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FEP Medical Policy Manual

FEP 8.01.64 Home Non-Invasive Positive Airway Pressure Devices for the Treatment of Respiratory Insufficiency and Failure

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Related Policies:

2.01.18 - Diagnosis of Obstructive Sleep Apnea Syndrome

Home Non-Invasive Positive Airway Pressure Devices for the Treatment of Respiratory Insufficiency and Failure

Description

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Respiratory failure is characterized by low arterial blood oxygen (hypoxemia, PaO_2) and/or high arterial carbon dioxide (hypercapnia, $PaCO_2$ > 45 mmHg). Chronic respiratory insufficiency or failure can occur with chronic obstructive pulmonary disease (COPD), thoracic restrictive disorders, and hypoventilation syndromes, and may result in poor quality of life, sleepiness, hospital admission, intubation, and death. Non-invasive positive airway pressure ventilation (NPPV) including continuous positive airway pressure (CPAP), bilevel positive airway pressure (BPAP), and home mechanical ventilators (HMV) that are pressure, rate and volume targeted are proposed for the treatment of COPD and other forms of chronic hypoventilation.

Chronic Obstructive Pulmonary Disease

Chronic obstructive pulmonary disease (COPD) is a common condition, affecting more than 5% of the population, and is associated with high morbidity and mortality. COPD is the fourth leading cause of death in the United States. It is a clinical syndrome with multiple etiologies that is characterized by chronic respiratory symptoms, structural pulmonary abnormalities, and/or lung function impairment. Chronic obstructive pulmonary disease is most frequently associated with cigarette smoking or other air pollutants, and a majority of patients with COPD in the United States have a history of cigarette smoking. Chronic obstructive pulmonary disease is progressive, with expiratory airflow limitation, air trapping/hyperinflation, and destruction of alveoli (emphysema). The Global Initiative for Chronic Obstructive Lung Disease (GOLD), defines COPD as "a heterogeneous lung condition characterized by chronic respiratory symptoms (dyspnea, cough, sputum production and/or exacerbations) due to abnormalities of the airways (bronchitis, bronchiolitis) and/or alveoli (emphysema) that cause persistent, often progressive, airflow obstruction".[1,2,]

Respiratory failure in patients with COPD is characterized by the inability to sustain normal gas exchange, leading to low arterial blood oxygen (hypoxemia, PaO_2) and/or high arterial carbon dioxide (hypercapnia, $PaCO_2$). Hypercapnia develops in about one-third of patients with COPD and is associated with poor quality of life, sleepiness, frequent hospital admissions due to exacerbations, and an increase in mortality compared to patients with COPD who are normocapnic. The hypercapnia is due in large part to poor lung biomechanics including low inspiratory muscle reserve, high CO_2 production, and a reduced ventilatory capability.[3,] The imbalance between the respiratory load and respiratory capability may in turn affect the ventilatory control center in the brain stem. Physiological changes in responsiveness to hypoxemia and hypercapnia during sleep can be particularly pronounced in patients with COPD, with overnight increases in $PaCO_2$ affecting daytime $PaCO_2$, possibly through bicarbonate retention or changes in cerebrospinal fluid.[4,] Patients with COPD may also have comorbid obstructive sleep apnea and/or obesity hypoventilation syndrome due to decreased ventilatory motor output and upper airway muscle activity during sleep.

Thoracic Restrictive Disorders Due to Neuromuscular Disease

Thoracic restrictive disorders result from a variety of underlying diseases all characterized by restrictive patterns on pulmonary function testing.[5,] Neuromuscular disorders such as muscular dystrophy, amyotrophic lateral sclerosis (ALS), polio, and phrenic neuropathies can result in weakness of the respiratory muscles affecting inspiration and expiration, ultimately resulting in hypoventilation. Impaired cough and swallowing associated with neuromuscular disease increases the risk of respiratory complications in these patients.[6,]Nocturnal hypoventilation due to muscular atonia during sleep leads to nocturnal hypercapnia. Frequent nocturnal episodes can result in renal compensation and ultimately result in daytime hypercapnia. Non-invasive positive airway pressure ventilation (NPPV) is often necessary for patients with thoracic restrictive disorders due to neuromuscular disease.

