**Subject:** Diaphragmatic Phrenic Nerve Stimulation  
**Policy Number:** NMP310  
**Effective Date:** December 2006  
**Updated:** April 2016

This National Medical Policy is subject to the terms in the IMPORTANT NOTICE At the end of this document

For Medicaid Plans: Please refer to the appropriate State’s Medicaid manual(s), publication(s), citation(s), and documented guidance for coverage criteria and benefit guidelines prior to applying Health Net Medical Policies

**The Centers for Medicare & Medicaid Services (CMS)**  
For Medicare Advantage members please refer to the following for coverage guidelines first:

<table>
<thead>
<tr>
<th>Use</th>
<th>Source</th>
<th>Reference/Website Link</th>
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<tbody>
<tr>
<td></td>
<td>National Coverage Manual Citation</td>
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<td></td>
<td>Local Coverage Determination (LCD)*</td>
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<td>Article (Local)*</td>
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<tr>
<td></td>
<td>None</td>
<td>Use Health Net Policy</td>
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Instructions  
- Medicare NCDs and National Coverage Manuals apply to ALL Medicare members in ALL regions.
Medicare LCDs and Articles apply to members in specific regions. To access your specific region, select the link provided under “Reference/Website” and follow the search instructions. Enter the topic and your specific state to find the coverage determinations for your region. *Note: Health Net must follow local coverage determinations (LCDs) of Medicare Administration Contractors (MACs) located outside their service area when those MACs have exclusive coverage of an item or service. (CMS Manual Chapter 4 Section 90.2)

If more than one source is checked, you need to access all sources as, on occasion, an LCD or article contains additional coverage information than contained in the NCD or National Coverage Manual.

If there is no NCD, National Coverage Manual or region specific LCD/Article, follow the Health Net Hierarchy of Medical Resources for guidance.

**Current Policy**
Health Net, Inc. considers diaphragmatic or phrenic nerve stimulation medically necessary, with FDA approved devices, for the treatment of severe, chronic ventilatory insufficiency in patients whose remaining phrenic nerve, lung, and diaphragm function is sufficient to accommodate electrical stimulation, when one of the following criteria is met:

1. Central alveolar hypoventilation either primary or secondary to a brainstem disorder; or
2. Member has high quadriplegia at the upper cervical level, at or above the C3 vertebral level.

And

When all of the following are met:
1. There is integrity and viability of the intrathoracic section of the phrenic nerve;
2. Diaphragmatic function is sufficient to accommodate chronic stimulation;
3. Baseline estimated pulmonary function test is known or likely to be adequate;
4. Patient has normal chest anatomy, a normal level of consciousness, and has the ability to participate in and complete the training and rehabilitation associated with the use of the device.

Health Net, Inc. considers diaphragmatic or phrenic nerve stimulation (D/P nerve stimulation) not medically necessary and contraindicated in patients with any of the following:

- Inadequate ventilation without the use of mechanical ventilation; or
- Respiratory failure or insufficiency is anticipated to be temporary; or
- Insufficient phrenic nerve or diaphragm function unable to accommodate electrical stimulation; or
- Major chest wall deformities; or
- Trauma to the midcervical spine (third, fourth, and fifth levels) usually does not allow pacing, because the cell bodies of the phrenic nerves are damaged; or
- Diseases of the lower motor neurons or the anterior horn cells, such as polio or amyotrophic lateral sclerosis, do not permit pacing.

**Codes Related To This Policy**

**NOTE:**
The codes listed in this policy are for reference purposes only. Listing of a code in this policy does not imply that the service described by this code is a covered or non-covered health service. Coverage is determined by the benefit documents and medical necessity criteria. This list of codes may not be all inclusive.

On October 1, 2015, the ICD-9 code sets used to report medical diagnoses and inpatient procedures have been replaced by ICD-10 code sets.

**ICD-9 Codes**

<table>
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<tr>
<th>Code</th>
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<tr>
<td>327.24</td>
<td>Idiopathic sleep related nonobstructive alveolar hypoventilation</td>
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<tr>
<td>327.25</td>
<td>Congenital central alveolar hypoventilation syndrome</td>
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<tr>
<td>344.00</td>
<td>Unspecified quadriplegia</td>
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<td>344.01</td>
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<td>344.02</td>
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<tr>
<td>518.5</td>
<td>Pulmonary insufficiency following trauma and surgery</td>
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<td>518.83</td>
<td>Chronic respiratory failure</td>
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<tr>
<td>786.09</td>
<td>Central alveolar hypoventilation</td>
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<tr>
<td>806.00</td>
<td>Fracture of vertebral column with spinal cord injury (cervical, C1-C4)</td>
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<td>907.2</td>
<td>Late effect of spinal cord injury</td>
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<td>952.00</td>
<td>Spinal cord injury without evidence of spinal bone injury (cervical, C1-C4)</td>
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**ICD-10 Codes**

