



Medical Coverage Policy

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Home Ventilators

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INSTRUCTIONS FOR USE

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Overview

This Coverage Policy addresses in-home ventilators for children and adults (HCPCS E0465-E0467 only). Ventilators are rental only DME and require on-going rental period authorizations.

Coverage Policy

Coverage for Durable Medical Equipment (DME), including in-home ventilators varies across plans. Please refer to the customer's benefit plan document for coverage details.

INVASIVE INTERFACE (HCPCS E0465)

A home ventilator with invasive interface (e.g., tracheostomy tube) is considered medically necessary if an individual has ANY of the following conditions:

- a neuromuscular disease
- a thoracic restrictive disease
- chronic respiratory failure consequent to chronic obstructive pulmonary disease
- congenital central hypoventilation syndrome

- chronic lung disease of infancy (e.g., bronchopulmonary dysplasia)
- obesity hypoventilation syndrome
- restrictive disorder of chest wall

NON-INVASIVE INTERFACE (HCPCS E0466)

A home ventilator with non-invasive interface (e.g., facemask) is considered medically necessary when ALL of the following criteria are met:

- Individual has ONE of the following AND lack of ventilator support would result in a life-threatening condition:
 - a neuromuscular disease
 - a thoracic restrictive disease
 - chronic respiratory failure consequent to chronic obstructive pulmonary disease
 - congenital central hypoventilation syndrome
 - chronic lung disease of infancy (e.g., bronchopulmonary dysplasia)
 - obesity hypoventilation syndrome
 - restrictive disorder of chest wall
- Individual has demonstrated failure of bilevel positive airway pressure (BPAP) to improve hypercapnia and/or oxygen saturation level
- Individual does not require ventilation continuously (24 hours/day)

CONTINUED USE

An in-home ventilator is considered medically necessary for continued use when all of the following criteria are met:

- the pretreatment clinical condition met initial criteria
- there is documentation of compliant usage

MULTI-FUNCTION (HCPCS E0467)

The Ventilation, Oxygen, Cough, Suction, Nebulization (VOCSN) multifunction ventilator (HCPCS E0467) is considered medically necessary for an individual who meets the medical necessity coverage criteria for a home ventilator AND has a requirement for MORE than one of the four additional functions (oxygen concentrator, cough stimulator, suction pump and nebulizer).

ADDITIONAL OR DUPLICATE DEVICES

An additional home ventilator device (HCPCS E0465-E0467) is considered medically necessary for EITHER of the following conditions:

- **For mobility when all of the following criteria are met:**
 - the individual meets the medical necessity coverage criteria for the specified home ventilator
 - the individual requires mechanical ventilation during mobility (e.g., for use in wheelchair) as prescribed in their plan of care
 - the primary ventilator is not portable or not suitable for use with a wheelchair
- **In addition to negative pressure ventilator when both of the following criteria are met:**
 - the individual meets the medical necessity coverage criteria for the specified home ventilator
 - a negative pressure ventilator is required for part of the day and a positive pressure ventilator is necessary during the rest of the day, as prescribed in their plan of care

An additional or duplicate home ventilator device (HCPCS E0465-E0467) is considered NOT medically necessary as a back-up device (similar device as the individual's primary ventilator, for multiple residences or to have in case of possible malfunction).

NOT MEDICALLY NECESSARY

A home ventilator device (HCPCS E0465-E0467) is considered NOT medically necessary for any of the following:

- a non-life-threatening condition
- when the sole purpose of the home ventilator is to function as a respiratory assistance device (RAD) including continuous positive airway pressure (CPAP), auto-titrating PAP (APAP), bilevel positive airway pressure (BPAP, BiPAP), adaptive servo-ventilation (ASV), average volume assured pressure support (AVAPS), or intelligent volume assured pressure support (iVAPS)
- treatment for obstructive sleep apnea

General Background

A ventilator, also known as a respirator, can help a person breathe when they are unable to breathe sufficiently on their own. A mechanical ventilator can take over the act of breathing completely or make breathing easier by assisting weakened respiratory muscles. Mechanical ventilation is indicated in respiratory failure, which is defined as insufficient oxygenation, insufficient alveolar ventilation, or both. The desired effect of mechanical ventilation is to maintain appropriate levels of Po₂ and Pco₂ in arterial blood while unloading the ventilatory muscles.

