and chronic restrictive hypoventilation syndrome related to obesity. Updated professional society guidelines and references.