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<td>G82.51</td>
<td>Quadriplegia, C1-C4 complete</td>
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<td>G82.52</td>
<td>Quadriplegia, C1-C4 incomplete</td>
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<td>J95.821</td>
<td>Acute postprocedural respiratory failure</td>
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<td>J96.00</td>
<td>Acute respiratory failure, unspecified whether with hypoxia or hypercapnia</td>
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<td>J96.10</td>
<td>Chronic respiratory failure, unspecified whether with hypoxia or hypercapnia</td>
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<td>R06.89</td>
<td>Other abnormalities of breathing</td>
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<tr>
<td>S12.000-S12.491</td>
<td>Fracture of first–fourth cervical vertebrae</td>
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<td>S14.101</td>
<td>Unspecified injury at C1 level of cervical spinal cord,</td>
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<tr>
<td>S14.102</td>
<td>Unspecified injury at C2 level of cervical spinal cord</td>
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<td>Unspecified injury at C3 level of cervical spinal cord</td>
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<td>S14.104</td>
<td>Unspecified injury at C4 level of cervical spinal cord</td>
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**CPT Codes**

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<th>Description</th>
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<tr>
<td>64580</td>
<td>Incision for implantation of neurostimulator electrode array; neuromuscular</td>
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**HCPCS Codes**
Diaphragmatic Phrenic Nerve Stimulation Apr 16

Scientific Rationale – Update April 2016
McDermott et al. (2015) completed a multicentre, open-label, randomised controlled trial at seven specialist ALS and respiratory centres in the UK. Eligible participants were aged 18 years or older with laboratory supported probable, clinically probable, or clinically definite ALS; stable riluzole treatment for at least 30 days; and respiratory insufficiency. We randomly assigned participants (1:1), via a centralised web-based randomisation system with minimisation that balanced patients for age, sex, forced vital capacity, and bulbar function, to receive either non-invasive ventilation plus pacing with the NeuRx RA/4 Diaphragm Pacing System or non-invasive ventilation alone. Patients, carers, and outcome assessors were not masked to treatment allocation. The primary outcome was overall survival, defined as the time from randomisation to death from any cause. Analysis was by intention to treat. This trial is registered, ISRCTN number 53817913. Between Dec 5, 2011, and Dec 18, 2013, we randomly assigned 74 participants to receive either non-invasive ventilation alone (n=37) or non-invasive ventilation plus diaphragm pacing (n=37). On Dec 18, 2013, the Data Monitoring and Ethics Committee (DMEC) recommended suspension of recruitment on the basis of overall survival figures. Randomly assigned participants continued as per the study protocol until June 23, 2014, when the DMEC advised discontinuation of pacing in all patients. Follow-up assessments continued until the planned end of the study in December, 2014. Survival was shorter in the non-invasive ventilation plus pacing group than in the non-invasive ventilation alone group (median 11·0 months [95% CI 8·3–13·6] vs 22·5 months [13·6–not reached]); adjusted hazard ratio 2·27, 95% CI 1·22–4·25; p=0·009). 28 (76%) patients died in the pacing group and 19 (51%) patients died in the non-invasive ventilation alone group. We recorded 162 adverse events (5·9 events per person-year) in the pacing group, of which 46 events were serious, compared with 81 events (2·5 events per person-year) in the non-invasive ventilation alone group, of which 31 events were serious. Addition of diaphragm pacing to standard care with non-invasive ventilation was associated with decreased survival in patients with ALS. Our results suggest that diaphragmatic pacing should not be used as a routine treatment for patients with ALS in respiratory failure. This study was funded by The National Institute for Health Research Health Technology Assessment Programme; the Motor Neurone Disease Association of England, Wales, and Northern Ireland.

Scientific Rationale – Update September 2012
National Institute for Health and Clinical Excellence (NICE, July 22, 2009), has a summary on ‘Phrenic Nerve Stimulation.’ Diaphragmatic pacing by phrenic nerve stimulation using implanted electrodes in the neck or chest is an alternative for some patients with spinal cord injury or neurological disease.

The Mark IV Breathing Pacemaker System (Avery Biomedical Devices Inc.) is approved for marketing in the United States. The original premarket approval was prior to 1987; modifications to the device were approved in March 2003. The Mark IV
is indicated for persons who require chronic ventilatory support because of upper motor neuron respiratory muscle paralysis or central alveolar hypoventilation and whose remaining phrenic nerve, lung, and diaphragm function are sufficient to accommodate electrical stimulation (Center for Devices and Radiological Health [CDRH], 2011).