Ventilators can be classified as either invasive or non-invasive. Invasive ventilators use an endotracheal tube or tracheostomy tube to deliver oxygen to the patient. Non-invasive ventilators use nasal masks, oral masks, nasal pillows, or a full-face mask to deliver oxygen to the patient.

Positive Pressure Ventilation

The majority of all mechanical ventilatory support is provided by positive pressure ventilation. Positive pressure ventilators developed as a more effective breathing option to the larger, bulkier negative pressure devices (e.g., iron lung). Since the 1980s, computer technology has enabled manufacturers to produce even smaller, lightweight ventilators that are easier to transport and operate, and are better suited for people living at home. Home ventilators may be used via:

- an invasive interface (i.e., tracheostomy) or
- a noninvasive interface (i.e., nose or mouth mask, chest shell).

After careful evaluation and pulmonary function tests to assess breathing and lung function and capacity, the physician recommends a type of ventilator and appropriate interfaces. In the USA, physicians work with respiratory therapists in home health care companies that provide the equipment, supplies, and education, training and monitoring. Individuals who need to use ventilation only at night have different equipment requirements than those who need to use a ventilator around the clock. Sometimes an individual may not be comfortable with a specific ventilator or interface and may need to change them in order to find the most comfortable and effective system. Some ventilator users alternate modes and interfaces during the day and night. There are some clinical scenarios that warrant both a primary ventilator and a secondary ventilator. An example is if an individual requires one type of ventilator (e.g., a negative pressure ventilator with a chest shell) during the day and needs a different type of ventilator (e.g., positive pressure ventilator with a face mask) during the night.

Multi-mode ventilators can provide many modes of ventilation, such as pressure support, pressure control, volume control, assist/control, synchronized intermittent mandatory ventilation (SIMV), bilevel positive pressure (BiPAP, BPAP) or continuous positive airway pressure (CPAP). Examples include LTV[®] series (CareFusion BD), iVent 201[®] (GE Healthcare), Trilogy 100, 200, 202 (Philips Respironics Inc), and VOCSN Unified Respiratory System (Ventec Life Systems, Inc.).

Utilizing a home ventilator as a respiratory assist device (RAD) (e.g., continuous positive airway pressure (CPAP), auto-titrating PAP (APAP), bilevel positive airway pressure (BPAP, BiPAP), adaptive servo-ventilation (ASV), average volume assured pressure support (AVAPS), or intelligent volume assured pressure support (iVAPS)) is considered not medically necessary. Numerous RADs have received FDA approval and may include minimum weight and/or age requirements that vary by device.

Food and Drug Administration (FDA)

Numerous positive and negative pressure ventilators are FDA-approved. One such example is the Ventilation, Oxygen, Cough, Suction, Nebulization (VOCSN) Unified Respiratory System (Ventec Life Systems, Inc., Bothell, Washington USA) which received 510(k) approval on April 7, 2017. Also known as the Ventec One-Circuit™, VOCSN is a multifunction ventilator that can provide five therapies or a mix of therapies from a single device. Therapies provided by VOCSN include: critical care ventilation; 6 L/min oxygen concentrator; touch button cough; hospital grade suction and high performance nebulizer. The VOCSN is intended to provide continuous or intermittent positive pressure ventilator support for the care of individuals who require mechanical ventilation. It may be used in invasive and noninvasive applications. The VOCSN is intended for pediatric through adult patients weighing at least 5 kg. It is intended for use in home, hospital, institutional and transport settings, including portable applications. The integral oxygen concentrator is intended for the administration of supplemental oxygen. The integral suction pump is intended for airway fluid removal and oral/ pharyngeal hygiene. The integral cough assist option is intended for patients who are additionally unable to cough or clear secretions effectively.