Hypoventilation Syndromes

Hypoventilation syndromes are nonspecific disorders characterized by hypercapnia ($PaCO_2$ >45 mm Hg) that is not otherwise categorized.[7,] Obesity hypoventilation syndrome (OHS), central respiratory depression due to substance or medication use, and decompensated hypercapnic respiratory failure that is not COPD are all included in this category. In patients with OHS, weight loss is useful in normalizing PaCO2; however, NPPV should be initiated early while weight loss is attempted.[8,]

Treatment With Non-invasive Positive Airway Pressure

A major goal of management of patients with chronic hypoventilation is to reduce hospitalizations and mortality. Long-term oxygen therapy is recommended for patients with poor clinical status and NPPV devices for patients with severe chronic hypercapnia and a history of hospitalization for acute respiratory failure. Non-invasive positive airway pressure ventilation devices include nocturnal continuous positive airway pressure (CPAP) for individuals with hypercapnia due to obstructive sleep apnea or hypoventilation and bilevel positive airway pressure (BPAP) devices or non-invasive home mechanical ventilators that are pressure, rate, and volume targeted. The objective of this evidence review is to describe which features of NPPV are required to improve the net health outcome in patients with COPD, thoracic restrictive disorders due to neuromuscular disease, or those with hypoventilation syndromes.

Benefits of nocturnal NPPV persist into the daytime with improved breathing patterns (lower frequencies and larger tidal volumes) and improved gas exchange. Explanations for the improvement in daytime respiration with nocturnal NPPV include increased respiratory drive, improved diaphragm function by unloading the respiratory muscles during sleep, increased CO_2 sensitivity, and reduction in air trapping and hyperinflation. It is not known which factors (eg, muscle unloading, gas exchange normalization, decrease in hyperinflation) underlie the benefits of NPPV on health outcomes. It is also unclear if the reduction in $PaCO_2$ has an effect on health outcomes or if it is only a marker of effective ventilation.[4,]

Respiratory Assist Devices

The Centers for Medicare and Medicaid Services (CMS) defines respiratory assist devices (RADs) as bilevel devices with or without back-up respiratory rate capability. While CPAP devices provide continuous air at a pressure that prevents the collapse of the airway during inspiration, BPAP devices work by increasing pressure during inspiration and lowering it during expiration (pressure cycled). In some devices a backup respiratory rate is triggered when the patient's nocturnal respiratory rate decreases below a set threshold. The backup rate is typically set 2 breaths below the patient's spontaneous respiratory rate during wakefulness.

Terminology on device features is described in Table 1.

Table 1. Device Features

Term	Definition	Description
Bilevel-S	Bilevel without a backup rate	Positive airway pressure that is higher during inspiration than expiration that is triggered by patient inspiration.
Bilevel-ST	Bilevel with a backup rate	Positive airway pressure that is higher during inspiration than expiration with a backup respiratory cycle length if the patient's breathing is slower than the preset rate.
VAPS	Volume-assured pressure support modes	Bilevel ST modes that use an algorithm to adjust inspiratory pressure support to meet a set tidal volume.
iVAPS	Intelligent volume-assured pressure support modes	Bilevel ST modes that use an algorithm to adjust inspiratory pressure support within a predetermined range to meet a set target ventilation.

Home Mechanical Ventilators

In some patients, nocturnal respiratory assist devices are insufficient to address the respiratory failure. Non-invasive home mechanical ventilators (HMV) are proposed for the treatment of chronic respiratory failure that is refractory to a respiratory assist device. Mechanical ventilators are devices that deliver more controlled breathing with bilevel ventilation at a higher pressure. The ventilators may also have additional features compared to BPAP machines such as alarms and battery backup power. Home mechanical ventilators can be used for patients with tracheostomy in the home, but may also be used with a non-invasive interface such as a mask or mouthpiece in patients who do not depend on 24 hour ventilation for survival. Current technology has decreased the size of home ventilators to around 10 pounds. In addition, some models may be wireless with battery backup, allowing greater mobility during the day.

Titration

Early studies with low intensity NPPV did not demonstrate health benefits in patients with hypercapnia. More recent studies have reinforced the importance of high-intensity NPPV (>18 cm H_2O) that is titrated to decrease hypercapnia. A high respiratory backup rate that is increased to the level of spontaneous breathing has also been shown to be important to achieve positive health outcomes. Manually set, laboratory or hospital titration of NPPV with pressure control and backup rate have been recommended for stable hypercapnic COPD.[9,] The goal of titration of inspiratory positive airway pressure is to achieve normocapnia, a reduction in transcutaneous CO_2, or maximum tolerable inspiratory pressure. A fast rise in inspiratory pressure (rise time) allows enough time for expiration within the normal rate of breathing. In patients with air trapping and hyperinflation, use of positive end-expiratory pressure can also be beneficial.