The NeuRx DPS RA/4 Respiratory System (Synapse Biomedical Inc.) accomplishes respiratory pacing using intramuscular electrodes inserted via laparoscopy. Under this report is intended to provide research assistance and general information only. It is not intended to be used as the sole basis for determining coverage policy. Under the Humanitarian Device Exemption (HDE), the FDA approved the device to be used by patients aged 18 years or older. The device is intended for use in patients with stable, high spinal cord injuries with stimulatable diaphragms, to allow the patients to breathe without the assistance of a mechanical ventilator for at least 4 continuous hours per day (Center for Devices and Radiological Health [CDRH], 2008).

There is a Clinical Trial on ‘Humanitarian Device Exemption Post-Approval Study of NeuRx Diaphragm Pacing System for Amyotrophic Lateral Sclerosis’. This study is currently recruiting participants, and was verified August 2012, with the ClinicalTrials.gov Identifier of NCT01605006. This post-approval study will follow 60 participants who have ALS, documented chronic hypoventilation, and bilateral phrenic nerve function, and who undergo the surgical implantation procedure to receive the NeuRx Diaphragm Pacing System device. Participants who are successfully implanted with the device will use it for daily diaphragm conditioning sessions. Participants will be followed for at least two years (until the last enrolled participant reaches the 2-year follow-up visit). Safety and probable benefit outcome measures will be assessed.

There is another Clinical Trial on ‘Diaphragmatic Pacemaker in Tetraplegic Patients With Spinal Cord Injuries’. This study is currently recruiting participants, and was verified in June 2011, with the ClinicalTrials.gov Identifier of NCT01385384. Permanent dependency of breathing apparatus due to spinal cord injury is traditionally treated with different types of mechanical ventilation. However, the electric ventilation became a possibility through their most current versions, such as diaphragmatic pacemakers. Diaphragmatic pacemakers rhythmically stimulates the diaphragm to replace the functions of the respiratory center that doesn't works well or is inaccessible. However, this modality has the prerequisite that the phrenic nerve and diaphragm muscle are normal. The reason for the development of diaphragmatic pacemaker freeing the patient from the ventilator. By using the mechanical energy of the diaphragm of the patient, the patient may come not need the ventilator tubing, tracheostomy, and with the help of their caregivers, the inconvenient mechanical ventilators. The enrollment in this study is only 5 and the estimated study completion date is July 2012, although it is not completed at this time.

**Scientific Rationale – Update October 2011**

Amyotrophic lateral sclerosis (ALS) is another cause of respiratory failure and is a form of motor neuron disease. ALS is a progressive and fatal disease, with the average lifespan of patients diagnosed with ALS only 3 to 5 years. The disease is characterized by motor neuron degeneration of the cerebral cortex, brainstem and spinal cord, although the cause is unknown. ALS causes progressive muscle weakening and wasting, and loss of ability to initiate and control voluntary movement. As the disease progresses, the mechanical function of the respiratory system is impaired, and pulmonary complications are a major cause of death in ALS.
Ounders et al. (2009) completed a study that summarized the largest series of surgical cases in ALS during multicenter prospective trials of the laparoscopic diaphragm pacing system (DPS) to delay respiratory failure. Fifty-one patients were implanted from March 2005 to March 2008 at 2 sites. Age at implantation ranged from 42 to 73 years and the percent predicted forced vital capacity (FVC) ranged from 20% to 87%. On preoperative blood gases, Pco2 was as high as 60. Using this protocol, there were no failures to extubate or 30-day mortalities. The DPS system increases the respiratory system compliance by decreasing posterior lobe atelectasis and can stimulate respirations at the end of each case. Although the results were positive and promising, this was a small study of only 51 patients. Larger, peer-reviewed comparative or randomized controlled studies are necessary to determine the safety, efficacy and long-term outcomes of diaphragmatic and phrenic nerve stimulation for individuals with ALS.

Scientific Rationale – Update February 2011
Khong et al (2010) reviewed the data on 19 patients treated with phrenic nerve stimulation. Of the 19 patients, 14 required pacing due to quadriplegia, one had congenital central hypoventilation syndrome and one had brainstem encephalitis. Information was unavailable for the remaining three patients. Currently, 11 of the pacers are known to be actively implanted, with the total pacing duration ranging from 1 to 21 years (mean 13 years). Eight of the 19 patients had revision surgeries. Four of these were to replace the original I-107 system (which had a 3-5-year life expectancy) with the current I-110 system, which is expected to perform electrically for the patient’s lifetime. Three patients had revisions due to mechanical failure. The remaining patients’ notes were incomplete. These data suggest that phrenic nerve stimulation can be used instead of mechanical ventilators for long-term ongoing respiratory support.