Literature Review

The Agency for Healthcare Research and Quality (AHRQ) Technology Assessment on Noninvasive Positive Pressure Ventilation in the Home (Final report 2/04/2020) evaluated home noninvasive positive pressure ventilation (NIPPV) in adults with chronic respiratory failure in terms of initiation, continuation, effectiveness, adverse events, equipment parameters and required respiratory services. Devices evaluated were home mechanical ventilators (HMV), bi-level positive airway pressure (BPAP) devices, and continuous positive airway pressure (CPAP) devices.

Of the included 68 studies, 14 evaluated home mechanical ventilator (HMV). Key messages include:

- In patients with COPD, home NIPPV as delivered by a BPAP device (compared to no device) was associated with lower mortality, intubations, hospital admissions, but no change in quality of life (low to moderate strength of the body of evidence [SOE]). NIPPV as delivered by a HMV device (compared individually with BPAP, CPAP, or no device) was associated with fewer hospital admissions (low SOE). In patients with thoracic restrictive diseases, HMV (compared to no device) was associated with lower mortality (low SOE). In patients with neuromuscular diseases, home BPAP (compared to no device) was associated with lower mortality and better quality of life (low SOE). In patients with obesity hypoventilation syndrome, HMV/BPAP mix (compared to no device) was associated with lower mortality (low SOE). BPAP (compared to no device) was associated with improved sleep quality.
- Current evidence is insufficient to assess the comparative effectiveness of many NIPPV device capabilities on patient outcomes; particularly comparing HMV to BPAP. Future studies should address which device capabilities are associated with improved patient outcomes.
- Criteria to initiate home NIPPV and home respiratory services were summarized in this report but varied and were not validated in comparative studies.
- Incidence of non-serious adverse events such as facial rash, dry eyes, mucosal dryness, and mask discomfort across devices was approximately 0.3 events over a median duration of device used of 6 months. The most commonly reported serious adverse event was acute respiratory failure. Based on direct comparisons, we found no statistically significant differences in number of treatment withdrawals or adverse events when comparing different devices or when comparing device use with no device use.

Professional Societies/Organizations

American Association for Respiratory Care (AARC): The AARC Clinical Practice Guideline Long-term Invasive Mechanical Ventilation in the Home (2007) refers to patients ventilated by positive pressure via a tracheostomy tube in the home. Patients requiring invasive long-term ventilator support have demonstrated:

- an inability to be completely weaned from invasive ventilatory support, or
- a progression of disease etiology that requires increasing ventilatory support.

Conditions that met these criteria may include but are not limited to ventilatory muscle disorders, alveolar hypoventilation syndrome, primary respiratory disorders, obstructive lung diseases, restrictive lung diseases, and cardiac disorders, including congenital anomalies.

Contraindications to invasive long term mechanical ventilation in the home include:

- the presence of a physiologically unstable medical condition requiring higher level of care or resources than available in the home
- patient's choice not to receive home mechanical ventilation
- lack of an appropriate discharge plan
- unsafe physical environment as determined by the patient's discharge planning team
- inadequate resources for care in the home

American Thoracic Society (ATS) Long-Term Noninvasive Ventilation in Chronic Stable Hypercapnic Chronic Obstructive Pulmonary Disease: The ATS Clinical Practice Guideline Long-Term Noninvasive Ventilation in Chronic Stable Hypercapnic Chronic Obstructive Pulmonary Disease (Macrea, et al., 2020) purpose is to summarize the available evidence and provide actionable recommendations addressing:

- patients with COPD, especially potential subgroups who might benefit from NIV therapy
- the ideal timing and location (e.g., hospital or sleep laboratory vs. home) for NIV initiation
- the identification of optimal modes and settings for chronic NIV therapy.

American Thoracic Society (ATS) Mechanical Ventilation in Adult Patients with Acute Respiratory Distress Syndrome: The ATS Clinical Practice Guideline Mechanical Ventilation in Adult Patients with Acute Respiratory Distress Syndrome (Fan, et al., 2016) is an evidence-based clinical practice guideline on the use of ventilatory strategies and associated cointerventions in adult patients with acute respiratory distress syndrome (ARDS), providing treatment recommendations on the basis of these interventions. It does NOT specifically address home setting or portable ventilation.