A suggested protocol for in-laboratory titration of NPPV in patients with COPD in the U.S. is described by Orr et al (2020).[4,] Titration of NPPV is usually performed in a monitored environment after the patient has stabilized, as studies have not found an improvement in health outcomes when NPPV is started soon after an acute exacerbation. Polysomnography or respiratory monitoring may be used during titration to evaluate the presence of obstructive sleep apnea or hypoventilation. The inspiratory pressure is typically started at 6 to 8 cm H_2O of pressure support above the expiratory pressure and titrated to reduce hypercapnia. A Bilevel-ST (with backup rate) or a VAPS (volume assured) may be used if a Bilevel-S (without backup rate) fails to adequately reduce hypercapnia. Although titration in European studies has been performed with a hospital stay, this is not feasible in the U.S., and titration might be performed over several weeks in the patient's home by an external durable medical equipment (DME) provider.

Pulmonary Rehabilitation

Pulmonary rehabilitation is a personalized intervention that includes physical activity (eg, activities of daily living, endurance exercises and muscle strengthening), health education, and psychological support. It may be performed in the hospital, outpatient clinic, or home, and has been shown to reduce mortality, exacerbation rate, intensive care admissions, and emergency department visits. Pulmonary rehabilitation is common in Europe but is less frequently provided in the U.S.

OBJECTIVE

The objective of this evidence review is to describe which features of non-invasive positive pressure ventilation are required to improve the net health outcome in individuals with chronic obstructive pulmonary disease, thoracic restrictive disorders due to neuromuscular disease, or hypoventilation syndromes.

POLICY STATEMENT

For patients with chronic obstructive pulmonary disease (COPD) without significant hypercapnia ($PaCO_2$ < 52 mmHg) and with obstructive sleep apnea, see policy 2.01.18 (Diagnosis of Obstructive Sleep Apnea Syndrome) and 8.01.64 (Medical Management of Obstructive Sleep Apnea Syndrome).

Nocturnal bilevel positive airway pressure (BPAP) with backup rate may be considered **medically necessary** for individuals with COPD and chronic respiratory failure (see Policy Guidelines) who meet either of the following:

- Chronic stable daytime (awake) hypercapnia ($PaCO_2$>52 mmHg) **OR**
- Daytime (awake) hypercapnia ($PaCO_2$>52 mmHg) at least 2 weeks after discharge from the hospital for an acute exacerbation with decompensated respiratory acidosis.

Non-invasive home mechanical ventilation may be considered **medically necessary** for individuals with COPD who meet the following criteria:

- Qualify for a BPAP device **AND** meet at least one of the following:
    - Higher pressure is needed to reduce hypercapnia than can be achieved with a bilevel device during titration (typically >25cm H_2O); OR
    - Severe hypoxemia requiring fraction of inspired oxygen (FIO_2) >40% or >5 L/min; **OR**
    - Daytime use (battery operated unit) is required to reduce hypercapnia.

Individuals with COPD who are started on BPAP at discharge from hospitalization may continue for up to 3 months to provide time to stabilize and complete reevaluation.

Non-invasive positive airway pressure for COPD is considered **investigational** under all other conditions.

Bilevel positive airway pressure may be considered **medically necessary** for individuals with thoracic restrictive disorders due to neuromuscular disease who meet any of the following:

- Pulmonary function tests:
    - Spirometry (upright or supine) with vital capacity <50% predicted or <80% predicted with associated symptoms (orthopnea, dyspnea, morning headaches, excessive daytime sleepiness, or unrefreshing sleep); OR
    - Maximal inspiratory pressure <60 cm H_2O or maximum expiratory pressure (MEP) <40 cm H_2O; OR
    - Peak cough flow (PCF) <270 L/min for age ≥12 years or PCF <5th percentile for age <12 years; OR
    - Sniff nasal inspiratory pressure (SNIP) <70 cm H_2O in males, SNIP <60 cmH_2O in females for age ≥12 years.