Scientific Rationale
Diaphragmatic or phrenic nerve stimulation (D/P nerve stimulation) or pacing is an alternative to mechanical ventilation for specific patients with chronic respiratory insufficiency. This is also known as electrophrenic respiration, in which an implanted pacemaker is used to stimulate the phrenic nerves with regular electrical pulses. Candidates for D/P nerve stimulation must have intact phrenic nerves and diaphragms as well as adequate residual pulmonary function.

The most important part of the respiratory system is the diaphragm, which is innervated by the cervical motor neurons C3-5 via the phrenic nerves. The viability of the phrenic nerves is best determined by percutaneous electrical stimulation at the neck, with fluoroscopy used to detect diaphragmatic movement. This produces negative pressure in the chest and allows air to enter the lungs as in normal breathing.

The diaphragm has both an expiratory action on the upper rib cage and an inspiratory action on the lower rib cage. This dome shaped muscle separates the abdominal and the thoracic cavity.

The diaphragm consists of the following two separate portions joined by a central tendon:
- The crural portion consists of fibers originating from the first three lumbar vertebral bodies and the medial and lateral ligaments. This part of the
Diaphragm receives its impulses from the fourth and fifth cervical segments and is not attached to the ribcage.

- The costal portion arises from the inner surfaces and upper margins of the lower six ribs and the sternum. This area derives its innervation from the third and fourth cervical segments and exerts an insertional force on the rib cage. Diaphragmatic or phrenic nerve stimulation (D/P nerve stimulation) or pacing is an alternative to mechanical ventilation for specific patients with chronic respiratory insufficiency. This is also known as electrophrenic respiration, in which an implanted pacemaker is used to stimulate the phrenic nerves with regular electrical pulses.

**Initial Evaluation for D/P Pacing**
Patients are considered candidates for diaphragmatic/ phrenic nerve pacing if they meet all of the following criteria:

- Relatively normal cognitive function.
- Complete respiratory paralysis without recovery for more than three months after the injury or onset of apnea. (This would include *Central Sleep Apnea or Ondine’s curse which is also known as (CCHS) congenital central hypoventilation syndrome)
- Viable lower motor neurons for the phrenic nerves.

**Assessment of phrenic nerve function**
Timing of the testing of the phrenic nerve function is somewhat controversial, but most agree that a minimum of three months should elapse following the acute event before testing is attempted. One study, for example, showed that phrenic nerves may be responsive when tested very early after injury, but subsequently become unresponsive within this initial three month period. The same investigators also demonstrated that initially unresponsive nerves may become responsive up to two years after injury. Thus, patients who are initially excluded as candidates for diaphragmatic pacing should be periodically retested for at least two years after the injury to determine if there has been recovery of the phrenic nerves.

**Indications for D/P nerve stimulator**
The device has been used successfully to treat hypoventilation caused by a variety of conditions including:

A. Respiratory paralysis resulting from lesions of the brain stem.
B. Alternative to management of patients with respiratory insufficiency who are dependent upon the usual therapy of intermittent or permanent use of a mechanical ventilator as well as maintenance of a permanent tracheotomy stoma.
C. Diaphragmatic pacing should be considered for patients who have a central nervous system cause for apnea. The lesion in the central nervous system must be above the third cervical level, since pacing of the diaphragm by phrenic nerve stimulation is possible only if the nerve cell bodies (located in the anterior horns of C3-5) are viable.
D. Basilar Meningitis
**Contraindications for D/P nerve Stimulator**

This procedure is contraindicated in the following scenarios:

A. Defects in the respiratory nerves and muscles.

B. Trauma to the mid-cervical spine (third, fourth, and fifth levels) usually does not allow pacing, because the cell bodies of the phrenic nerves are damaged. Likewise, diseases of the lower motor neurons or the anterior horn cells, such as polio or amyotrophic lateral sclerosis, do not permit pacing.

C. Failure of the diaphragms to contract with percutaneous stimulation of the phrenic nerves.

D. Coma.

E. Severe primary pulmonary disease.

**Pacing System**

The pacing system consists of electrodes sutured to the subcutaneous tissue of the phrenic nerves in the neck or in the chest. The external transmitter controls the intensity, duration and rate of impulse, and thus the respiratory rate. This device is small and light with an antenna secured externally over the receiver. The equipment needed to receive D/P nerve stimulation treatment is small enough to be worn in a pocketed belt or vest, and allows considerable freedom for patients who may be ambulatory or use a wheelchair.

Fluoroscopy is used to determine the current that produces maximal diaphragm excursion. The intensity and duration of the stimulus are adjusted during a training and rehabilitation period. This would establish the optimal contraction of the diaphragm and thus the maximal tidal volume using the lowest applied current. The stimulation rate is set to the most suitable age and clinically appropriate rate.