- The recommendations for the following interventions for the treatment of ARDS are strong:
 - Mechanical ventilation using lower tidal volumes (4–8 ml/kg predicted body weight) and lower inspiratory pressures (plateau pressure, 30 cm H₂O) (moderate confidence in effect estimates)
 - Prone positioning for more than 12 h/d in severe ARDS (moderate confidence in effect estimates)
- The recommendation against the following intervention for the treatment of ARDS is strong:
 - Routine use of high-frequency oscillatory ventilation in patients with moderate or severe ARDS (high confidence in effect estimates)
- The recommendation for the following interventions for the treatment of ARDS is conditional:
 - Higher positive end-expiratory pressure in patients with moderate or severe ARDS (moderate confidence in effect estimates)
 - Recruitment maneuvers in patients with moderate or severe ARDS (low confidence in effect estimates)
- Additional evidence is necessary to make a definitive recommendation for or against the use of extracorporeal membrane oxygenation in patients with severe ARDS.

American Thoracic Society (ATS) Pediatric Chronic Home Invasive Ventilation: The ATS Clinical Practice Guideline Pediatric Chronic Home Invasive Ventilation (Sterni, et al., 2016) is an evidence-based clinical practice guideline for health care professionals for the safe hospital discharge and home/community management of children requiring chronic invasive ventilation. The recommendations presented are based on the current evidence and expert opinion and will require an update as new evidence and/or technologies become available. Recommendations regarding home equipment include:

- The ATS suggest the following pieces of equipment for use in the home when caring for a patient on home mechanical ventilation: the ventilator, a back-up ventilator, batteries, a self-inflating bag and mask,

suctioning equipment (portable), heated humidifier, supplemental oxygen for emergency use, nebulizer, and a pulse oximeter (nonrecording) (Strength of Recommendation: Conditional; Quality of Evidence: Very Low).

- The ATS suggest that a mechanical insufflation–exsufflation (MI-E) device be used to help maintain airway patency in patients requiring home mechanical ventilation with ineffective cough, including, but not limited to, those with neuromuscular disease with poor respiratory muscle strength (Strength of Recommendation: Conditional; Quality of Evidence: Very Low).
- For children requiring chronic home invasive ventilation, the ATS suggest monitoring, especially when the child is asleep or unobserved, with a pulse oximeter rather than use of a cardiorespiratory monitor or sole use of the ventilator alarms (Strength of Recommendation: Conditional; Quality of Evidence: Very Low).
- For children requiring chronic home invasive ventilation, the ATS recommends regular maintenance of home ventilators and all associated equipment as outlined by the manufacturer.

American Thoracic Society (ATS) Management of COPD Exacerbations: The ERS/ATS Clinical Practice Guideline on the Management of COPD Exacerbations (Wedzicha 2017) notes the following:

- Should NIV be used in patients who are hospitalized with a COPD exacerbation associated with acute or acute-on-chronic respiratory failure? For hospitalized patients with acute or acute-on-chronic hypercapnic respiratory failure due to a COPD exacerbation, we recommend the use of NIV (strong recommendation, low quality of evidence).
- Should a home-based management programme (hospital-at-home) be implemented in patients with COPD exacerbations? For patients with a COPD exacerbation who present to the emergency department or hospital, we suggest a home-based management programme (hospital-at-home; conditional recommendation, moderate quality of evidence).

American Thoracic Society (ATS) and European Respiratory Society (ERS) Clinical Practice Guideline Noninvasive Ventilation for Acute Respiratory Failure: The ATS/ERS guideline does NOT specifically address home setting or portable ventilation (Rochweg 2017). It makes recommendations regarding the current use of NIV for various forms of respiratory failure encountered in acute care settings.