- Hypercapnia
    - Chronic stable daytime (awake) hypercapnia with $PaCO_2$ ≥45 mmHg (capillary blood gas can be used in children); OR
    - Venous blood gas PCO_2, end-tidal PCO_2, or transcutaneous PCO_2, ≥50 mmHg; OR
- Hypoxia
    - Overnight oximetry in-laboratory or home sleep test with saturation <88% for 5 minutes;
    - Overnight oximetry: SpO2 ≤ 90% for ≥ 2% of sleep time.

Non-invasive home mechanical ventilation may be considered **medically necessary** for individuals with thoracic restrictive disorders due to neuromuscular disease who meet the following:

- Qualify for a BPAP device; AND
    - BPAP fails; OR
    - Have extreme loss in function with vital capacity <30%; OR
    - Non-invasive ventilation is needed for >10 hours per day; OR
    - Severe breathlessness (e.g., with speaking at rest); OR
    - Worsening daytime hypercapnia with need for mouthpiece ventilation; OR
    - Daytime use (battery operated unit) is required to reduce hypercapnia or dyspnea.

Bilevel positive airway pressure may be considered **medically necessary** for individuals with hypoventilation syndromes who meet the following criteria:

- Awake or sleep hypoventilation with hypercapnia (one of the following is met):
    - Awake hypoventilation with chronic stable daytime (awake) hypercapnia ($PaCO_2$ ≥ 45 mmHg); OR
    - Venous blood gas PCO_2, end-tidal PCO_2, or transcutaneous PCO_2 ≥50 mmHg; OR
    - Sleep hypoventilation with hypercapnia:
        - ≥10 mmHg increase from baseline awake PCO_2 and to a value > 50 mmHg for ≥10 min; OR
        - PCO_2 ≥55 mmHg for ≥10 min; AND
- Low clinical suspicion for COPD or neuromuscular disease; AND
- One of the following conditions are met:
    - Obesity with body mass index (BMI) ≥30 kg/m^2; OR
    - Decreased respiratory drive due to opioid or substance use; OR
    - Advanced lung disease other than COPD (e.g., end-stage or advanced interstitial lung disease); AND
- Individual was discharged from inpatient stay with persistent awake hypoventilation (hypercapnia) on BPAP.
    - A reassessment with a provider within 3 months (30 to 90 days) is required and an attended polysomnogram (PSG) should be performed to assess appropriateness of positive airway pressure modality (home sleep apnea test is acceptable if attended PSG is not obtainable); OR
- Individual is ambulatory and sleep study indicates that BPAP is necessary for sleep-disordered breathing, or individual with severe obstructive sleep apnea is continuous positive airway pressure intolerant or continuous positive airway pressure was proven ineffective.

Non-invasive home mechanical ventilation may be considered **medically necessary** for individuals with hypoventilation syndromes who meet the following:

- Qualify for a BPAP device and at least one of the following:
    - Higher pressure is needed to reduce hypercapnia than can be achieved with a bilevel continuous positive airway pressure device during titration (typically >25 cm H_2O); OR
    - Severe hypoxemia requiring FIO_2 >40% or >5 L/min; OR
    - Daytime use (battery operated unit) is required to reduce hypercapnia; OR
- Tried and failed BPAP device with persistent hypercapnia despite 3 months of adequate adherence to prescribed positive airway pressure therapy with:
    - Awake $PaCO_2$ ≥45 mmHg; OR
    - Awake venous blood gas PCO_2, end-tidal PCO_2, or transcutaneous PCO_2 ≥50 mmHg.

POLICY GUIDELINES

Respiratory failure in individuals with chronic disease is characterized by the inability to sustain normal gas exchange, leading to low arterial blood oxygen (hypoxemia, PaO_2) and/or high arterial carbon dioxide (hypercapnia, $PaCO_2$). Assessment of hypoxemia would lead to supplemental oxygen administration. Stable clinical state is defined as free of exacerbations for at least 4 weeks with pH over 7.35.

Compliance with treatment of at least 4 hours per 24 hours should be documented after the first 3 months of use. There are limited data on which to base compliance assessment. Assessment could be further based on an *average* of at least 4 hours per 24 hours over a consecutive 30-day period or use of 4 hours per 24 hours for at least 65% of the days in a consecutive 30-day period.