A Diaphragmatic / Phrenic nerve stimulator class III device currently FDA premarket (PMA) approved (i.e., for marketing in the United States) is manufactured by Avery Laboratories, Inc. (Commack, NY). The original premarket (PMA) approval was prior to 1987; recent modifications to the device were approved in January 2001. The modified device, marketed under the trade name Avery Mark IV is indicated for persons who require chronic ventilatory support because of upper motor neuron respiratory muscle paralysis or central alveolar hypoventilation and whose remaining phrenic nerve, lung and diaphragm function is sufficient to accommodate electrical stimulation (FDA, 2002).

Meyers et al. (2006) performed the largest series published to date, including 165 patients, of whom 27% were paced on a full-time basis and 63% on a part-time basis. Phrenic nerve pacing met the ventilatory requirements of 47% of the patients and was partially successful in 36%. Phrenic nerve pacing remains a relatively new procedure for selected patients with partial or complete respiratory insufficiency caused by a variety of conditions. This would include respiratory paralysis resulting from lesions of the brain stem and cervical spinal cord and chronic pulmonary disease with ventilatory insufficiency. This procedure is intended to be an alternative for patients with respiratory insufficiency who are dependent upon a mechanical ventilator as well as maintenance of a permanent tracheotomy stoma. Any patient considered for this technology must have an intact phrenic nerve and diaphragm.

Elefteriades et al.,(2002) have performed studies over a 10-15 year span on pacing devices. They have reported no clinical deterioration in pacing parameters or
respiratory measurements from continuous pacing. Pathologic studies have demonstrated integrity of the phrenic nerve and preserved diaphragm histology.

Garrido-Garcia et al. (1998) presented a prospective uncontrolled trial of patients with chronic ventilatory failure treated with electrophrenic respiration. The study included 13 males and nine females with a mean age of 12. The etiology included 13 patients with tetraplegia and five patients with sequelae of surgical treatment of intracranial lesions, and four with central alveolar hypoventilation. The mean duration of the conditioning period was 3-4 months. Eighteen patients (81.8%) achieved permanent, diaphragmatically paced breathing with bilateral stimulation and in four (18.2%) patients pacing was used only during sleep. Five patients died (22.7%) (i.e., two died during the hospital stay, three at home; two deaths had unknown causes, three deaths were due to lack of at-home care, recurrence of an epidermoid tumor and accidental disconnection of the mechanical ventilation before beginning the conditioning period). Two cases were considered failures (i.e., one patient had transitory neurapraxia lasting 80 days and the other had an ischemic spinal cord syndrome with progressive deterioration of the left-side response to stimulation). One patient had right phrenic nerve entrapment by scar tissue and four suffered infections. The follow-up periods since pacemaker implantation were: one for 11 years; four for 10 years, and 17 patients for less than five years. The authors concluded that this study demonstrated that complete stable ventilation can be achieved using diaphragmatic pacing and that it improves the prognosis and life quality of patients with severe chronic respiratory failure.

Complications of D/P Pacing
Complications of diaphragmatic / phrenic pacing include those related to the surgery and those related to the process of pacing. These include any of the following:

1. Infection following surgery.
2. Pulmonary complications following a thoracic surgical approach.
3. Dislodgement of the pacer electrode.
4. Transmission of pacer impulses to the brachial plexus, with rhythmic jerking of the upper extremity (seen with neck placement of the electrode).
5. Malfunction of hardware.

Central Alveolar Hypoventilation Syndrome
‘Central Alveolar Hypoventilation Syndrome’ is characterized by ventilatory impairment, resulting in arterial oxygen desaturation content. A failure of ventilation promptly increases the partial pressure of carbon dioxide measured by arterial blood gas analysis (PaCO2). This condition is worsened by sleep, and occurs in patients with normal mechanical properties of the lung.

Alveolar hypoventilation is caused by several disorders that are collectively referred to as hypoventilation syndromes. Alveolar hypoventilation also is a cause of hypoxemia, a deficient oxygenation of the blood. Thus, patients who hypoventilate may develop clinically significant hypoxemia. Alveolar hypoventilation includes the presence of hypoxemia along with hypercapnia, the presence of an excess of carbon dioxide in the blood. The combination of the decreased oxygenation and increased carbon dioxide in the blood, aggravates the clinical manifestations seen with hypoventilation syndromes.
Diaphragmatic pacing has also been used to treat patients with central alveolar hypoventilation syndrome. Yasuma et al (1998) noted that the respiratory assistance by the diaphragm pacemaker or the use of a mechanical ventilator as a backup was highly useful for the home care of a patient with central alveolar hypoventilation.