American Academy of Neurology (AAN): The AAN provides the following recommendations in their practice parameter update on the care of the patient with Amyotrophic Lateral Sclerosis (Miller, et al., 2009; Reaffirmed on January 11, 2020):

Respiratory Management

- Nocturnal oximetry may be considered to detect hypoventilation (regardless of the forced vital capacity, FVC) (Level C*).
- Supine FVC and maximal inspiratory pressure (MIP) may be considered useful in routine respiratory monitoring, in addition to the erect FVC (Level C).
- Sniff nasal pressure (SNP) may be considered to detect hypercapnia and nocturnal hypoxemia (Level C).
- Noninvasive ventilation (NIV) may be considered to enhance quality of life (QOL) in patients with ALS who have respiratory insufficiency (Level C).
- Tracheostomy invasive ventilation (TIV) may be considered to preserve QOL in patients with ALS who want long-term ventilator support (Level C).
- 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 (Level C).
- Mechanical insufflation/exsufflation (MIE) may be considered to clear secretions in patients with ALS who have reduced peak cough flow, particularly during an acute chest infection (Level C).
- There are insufficient data to support or refute high frequency chest wall oscillation (HFCWO) for clearing airway secretions in patients with ALS (Level U*).

Recommendations for Future Research:

- Evaluate SNP as a criterion for NIV initiation.

- Evaluate the impact of early NIV initiation on survival and quality of life.
- Assess the impact of executive dysfunction on NIV compliance.
- Evaluate the effect of hypoventilation on executive dysfunction.
- Compare techniques for clearing upper airway secretions at various stages of respiratory and bulbar dysfunction.
- Evaluate pulmonary tests, compliance with NIV, and outcomes in patients with bulbar dysfunction.

* C = Possibly effective, ineffective or harmful (or possibly useful/predictive or not useful/predictive) for the given condition in the specified population. (Level C rating requires at least one Class II study or two consistent Class III studies.)

U = Data inadequate or conflicting; given current knowledge, treatment (test, predictor) is unproven.

American Academy of Neurology (AAN) Consensus-based care recommendations for adults with myotonic dystrophy type 1: The AAN care recommendations (Ashizawa, et al 2018) note the following:

- Under section Life threatening symptoms—Clinical care recommendations
 - Vaccinate for pneumonia and flu; treat respiratory infections quickly and use cough assistance and mechanical ventilation as needed along with obtaining consultations from respiratory therapy and pulmonary medicine groups.
 - Some patients will eventually require either nighttime ventilator support or full-time ventilation. Most patients with chronic respiratory insufficiency respond to noninvasive ventilatory support (NIV). Patients experiencing acute respiratory failure require endotracheal intubation with positive pressure ventilation.
 - For chronic respiratory insufficiency, use supplemental oxygen with caution and in conjunction with NIV (see Surgery, anesthesia, and pain).
 - If surgery is planned, reassess clearance capacity if needed, possible adaptation to NIV or cough assistance.
- Under section re Excessive daytime sleepiness (EDS) symptoms:
 - If nocturnal or daytime hypoventilation is suspected, consider noninvasive positive pressure ventilation, and refer to a pulmonologist with experience in neuromuscular diseases re: possible need for NIV launching.
- Under Pregnancy / Obstetrical management:
 - Include a pediatric or neonatal specialist present at delivery; intensive neonatal care is recommended for neonates that may have DM1; anticipate need for feeding tube and ventilator support (recommended even if the fetus is known to be unaffected).

Use Outside of the US

The German National Guideline for Treating Chronic Respiratory Failure with Invasive and Non-Invasive Ventilation (Windisch, et al., 2018) notes that invasive and non-invasive home mechanical ventilation have become a well-established treatment option. Consequently, new chapters have been added to the guideline.

Non-Invasive Ventilation Recommendations

- Home access to non-invasive ventilation must be granted after considering the technical advantages and disadvantages, the individual patient tolerance levels, and the results of clinical testing.
- Commercial masks are usually sufficient. Customized masks must only be used under exceptional circumstances such as in neuromuscular disease patients, with high ventilation pressures or long ventilation times, or in patients with sensitive skin.
- Every patient should have a spare mask.
- Hybrid or special ventilation modes such as mandatory target volume cannot be recommended on a general basis.
- A second ventilation machine and an external battery are necessary for ventilation times ≥ 16 h/24 h.

