Description:

Electrophrenic pacing, also known as diaphragmatic/phrenic (D/P) nerve stimulation or breathing pacemaker, is intended as an alternative to mechanical ventilation in selected individuals with ventilatory insufficiency (or failure) who have retained adequate function in their phrenic nerves and diaphragm. The D/P nerve stimulator is an implanted device that acts as a pacemaker by providing regular electrical pulses to stimulate the phrenic nerves. Stimulation of the nerves then causes the diaphragm to contract, which produces negative pressure in the chest, allowing air to enter the lungs. 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 individuals who may be ambulatory or confined to a wheelchair. The stimulator consists of an externally worn transmitter and implanted receiver with electrodes. The receiver and electrodes for these devices may be placed either by open thoracotomy or laparoscopically.

Electrophrenic pacemakers are contraindicated in the following situations:

- Pre-operative screening tests do not demonstrate that phrenic nerves, lungs, and diaphragm can sustain ventilation by electrical stimulation;
- The patient has another serious disorder that might affect nerve conduction (e.g., tumors, vascular disease, diabetes, multiple sclerosis).

The diaphragmatic/phrenic pacing systems (Mark IV™ Avery Biomedical Devices, Inc. and NeuRx Diaphragm Pacing System (DPS) Synapse Biomedical Inc.) have been approved by the U.S. Food and Drug Administration (FDA).
**POSITION STATEMENT:**
The use of a FDA approved electrophrenic pacemaker meets the definition of medical necessity for members with permanent, severe hypoventilation related to one of the following conditions:

- **Quadriplegia** (high C3 or above)
- Central *alveolar hypoventilation* syndrome.

The use of a FDA approved electrophrenic pacemaker meets the definition of medical necessity when used as an alternative to invasive mechanical ventilation in members with motor neuron disease, for example *amyotrophic lateral sclerosis (ALS)*, when ALL of the following criteria are met:

- Diaphragm movement with stimulation is visible under fluoroscopy; **AND**
- Stimulation of the diaphragm directly results in sufficient muscle activity to accommodate independent breathing without the support of a ventilator; **AND**
- Member 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.

The use of electrophrenic pacemaker does not meet the definition of medical necessity when:

- Member can subsist independently of a mechanical respirator; **OR**
- Respiratory insufficiency is temporary.

The use of an electrophrenic pacemaker is considered experimental or investigational, as there is insufficient clinical evidence to support the effectiveness of this therapy for all other applications, and specifically for the following:

- Chronic obstructive pulmonary disease;
- Treatment of hiccups;
- Young children and infants.

**BILLING/CODING INFORMATION:**
There is no specific code describing electrophrenic pacemaker devices.

The following codes may be used for implantation and revision or removal of these devices:

<table>
<thead>
<tr>
<th>CPT Coding</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>64585</td>
<td>Revision or removal of peripheral neurostimulator electrode array</td>
</tr>
<tr>
<td>Code</td>
<td>Description</td>
</tr>
<tr>
<td>-------</td>
<td>-----------------------------------------------------------------------------</td>
</tr>
<tr>
<td>64590</td>
<td>Incision or replacement of peripheral or gastric neurostimulator pulse generator or receiver, direct or inductive coupling</td>
</tr>
<tr>
<td>64595</td>
<td>Revision or removal of peripheral or gastric neurostimulator pulse generator or receiver</td>
</tr>
</tbody>
</table>
**HCPCS Coding**

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>L8696</td>
<td>Antenna (external) for use with implantable diaphragmatic/phrenic nerve stimulation device, replacement, each</td>
</tr>
</tbody>
</table>

**REIMBURSEMENT INFORMATION:**
Refer to section entitled POSITION STATEMENT.

**PROGRAM EXCEPTIONS:**

Federal Employee Program (FEP): Follow FEP guidelines.

State Account Organization (SAO): Follow SAO guidelines.

Medicare Advantage products:

The following National Coverage Determination (NCD) was reviewed on the last guideline reviewed date: Phrenic Nerve Stimulator, (160.19) located at cms.gov. No Local Coverage Determination (LCD) were found at the time of the last guideline reviewed date.

**DEFINITIONS:**

Alveolar ventilation: the volume of gas (air) expired from the alveoli to the outside of the body per minute.

Amyotrophic lateral sclerosis (ALS): a debilitating disease with varied etiology characterized by rapidly progressive weakness, muscle atrophy, muscle spasticity, difficulty speaking (dysarthria), difficulty swallowing (dysphagia), and difficulty breathing (dyspnea); also known as Lou Gehrig’s disease.

Diaphragm: the thin muscle below the lungs and heart that separates the chest from the abdomen.

Hypoventilation: a state in which an abnormally low amount of air enters the lungs.

Phrenic nerve: is mainly the motor nerve of the diaphragm.

Quadriplegia: paralysis of all four limbs (i.e., arms, legs).

**RELATED GUIDELINES:**

Percutaneous Electric Nerve Stimulation (PENS), 02-61000-03

**OTHER:**
Other terms for describing these devices:

Diaphragm pacer
Diaphragm pacing
Electrophrenic pacemaker
Phrenic nerve pacer
Phrenic nerve stimulator
REFERENCES:


COMMITTEE APPROVAL:
This Medical Coverage Guideline (MCG) was approved by the BCBSF Medical Practice & Coverage Committee on 08/23/18.
**GUIDELINE UPDATE INFORMATION:**

<table>
<thead>
<tr>
<th>Date</th>
<th>Update Details</th>
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<tbody>
<tr>
<td>06/15/04</td>
<td>New Medical Coverage Guideline.</td>
</tr>
<tr>
<td>06/15/05</td>
<td>Scheduled review; no change in coverage statement.</td>
</tr>
<tr>
<td>07/15/06</td>
<td>Scheduled review: no change in coverage statement; added coding information.</td>
</tr>
<tr>
<td>07/15/07</td>
<td>Scheduled review; reformatted guideline; updated references.</td>
</tr>
<tr>
<td>07/15/08</td>
<td>Reviewed; position statement revised to include indications not considered medically necessary; updated references.</td>
</tr>
<tr>
<td>06/15/09</td>
<td>Scheduled review; no change in position statement; references updated.</td>
</tr>
<tr>
<td>10/01/09</td>
<td>HCPCS 4th quarter update; deleted ICD-9 diagnosis code 348.8 was removed from the guideline.</td>
</tr>
<tr>
<td>01/01/11</td>
<td>Revision; related ICD-10 codes added.</td>
</tr>
<tr>
<td>01/01/12</td>
<td>Annual HCPCS coding update: removed 64577.</td>
</tr>
<tr>
<td>02/15/13</td>
<td>Revision of position statement regarding covered indications; references updated.</td>
</tr>
<tr>
<td>05/11/14</td>
<td>Revision: Program Exceptions section updated.</td>
</tr>
<tr>
<td>01/01/15</td>
<td>Annual coding update: added L8696</td>
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<tr>
<td>11/01/15</td>
<td>Revision: ICD-9 Codes deleted.</td>
</tr>
<tr>
<td>10/01/16</td>
<td>Revision; billing/coding information section updated.</td>
</tr>
<tr>
<td>01/20/17</td>
<td>Revised 64585, 64590 and 64595 code descriptor.</td>
</tr>
<tr>
<td>09/15/18</td>
<td>Review; no change in position statement. Added FDA statement to description section. Updated references.</td>
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</tbody>
</table>