The Centers for Medicare and Medicaid Services (CMS) classifies a respiratory assist device as a bilevel positive airway pressure (BPAP) device with or without backup respiratory rate capability. Treatment modalities that are reported with the E0471 code include BiPAP ST, ASV, BiPAP AutoSV, iVAPS, AVAPS. BPAP units with batteries have a battery life that is shorter than home mechanical ventilators and are infrequently used in the U.S.

CMS defines non-invasive mechanical ventilators as life supporting/sustaining devices used in various settings, including home, hospital, and institutional settings. The non-invasive mechanical ventilators should have at least 6 pressure modes and 3 volume modes, and allow for both invasive or non-invasive use. For examples, see the Regulatory Status section.

Although most patients with comorbid COPD and obstructive sleep apnea can be effectively treated with continuous or auto-adjusting positive airway pressure, approximately 10% of patients will need BPAP to tolerate the required pressure. These devices are reviewed in reference medical policy 2.01.18 (Diagnosis of Obstructive Sleep Apnea Syndrome) and 8.01.64 (Medical Management of Obstructive Sleep Apnea Syndrome).

Respiratory therapy in the home may be provided for patients who are treated with E0466, E0470, or E0471 devices.

BENEFIT APPLICATION

Experimental or investigational procedures, treatments, drugs, or devices are not covered (See General Exclusion Section of brochure).

FDA REGULATORY STATUS

Numerous CPAP and BPAP devices are available in the U.S. Examples of HMV devices that have both invasive and non-invasive interfaces and are available in the U.S. are described in Table 2.

Table 2. Select Home Mechanical Ventilators with Non-invasive Interface

Device	Manufacturer	FDA clearance	Date	FDA product code
Trilogy™ Evo Ventilator	Respironics	K181166	2019	NOU, CBK
Vivo 60	Breas	K160481	2016	NOU, CBK, DQA, CCK
Astral 100/150	ResMed	K152068	2016	NOU, CBK
Newport™	Medtronic	K121891	2012	NOU, CBK
iVent	GE Healthcare	K092135	2009	NOU, CBK
LTV	Cardinal Health	K083688	2009	CBK
Puritan Bennett 540	Covidien	K082966	2008	CBK

RATIONALE

Summary of Evidence

For individuals who have chronic obstructive pulmonary disease (COPD) and obstructive sleep apnea (OSA) who receive continuous positive airway pressure (CPAP) , the evidence includes observational studies. Relevant outcomes are mortality, symptoms, morbid events, functional outcomes, quality of life, and hospitalization. Studies of patients with both COPD and OSA who do or do not use CPAP show a mortality benefit in patients with overlap syndrome who are treated with positive airway pressure. The greatest benefits occur in patients with COPD and hypercapnia and in older adults, and individuals with more comorbid conditions and higher complexity ratings. It should be noted that the threshold for what was considered hypercapnia was lower than in other studies on bilevel positive airway pressure (BPAP) that used a threshold of arterial blood carbon dioxide ($PaCO_2$) > 52 mm Hg. Although the literature indicates that patients with COPD should be screened for OSA due to increased mortality in overlap syndrome, no studies were identified to indicate that CPAP would be prescribed in any manner other than would typically be recommended for patients with clinically significant OSA (see Evidence Review 2.01.18). Patients with overlap syndrome can be treated with CPAP and, when CPAP is not tolerated, with BPAP. The evidence is sufficient to determine that the technology results in an improvement in the net health outcome.

For individuals who have COPD and chronic respiratory failure who receive BPAP, the evidence includes randomized controlled trials (RCTs) and systematic reviews of RCTs. Relevant outcomes are mortality, symptoms, morbid events, functional outcomes, quality of life, and hospitalization. The primary limitation of the evidence base is the heterogeneity of patient selection criteria and treatment parameters. The most recent trials indicate that bilevel non-invasive positive airway pressure ventilation (NPPV) improves hypercapnia in both patients with stable hypercapnia and in patients who have stabilized following an acute exacerbation. There is evidence that some health outcomes including function, readmissions, and death are improved; however, the strength of evidence is low. Several factors have been reported to be important to achieve benefit of NPPV. These are severe hypercapnia with $PaCO_2$ > 52 mmHg, use for at least 5 hours per night, and treatment with high intensity pressure. In addition, for patients with hypercapnia following an acute exacerbation, titration should occur at least 2 weeks after hospitalization when hypercapnia has stabilized. Under these conditions, the evidence is sufficient to determine that the technology results in an improvement in the net health outcome.