Girsch, et al. (1996) noted that ventilatory insufficiency due to central hypoventilation syndrome and spinal cord injury (SCI) can be treated even in children with diaphragm pacing, provided the indication for implantation, containing medical and social aspects, was made correctly.

**Ondine’s Curse**

Ondine’s Curse, is a form of primary alveolar hypoventilation, in which the cause of hypoventilation and hypercapnia is not known. Patients with primary alveolar hypoventilation have normal alveolar-arterial oxygen gradients and are able to voluntarily hyperventilate and normalize their PaCO2.

Congenital central hypoventilation syndrome (CCHS), also known as Ondine’s curse, is a very rare and serious form of ‘Central Sleep Apnea’. It is a failure from birth of the central nervous system’s autonomic control over breathing while asleep. There are usually no breathing problems while awake, since the voluntary control of ventilation, which operates during waking hours, is generally intact. The exact pathophysiology of CCHS remains unknown but has been the subject of intense research. About 1 in every 200,000 children has this condition. In 2006, there are only about 200 known cases worldwide.

Associated with this syndrome, there can, in addition, be problems with the motility of the esophagus and the colon. Reduced esophageal motility causes food to move too slowly down to the stomach. Reduced motility of the colon produces a condition called Hirschsprung disease (megacolon) with severe constipation and even obstruction. In addition, the heart rate may be abnormally slow.

Patients who have congenital central hypoventilation syndrome (CCHS) generally require tracheotomies and lifetime mechanical ventilation on a respirator in order to survive. Most people with congenital Ondine’s curse do not survive infancy, although they can be kept alive with a ventilator. Mitsuyama et al (2003) performed a study that confirmed that phrenic nerve pacing is an appropriate alternative to a mechanical ventilator for patients with CCHS.

Chen and Keens (2004) reported that all patients with CCHS require lifelong ventilatory support during sleep, but some will be able to maintain adequate ventilation without assistance while awake once past infancy. Daytime diaphragm pacing in children with CCHS provides greater mobility than mechanical ventilation. Modalities of home mechanical-assisted ventilation include positive pressure ventilation via tracheostomy, non-invasive positive pressure ventilation (bi-level ventilation), negative pressure ventilation and diaphragmatic pacers.

Shaul, et al. (2002) stated that diaphragmatic pacing can provide chronic ventilatory support for children who suffer from congenital central hypoventilation syndrome (CCHS) or cervical spinal cord injury. Between 1997-2000, nine children ranging in age from 5-15 and suffering from these disorders underwent thoracoscopic placement of bilateral phrenic nerve electrodes. Four patients experienced postoperative complications (i.e.,

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pneumonia, atelectasis, bradycardia, and pneumothorax). Average follow-up has been 30 months. Eight patients have reached their long-term pacing goals. The authors concluded that phrenic nerve electrodes can be implanted thoracoscopically and allow the successful use of diaphragmatic pacing therapy. Avoidance of thoracotomy with its associated perioperative morbidity and scarring may encourage wider diaphragmatic pacing in children.

Weese-Mayer et al, per the American Thoracic Society (1999), reports that as the infant becomes ambulatory the possibility of diaphragm pacing by phrenic nerve stimulation should be considered to allow for increased mobility and improved quality of life. The 24 h/d supported patients will still require a tracheostomy for the nighttime mechanical ventilation. Although diaphragm pacing is not typically recommended for the young child who requires only nighttime support (the benefits do not outweigh the risks), for the older child this might be an appropriate consideration.

Flageole et al (1995) stated that pediatric surgeons should be aware of congenital central hypoventilation syndrome (CCHS) because it may be treated with surgically implanted electrodes that allow for pacing of the diaphragm. The technique has an acceptable complication rate, and it can greatly decrease the impact of the disease on the lifestyle and activity of the patient.

Spinal Cord Injury
Spinal cord injury (SCI) can cause various degrees of neurological impairment depending on the location and severity of the injury. The degree of impairment is the neurological extent of injury, namely the amount of tissue trauma to the spinal cord at the level of injury. If the spinal cord is seriously damaged at the injury site, there is complete loss of sensation and voluntary muscle control below the level of lesion. On the other hand, if the damage is not complete, some sensory and/or motor functions may still be preserved. Thus, a complete injury to the cervical spine will result in quadriplegia, while an incomplete injury to the cervical spine will result in quadriparesis. In this policy, diaphragmatic or phrenic nerve stimulation specifically refers to the interruption of neuronal conduction at the upper cervical level, at or above the C3 vertebrae.