Invasive Ventilation Recommendations

- The tracheostoma should be surgically created; proof of stability is absolutely necessary for percutaneous dilational tracheostomata.
- A second ventilation machine and an external battery are essential for ventilation times ≥ 16 h/24 h.
- A pulse oximeter is necessary for selective measurements and possibly for continuous measurements in certain disease groups (i.e., spinal cord transection, paediatrics).
- A small-diameter spare cannula must be made available.
- For application of speaking valves, a cuffless cannula, a completely unblocked cannula, or a fenestrated cannula must be used.
- Conditioning (humidification and moistening) of the ventilation air is mandatory.
- Two aspirator devices are necessary.

Obstructive Airway disease Recommendations

- Non-invasive ventilation is the primary therapeutic option for home mechanical ventilation of chronic obstructive pulmonary disease patients with chronic respiratory failure.
- Home non-invasive ventilation must be initiated when the patient presents with chronic hypercapnia in combination with the typical symptoms of respiratory failure.
- Home non-invasive ventilation must be initiated when hypercapnia persists for more than 2 weeks after completing acute ventilation therapy for acute respiratory acidosis.
- The aim of the ventilation therapy is to normalise PaCO₂; this may require sufficiently high ventilation intensity with effective ventilation pressures as well as the use of controlled ventilation modes.

Thoracic-Restrictive Lung Diseases Recommendations

- Non-invasive ventilation is the primary therapeutic option for home mechanical ventilation of thoracic-restrictive disease patients with chronic respiratory failure. The most important criteria for beginning long-term non-invasive ventilation are hypercapnia in combination with both the typical symptoms of chronic respiratory failure and the reduction in quality of life.
- For symptoms of hypoventilation in the absence of hypercapnia, a sleep study should take place.
- Patients with severe restrictive ventilatory dysfunction but without manifest hypercapnia must be closely monitored.

Obesity Hypoventilation Syndrome Recommendations

- Continuous positive airway pressure or non-invasive ventilation are the primary treatment options for home mechanical ventilation in patients with obesity hypoventilation syndrome.
- A primary attempt at continuous positive airway pressure therapy must take place under poly(somno)graphic evaluation. Primary non-invasive ventilation can be indicated in the presence of significant comorbidities or severe hypercapnia.
- A switch to non-invasive ventilation is indicated in the following cases: continuous positive airway pressure intolerance, persistent sleep-related or aggravated hypoventilation, and/or persistent sleep-related respiratory dysfunction despite continuous positive airway pressure use.
- Concomitant weight loss should be aimed for.
- A therapeutic switch from non-invasive ventilation to continuous positive airway pressure should be considered under stable conditions, particularly after weight reduction; if applicable, omission of mechanical ventilation can be considered.
- If obesity hypoventilation syndrome is clinically suspected, appropriate diagnostic tests for obesity hypoventilation syndrome should take place before undergoing general anaesthetics.

Neuromuscular Diseases Recommendations

- Clinical assessment and determination of forced vital capacity in neuromuscular disease patients should take place at 3- to 12-month intervals. A forced vital capacity $< 70\%$ is an indicator for poly(somno)graphy and PtcCO₂ measurement.
- In the event of mild hypercapnia (PaCO₂ ≥ 45 mm Hg or diurnal normocapnia with a nocturnal rise in PtcCO₂ of ≥ 10 mm Hg), non-invasive ventilation should be implemented as the primary therapy option for home mechanical ventilation in neuromuscular disease patients with chronic respiratory failure, if diurnal symptoms persist.

- There is no indication for prophylactic implementation of non-invasive ventilation in the absence of chronic respiratory failure.
- The measurement of coughing capacity is obligatory in neuromuscular disease patients. The presence of weak cough (peak cough flow < 270 L/min) is an indication for the introduction of secretion management.