For individuals who have COPD and chronic respiratory failure when BPAP is inadequate who receive home mechanical ventilation (HMV), the evidence includes observational studies and an analysis of administrative claims data. Relevant outcomes are mortality, symptoms, morbid events, functional outcomes, quality of life, and hospitalization. There is low strength of evidence based on observational studies and claims data that NPPV reduces the number of hospital admissions or number of patients with hospitalization compared to either no device or BPAP. Due to the severity of the condition, high quality prospective controlled trials are unlikely in patients who have failed BPAP. HMV may be appropriate in situations where BPAP is not adequate to obtain needed pressures or when daytime use and battery backup is needed. The evidence is sufficient to determine that the technology results in an improvement in the net health outcome.

For individuals who have hypoventilation as a result of thoracic disorder due to neuromuscular disease who receive BPAP, the evidence includes systematic reviews. Relevant outcomes are mortality, symptoms, morbid events, functional outcomes, quality of life, and hospitalization. Clinical trials included in a systematic review of 10 RCTs (or quasi-RCTs) evaluated the use of nocturnal NPPV (primarily BPAP) in individuals with neuromuscular or chest wall disorders. One-year mortality rates were significantly reduced with NPPV use (risk ratio, 0.62; 95% confidence interval [CI], 0.42 to 0.91).

Patients treated with NPPV also had lower hospital admission rates and greater symptom improvement. Although the studies were limited by heterogeneity, nocturnal NPPV was found to improve outcomes in patients with restrictive thoracic disorders including neuromuscular disease. A systematic review of observational studies in children with neuromuscular diseases found improved mortality with NPPV compared with standard of care. The evidence is sufficient to determine that the technology results in an improvement in the net health outcome.

For individuals who have hypoventilation as a result of thoracic disorder due to neuromuscular disease who receive HMV, the evidence includes a systematic review of observational studies. One observational study comparing BPAP to HMV found no difference in survival between these ventilation methods, although more patients received effective ventilation with HMV. Due to the severity of the condition, high quality prospective controlled trials are unlikely in patients who have failed BPAP. HMV may be appropriate in situations where BPAP is not adequate to obtain needed pressures or when daytime use and battery backup is needed. The evidence is sufficient to determine that the technology results in an improvement in the net health outcome.

For individuals who have obesity hypoventilation syndrome (OHS) and OSA who receive CPAP, the evidence includes RCTs. In the largest RCT, $PaCO_2$ improved with both CPAP and NPPV (mixed BPAP/mechanical ventilation) compared with lifestyle interventions. The were was no significant difference between CPAP and NPPV in $PaCO_2$ in the short-term or in hospitalized days at long-term follow-up. An RCT comparing CPAP and BPAP found similar outcomes with these treatments. The evidence is sufficient to determine that the technology results in an improvement in the net health outcome.

For individuals who have OHS and chronic respiratory failure who receive BPAP, the evidence includes systematic reviews and RCTs. Relevant outcomes are mortality, symptoms, morbid events, functional outcomes, quality of life, and hospitalization. The majority of evidence specific to BPAP in OHS without OSA comes from a single RCT. In patients with OHS without OSA, BPAP resulted in better $PaCO_2$ outcomes than lifestyle modifications in the short-term, but long-term outcomes failed to find significant improvement in hospitalization days between groups. The evidence is sufficient to determine that the technology results in an improvement in the net health outcome.

For individuals who have OHS and chronic respiratory failure when BPAP is inadequate who receive HMV, there are no randomized or nonrandomized studies. Due to the severity of the condition, high quality prospective controlled trials are unlikely in patients who have failed BPAP. HMV may be appropriate in situations where BPAP is not adequate to obtain needed pressures or when daytime use and battery backup is needed. The evidence is sufficient to determine that the technology results in an improvement in the net health outcome.

For individuals with hypoventilation as a result of hypoventilation syndromes unrelated to OHS, the evidence is limited to case reports and case series primarily in patients with congenital central hypoventilation. In some cases NPPV minimized the need for invasive mechanical ventilation. Due to the severity of the condition, high quality prospective controlled trials are unlikely in patients who have hypoventilation due to hypoventilation syndromes. The evidence is sufficient to determine that the technology results in an improvement in the net health outcome.