Creasey, et al. (1996) reported that electrical stimulation has been used for over 25 years to restore breathing to patients with high quadriplegia causing respiratory paralysis and to patients with central alveolar hypoventilation. Three groups have developed electrical pacing systems for long-term support of respiration. The systems differ principally in the electrode design and stimulation waveform. Approximately 1,000 people have received one of the three phrenic pacing devices, most with strongly positive results: reduced risk of tracheal problems and chronic infection, the ability to speak and smell more normally, reduced risk of accidental interruption of respiration, greater independence, and reduced costs and time for ventilatory care.

Glenn et al. (1984) reported a case study of five patients with respiratory paralysis and quadriplegia. The patients required full-time ventilatory support by continuous electrical pacing of both hemidiaphragm simultaneously for 11-33 months, through the application of a low frequency stimulus to the phrenic nerves. The authors reported that the strength and endurance of the diaphragm muscle increased with pacing. Biopsy specimens taken from two patients who had uninterrupted stimulation
for six and 16 weeks respectively showed changes suggestive of the development of fatigue-resistant muscle fibers. When comparing these results with those of earlier experience with intermittent unilateral stimulation of the diaphragm in 17 patients with respiratory paralysis it was found that continuous bilateral pacing using low-frequency stimulation appeared to be superior because of more efficient ventilation of both lungs. The authors report that for patients with respiratory paralysis and intact phrenic nerves, continuous simultaneous pacing of both hemidiaphragm with low-frequency stimulation and a slow respiratory rate is a satisfactory method of providing full-time ventilatory support.

Diaphragmatic/phrenic (D/P) nerve stimulation is generally considered not medically necessary for patients who can achieve adequate ventilation without the use of mechanical ventilation or when respiratory failure or insufficiency is anticipated to be temporary. Results from a number of uncontrolled prospective and retrospective studies indicate that D/P pacing is a viable alternative for carefully selected patients who would otherwise be dependent on mechanical ventilators (Hayes, 2005). Thus, candidates for diaphragm pacing are potentially ambulatory patients who require ventilatory support 24 h/d via tracheotomy and who do not exhibit significant ventilator-related lung damage. Diaphragm pacer settings must provide adequate alveolar ventilation and oxygenation during rest and daily activities.

Evidence evaluated for this policy was obtained primarily from a search of the peer-reviewed published literature, over a twenty year time period. A limited number of prospective and retrospective studies evaluating D/P nerve stimulation were identified in the search. Clinical trials that have studied the efficacy of D/P stimulation have included various patient populations, including both adult and pediatric patients with spinal cord injuries, congenital central alveolar hypoventilation syndrome and other causes of respiratory failure. None of the studies were controlled trials, and none provided a direct comparison between D/P nerve stimulation and noninvasive methods of ventilatory support, although most provided long follow-up times, usually years.

The use of diaphragmatic pacing as a means to improve ventilation and eliminate the need for continuous positive pressure ventilatory support was introduced over thirty years ago. D/P pacing can lead to a significant improvement in the quality of life of ventilator-dependent quadriplegic individuals. It can also help improve pulmonary function and reduce the incidence of pulmonary infections. Most patients are able to speak while being paced, which contributes substantially to its clinical value.

**Review History**

<table>
<thead>
<tr>
<th>Date</th>
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<tr>
<td>December 2006</td>
<td>Medical Advisory Council</td>
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<tr>
<td>December 2007</td>
<td>Update – no revisions</td>
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<tr>
<td>February 2011</td>
<td>Update – no revisions</td>
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<tr>
<td>October 2011</td>
<td>Update. No revisions</td>
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<tr>
<td>September 2013</td>
<td>Update – no revisions. Code updates</td>
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<tr>
<td>April 2014</td>
<td>Update – no revisions.</td>
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<tr>
<td>April 2015</td>
<td>Update – no revisions.</td>
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**This policy is based on the following evidence-based guideline:**