Secretion Management Recommendations

- In neuromuscular disease patients with impaired cough and invasive ventilation, secretion management is obligatory and must be carried out regularly and adapted to the underlying disease.
- The indication for a mechanical In-Exsufflator device must be established by a physician with ventilation competency, who also then takes over the responsibility for the patient's therapy.
- Adaptation of the mechanical In-Exsufflator to the patient can be performed by a respiratory therapist.
- Switching between different types of mechanical In-Exsufflators that are not structurally identical may not be carried out without renewed testing of the effectiveness and patient compatibility of the devices; this is because the performance of each device may differ significantly.
- Secretion management in chronic obstructive pulmonary disease patients with high secretion load must include measures for secretolysis (medicinal/non-medicinal) and measures to ease coughing such as special coughing techniques (huffing) and positive expiratory pressure systems.
- Mechanical In-Exsufflators should not be used in chronic obstructive pulmonary disease patients.
- Suctioning in invasively ventilated patients must take place strictly via endotracheal placement of the suctioning catheter (max. 1 cm beyond the cannula), independent of the underlying condition.

Spinal Cord Transection Recommendations

- Permanent pulse oximetry monitoring is necessary in partly and fully dependent mechanically ventilated patients with spinal cord transection.
- Capnographic monitoring of patients with spinal cord transection injuries is indicated in many situations in the home mechanical ventilation setting.

Special Features of Pediatric Ventilation

- Due to their difference from adults, the care and ventilation of children with chronic respiratory failure must be carried out by a multidisciplinary team.
- Safe and successful home mechanical ventilation therapy must take into account the family and home environment of the patient and be accompanied by offers of medical, nursing, and psychosocial support.
- Continuous monitoring of oxygen saturation (at the least), and PCO₂ (if necessary) must be carried out in children who are dependent on mechanical ventilation for survival.
- The development of paediatric competence centres that initiate home mechanical ventilation should be aimed at for better care of long-term-ventilated children (Windisch, et al., 2018).

The Canadian Thoracic Society Clinical Practice Guideline on Home Mechanical Ventilation (McKim, et al, 2011) notes that guideline applies to all adult individuals who are at risk for or are using HMV. Individuals with amyotrophic lateral sclerosis (ALS), central hypoventilation syndrome (CHS), chronic obstructive pulmonary disease (COPD), kyphoscoliosis, obesity hypoventilation syndrome (OHS), spinal cord injury (SCI), Duchenne muscular dystrophy (DMD), muscular dystrophies (MDs) other than DMD, myopathies and myotonic dystrophy (Steinert's muscular dystrophy [SMD]) are of special interest and are considered individually in the present clinical practice guideline.

In an update to the 2011 guideline on home mechanical ventilation, the Canadian Thoracic Society (Rimmer, et al., 2019) published a clinical practice guideline that focuses specifically on patients with amyotrophic lateral sclerosis. The guideline provided the following recommendations in regard to non-invasive ventilation (NIV):

- "We recommend NIV for patients with ALS meeting initiation criteria provided it is in keeping with patient preferences and values (GRADE 1B)
- We recommend that practitioners offer NIV to patients with any one of the following:
 - Orthopnea* (GRADE 1B)
 - Daytime arterial or capillary pCO₂>45 mmHg (GRADE 1B)

- Sleep disordered breathing as defined by oxygen saturation < 90% for > 5% of the night or < 88% for 5 consecutive minutes or a 10mmHg increase in TcCO₂ during sleep AND any of the following symptoms: dyspnea, morning headache, daytime sleepiness, or non-refreshing sleep. (GRADE 2B)
- FVC < 50% predicted** (GRADE 1B)
- FVC sitting or supine <80% predicted with symptoms as indicated in Recommendation 2.1 (a) and any other indicator of respiratory muscle involvement including SNIP <-50cmH₂O, or MIP< -65 cmH₂O in males and < -55 in females. (GRADE 1C)
- SNP < -40 cmH₂O or MIP < -40 cmH₂O** (GRADE 1C)
- We suggest that practitioners discuss the option of tracheostomy ventilation with ALS patients. (GRADE 2B)
- We strongly recommended that practitioners discuss this option well in advance of acute respiratory failure. (GRADE 1C)”

Strength of recommendations grading system:

- 1A: Strong recommendation, high-quality evidence; benefits clearly outweigh risk and burdens or vice versa; consistent evidence from randomized controlled trials without important limitations or exceptionally strong evidence from observational studies.
- 1B: Strong recommendation, moderate-quality evidence; benefits clearly outweigh risk and burdens or vice versa; evidence from randomized controlled trials with important limitations (inconsistent results, methodologic flaws, indirect or imprecise) or very strong evidence from observational studies.
- 1C: Strong recommendation, low- or very-low-quality evidence; benefits clearly outweigh risk and burdens or vice versa; evidence for at least one critical outcome from observational studies, case series, or randomized controlled trials, with serious flaws or indirect evidence.
- 2A: Weak recommendation, high-quality evidence; benefits closely balanced with risks and burden; consistent evidence from randomized controlled trials without important limitations or exceptionally strong evidence from observational studies.
- 2B: Weak recommendation, moderate-quality evidence; benefits closely balanced with risks and burden; evidence from randomized controlled trials with important limitations (inconsistent results, methodologic flaws, indirect or imprecise) or very strong evidence from observational studies.
- 2C: Weak recommendation, low or very-low-quality evidence; uncertainty in the estimates of benefits, risks, and burden; benefits, risk, and burden may be closely balanced; evidence for at least one critical outcome from observational studies, case series, or randomized controlled trials, with serious flaws or indirect evidence.

In another update to the 2011 guideline on home mechanical ventilation, the Canadian Thoracic Society (Kaminska, et al., 2021) published a clinical practice guideline that focuses specifically on patients with chronic hypercapnic respiratory failure due to advanced COPD. The guideline provided the following recommendations in regards to non-invasive ventilation (NIV):

- “In patients with stable severe COPD and chronic hypercapnic respiratory failure (PaCO₂ ≥ 52 mm Hg), we suggest long-term NIV to improve survival. (weak/conditional recommendation, low certainty evidence)
- In patients with severe COPD on LTOT who remain significantly hypercapnic (persistent PaCO₂> 52 mmHg) for at least 2 weeks after discontinuing NIV for an acute exacerbation, we suggest long-term NIV to delay hospital readmission. (weak/conditional recommendation, very low certainty evidence)
- We suggest high-intensity non-invasive ventilation instead of low-intensity non-invasive ventilation to improve PaCO₂ in patients with COPD and chronic hypercapnic respiratory failure (persistent PaCO₂> 52 mmHg). (weak/conditional recommendation, low certainty evidence)
- We do not recommend the use of volume-assured pressure-preset NIV over standard pressure-preset NIV in patients with COPD and chronic hypercapnic respiratory failure. (strong recommendation, low certainty evidence)”

The European Respiratory Society published recommendations (Ergan, et al., 2019) for the use of long-term home non-invasive ventilation (LTH-NIV) for the management of COPD:

- “The ERS task force suggests LTH-NIV be used for patients with chronic stable hypercapnic COPD (conditional recommendation, low certainty evidence).
- The ERS task force suggests LTH-NIV be used in patients with COPD following a life-threatening episode of acute hypercapnic respiratory failure requiring acute NIV, if hypercapnia persists following the episode (conditional recommendation, low certainty evidence).
- The ERS task force suggests using fixed pressure support mode as first-choice ventilator mode in patients with COPD using LTH-NIV (conditional recommendation, very low certainty evidence).”

Medicare Coverage Determinations

	Contractor	Determination Name/Number	Revision Effective Date
NCD	National	Durable Medical Equipment (280.1)	05/05/2005
LCD		No Determination found	

Note: Please review the current Medicare Policy for the most up-to-date information. (NCD = National Coverage Determination; LCD = Local Coverage Determination.)

Coding Information

- Note:** 1) This list of codes may not be all-inclusive.
 2) Deleted codes and codes which are not effective at the time the service is rendered may not be eligible for reimbursement.

Considered Medically Necessary when criteria in the applicable policy statements listed above are met:

HCPCS Codes	Description
E0465	Home ventilator, any type, used with invasive interface, (e.g., tracheostomy tube)
E0466	Home ventilator, any type, used with non-invasive interface, (e.g., mask, chest shell)
E0467	Home ventilator, multi-function respiratory device, also performs any or all of the additional functions of oxygen concentration, drug nebulization, aspiration, and cough stimulation, includes all accessories, components and supplies for all functions

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