SUPPLEMENTAL INFORMATION

Practice Guidelines and Position Statements

Guidelines or position statements will be considered for inclusion in 'Supplemental Information' if they were issued by, or jointly by, a US professional society, an international society with US representation, or National Institute for Health and Care Excellence (NICE). Priority will be given to guidelines that are informed by a systematic review, include strength of evidence ratings, and include a description of management of conflict of interest.

American College of Chest Physicians

In 2023, the American College of Chest Physicians (ACCP) published clinical practice guidelines for respiratory management of patients with neuromuscular weakness.[50,] Most evidence is based on observational data from patients with amyotrophic lateral sclerosis. The guidelines recommend non-invasive ventilation (NIV) for patients with neuromuscular disease and chronic respiratory failure for patients who meet the following pulmonary function test criteria:

- Forced vital capacity (FVC) <80% predicted with symptoms or FVC <50% predicted without symptoms;
- Maximum inspiratory pressure (MIP) <60 cm H_2O or maximum expiratory pressure (MEP) <40 cm H_2O;
- Peak cough flow (PCF) <270 L/min for age ≥12 years or PCF <5th percentile for age <12 years;
- Sniff nasal inspiratory pressure (SNIP) <70 cm H_2O in male patients, SNIP <60 cmH_2O in female patients for age ≥12 years.

The panel found no strong evidence to support one method of NIV over another.

American College of Chest Physicians et al

In 2021, the ACCP, the American Association for Respiratory Care, the American Academy of Sleep Medicine, and the American Thoracic Society published a technical expert panel report on optimal NIV for chronic obstructive pulmonary disease (COPD), thoracic restrictive disorders, and hypoventilation syndromes.[51,52,53,]

Chronic Obstructive Pulmonary Disease

For COPD the panel recommends that overnight oxygen saturation should not be part of the criteria for bilevel positive airway pressure (BPAP) and that home mechanical ventilators be considered when patients need any of the following:[51,]

- "Higher inspiratory pressures than those deliverable by E0471,
- FIO_2 [fraction of inspired oxygen] higher than 40% or 5 L/min nasally,
- Ventilator support for 10 h per day or greater (ie, daytime use),
- Both sophisticated alarms and accompanying internal battery (high-dependency patient),
- Mouthpiece ventilation during the day,
- Persistence of hypercapnia with $PaCO_2$ [arterial blood carbon dioxide]> 52 mm Hg despite adequate adherence to BPAP therapy"
- The panel strongly recommended the use of respiratory therapists in the home for initiation and ongoing support for positive pressure ventilation with either BPAP or home ventilators.

Thoracic Restrictive Disorders

For thoracic restrictive disorders, the panel recommends BPAP for patients with any of the following:[52,]

- "Spirometry (upright or supine) with vital capacity <50% predicted or <80% predicted with associated symptoms (i.e., orthopnea, dyspnea, morning headaches, excessive daytime sleepiness, or unrefreshing sleep),
- Force testing with maximal inspiratory pressure <60 cm H_2O,
- Hypercapnia:
    - Chronic stable daytime (awake) hypercapnia with $PaCO_2$ >45 mmHg,
    - Venous blood gas PCO_2, end-tidal PCO_2, or transcutaneous PCO_2, >50 mmHg, or
- Hypoxia:
    - Overnight oximetry in-laboratory or home sleep test with saturation <88% for 5 minutes."

Home mechanical ventilation is recommended in patients with vital capacity <30% or if BPAP fails.

Hypoventilation Syndromes

For patients with hypoventilation syndromes who are obese the recommendations include:[53,]

- BPAP (spontaneous/timed) or volume-assured pressure support (VAPS) for those who are discharged from the hospital, for those with obesity hypoventilation syndrome (OHS) without obstructive sleep apnea, and for those who have failed continuous positive airway pressure (CPAP).

For patients with hypoventilation syndromes due to reduced respiratory drive or advanced lung disease that is not COPD, BPAP (spontaneous/timed) or VAPS is recommended. Patients with hypoventilation syndromes who fail BPAP/VAPS should receive home mechanical ventilation.

American Thoracic Society

Chronic Obstructive Pulmonary Disease

In 2020, the American Thoracic Society published an evidence-based clinical practice guideline on long-term non-invasive ventilation in chronic stable hypercapnic COPD.[2,] The society included the recommendations in Table 3 , all of which were conditional due to moderate to very low certainty in the evidence base.