References – Update April 2016

References – Update April 2015

References – Update April 2014

References – Update September 2013

References Update – September 2012

References Update – October 2011

References Update – February 2011

References Initial


**Important Notice**

**General Purpose.**
Health Net's National Medical Policies (the "Policies") are developed to assist Health Net in administering plan benefits and determining whether a particular procedure, drug, service or supply is medically necessary. The Policies are based upon a review of the available clinical information including clinical outcome studies in the peer-reviewed published medical literature, regulatory status of the drug or device, evidence-based guidelines of governmental bodies, and evidence-based guidelines and positions of select national health professional organizations. Coverage determinations are made on a case-by-case basis and are subject to all of the terms, conditions, limitations, and exclusions of the member's contract, including medical necessity requirements. Health Net may use the Policies to determine whether under the facts and circumstances of a particular case, the proposed procedure, drug, service or supply is medically necessary. The conclusion that a procedure, drug, service or supply is medically necessary does not constitute coverage. The member's contract defines which procedure, drug, service or supply is covered, excluded, limited, or subject to dollar caps. The policy provides for clearly written, reasonable and current criteria that have been approved by Health Net's National Medical Advisory Council (MAC). The clinical criteria and medical policies provide guidelines for determining the medical necessity criteria for specific procedures, equipment, and services. In order to be eligible, all services must be medically necessary and otherwise defined in the member's benefits contract as described this "Important Notice" disclaimer. In all cases, final benefit determinations are based on the applicable contract language. To the extent there are any conflicts between medical policy guidelines and applicable contract language, the contract language prevails. Medical policy is not intended to override the policy that defines the member’s benefits, nor is it intended to dictate to providers how to practice medicine.

**Policy Effective Date and Defined Terms.**
The date of posting is not the effective date of the Policy. The Policy is effective as of the date determined by Health Net. All policies are subject to applicable legal and regulatory mandates and requirements for prior notification. If there is a discrepancy between the policy effective date and legal mandates and regulatory requirements, the requirements of law and regulation shall govern. * In some states, prior notice or posting on the website is required before a policy is deemed effective. For information regarding the effective dates of Policies, contact your provider representative. The Policies do not include definitions. All terms are defined by Health Net. For information regarding the definitions of terms used in the Policies, contact your provider representative.

**Policy Amendment without Notice.**
Health Net reserves the right to amend the Policies without notice to providers or Members. In some states, prior notice or website posting is required before an amendment is deemed effective.

**No Medical Advice.**
The Policies do not constitute medical advice. Health Net does not provide or recommend treatment to members. Members should consult with their treating physician in connection with diagnosis and treatment decisions.
No Authorization or Guarantee of Coverage.
The Policies do not constitute authorization or guarantee of coverage of particular procedure, drug, service or supply. Members and providers should refer to the Member contract to determine if exclusions, limitations, and dollar caps apply to a particular procedure, drug, service or supply.

Policy Limitation: Member’s Contract Controls Coverage Determinations.
Statutory Notice to Members: The materials provided to you are guidelines used by this plan to authorize, modify, or deny care for persons with similar illnesses or conditions. Specific care and treatment may vary depending on individual need and the benefits covered under your contract. The determination of coverage for a particular procedure, drug, service or supply is not based upon the Policies, but rather is subject to the facts of the individual clinical case, terms and conditions of the member’s contract, and requirements of applicable laws and regulations. The contract language contains specific terms and conditions, including pre-existing conditions, limitations, exclusions, benefit maximums, eligibility, and other relevant terms and conditions of coverage. In the event the Member’s contract (also known as the benefit contract, coverage document, or evidence of coverage) conflicts with the Policies, the Member’s contract shall govern. The Policies do not replace or amend the Member’s contract.

Policy Limitation: Legal and Regulatory Mandates and Requirements
The determinations of coverage for a particular procedure, drug, service or supply is subject to applicable legal and regulatory mandates and requirements. If there is a discrepancy between the Policies and legal mandates and regulatory requirements, the requirements of law and regulation shall govern.

Reconstructive Surgery
CA Health and Safety Code 1367.63 requires health care service plans to cover reconstructive surgery. “Reconstructive surgery” means surgery performed to correct or repair abnormal structures of the body caused by congenital defects, developmental abnormalities, trauma, infection, tumors, or disease to do either of the following:

1. To improve function or
2. To create a normal appearance, to the extent possible.

Reconstructive surgery does not mean “cosmetic surgery,” which is surgery performed to alter or reshape normal structures of the body in order to improve appearance.

Requests for reconstructive surgery may be denied, if the proposed procedure offers only a minimal improvement in the appearance of the enrollee, in accordance with the standard of care as practiced by physicians specializing in reconstructive surgery.

Reconstructive Surgery after Mastectomy
California Health and Safety Code 1367.6 requires treatment for breast cancer to cover prosthetic devices or reconstructive surgery to restore and achieve symmetry for the patient incident to a mastectomy. Coverage for prosthetic devices and reconstructive surgery shall be subject to the co-payment, or deductible and coinsurance conditions, that are applicable to the mastectomy and all other terms and conditions applicable to other benefits. “Mastectomy” means the removal of all or part of the breast for medically necessary reasons, as determined by a licensed physician and surgeon.

Policy Limitations: Medicare and Medicaid
Policies specifically developed to assist Health Net in administering Medicare or Medicaid plan benefits and determining coverage for a particular procedure, drug, service or supply for Medicare or Medicaid members shall not be construed to apply to any other Health Net plans and members. The Policies shall not be interpreted to limit the benefits afforded Medicare and Medicaid members by law and regulation.