Table 3. American Thoracic Society Recommendations for COPD

Recommendation	Strength of Recommendation	Level of Certainty
"We suggest the use of nocturnal noninvasive ventilation (NIV) in addition to usual care for patients with chronic stable hypercapnic COPD."	Conditional	Moderate
"We suggest that patients with chronic stable hypercapnic COPD undergo screening for obstructive sleep apnea before initiation of long-term NIV."	Conditional	Very low
"We suggest not initiating long-term NIV during an admission for acute on-chronic hypercapnic respiratory failure, favoring instead reassessment for NIV at 2 - 4 weeks after resolution."	Conditional	Low
"We suggest not using an in-laboratory overnight polysomnogram (PSG) to titrate NIV in patients with chronic stable hypercapnic COPD who are initiating NIV."	Conditional	Very low
"We suggest NIV with targeted normalization of $PaCO_2$ in patients with hypercapnic COPD on long-term NIV."	Conditional	Low

COPD: chronic obstructive pulmonary disease; NIV: non-invasive ventilation; PaCO2: pressure of carbon dioxide; PSG: polysomnogram.
Hypercapnic COPD defined as PaCO2 > 45 mmHg.

Obesity Hypoventilation Syndrome

In 2019, the American Thoracic Society published a clinical practice guideline on OHS.[54,] These guidelines recommend positive airway pressure for patients with OHS. Generally CPAP is recommended over other NIV because the majority (>70%) of patients have concomitant obstructive sleep apnea (OSA). The guidelines do recommend non-invasive positive airway pressure ventilation (NPPV) initiation at discharge for patients hospitalized with respiratory failure suspected of having OHS until they undergo outpatient workup and titration of positive airway pressure therapy. Both recommendations were conditional with very low level of certainty in the evidence.

Global Initiative for Chronic Obstructive Pulmonary Disease

The Global Initiative for Chronic Obstructive Pulmonary Disease (GOLD) published a revised report for 2023.[1,] GOLD guidelines recommend at least 1 of the following as an indication for non-invasive mechanical ventilation :

- Respiratory acidosis ($PaCO_2$ ≥45 mmHg and arterial pH ≤7.35);
- Severe dyspnea with clinical signs suggestive of respiratory muscle fatigue, increased work of breathing, or both;
- Persistent hypoxemia despite supplemental oxygen therapy.

National Institute for Health and Care Excellence Global

In 2019, the United Kingdom's NICE published a guideline for the diagnosis and management of COPD.[55,] NICE recommends that patients with COPD who have chronic hypercapnic respiratory failure despite adequate pharmacologic and oxygen therapy should be referred to a specialist center for consideration of long-term, non-invasive ventilation.

U.S. Preventive Services Task Force Recommendations

Not applicable.

Medicare National Coverage

There is no national coverage determination. In the absence of a national coverage determination, coverage decisions are left to the discretion of local Medicare carriers.

The Centers for Medicare and Medicaid Services requested topic review by the Agency for Healthcare Research and Quality (AHRQ). The technology assessment was published February 2020.[20,]

REFERENCES

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POLICY HISTORY - THIS POLICY WAS APPROVED BY THE FEP® PHARMACY AND MEDICAL POLICY COMMITTEE ACCORDING TO THE HISTORY BELOW:

Date	Action	Description
March 2022	New policy	Policy created with literature review through November 9, 2021. Considered medically necessary under specified conditions.
July 2022	Replace policy	Criteria for second policy statement refined from "after resolution" to "after discharge from the hospital". Clarification added to second Policy Guideline: "Compliance with treatment of at least 4 hours per 24 hours should be documented after the first 3 months of use. There are limited data on which to base compliance assessment. A percentage of days used could be considered, whereby assessment could be based on an average of at least 4 hours per 24 hours over a consecutive 30-day period or use of 4 hours per 24 hours for at least 65% of the days in a consecutive 30-day period." Intent unchanged.
June 2023	Replace policy - correction only	Updated title for related policy 2.01.18.
December 2023	Replace policy	Policy updated with literature review through May 8, 2023; references added. Title changed to remove "Chronic Obstructive Pulmonary Disease." Medically necessary policy statements added for thoracic restrictive disorders due to neuromuscular disease and hypoventilation syndromes for individuals who meet criteria.