02-61000-33

Original Effective Date: 06/15/04

Reviewed: 02/26/26

Revised: 10/01/26

Subject: Diaphragmatic/Phrenic Nerve Stimulation (Electrophrenic Pacemaker)

THIS MEDICAL COVERAGE GUIDELINE IS NOT AN AUTHORIZATION, CERTIFICATION, EXPLANATION OF BENEFITS, OR A GUARANTEE OF PAYMENT, NOR DOES IT SUBSTITUTE FOR OR CONSTITUTE MEDICAL ADVICE. ALL MEDICAL DECISIONS ARE SOLELY THE RESPONSIBILITY OF THE PATIENT AND PHYSICIAN. BENEFITS ARE DETERMINED BY THE GROUP CONTRACT, MEMBER BENEFIT BOOKLET, AND/OR INDIVIDUAL SUBSCRIBER CERTIFICATE IN EFFECT AT THE TIME SERVICES WERE RENDERED. THIS MEDICAL COVERAGE GUIDELINE APPLIES TO ALL LINES OF BUSINESS UNLESS OTHERWISE NOTED IN THE PROGRAM EXCEPTIONS SECTION.

Position Statement	Billing/Coding	Reimbursement	Program Exceptions	Definitions	Related Guidelines
Other	References	Updates			

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** for all other applications, including but not limited to the following. The evidence is insufficient to determine the effects of electrophrenic pacemaker on health outcomes.

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

BILLING/CODING INFORMATION:

There is no specific code describing electrophrenic pacemaker devices.

CPT Coding

64585	Revision or removal of peripheral neurostimulator electrode array
64590	Incision or replacement of peripheral or gastric neurostimulator pulse generator or receiver, direct or inductive coupling
64595	Revision or removal of peripheral or gastric neurostimulator pulse generator or receiver

HCPCS Coding

L8697	External accessory for use with implantable phrenic nerve stimulation device, replacement, each

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.

If this Medical Coverage Guideline contains a step therapy requirement, in compliance with Florida law 627.42393, members or providers may request a step therapy protocol exemption to this requirement if based on medical necessity. The process for requesting a protocol exemption can be found at Coverage Protocol Exemption Request

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:

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3. DiMarco AF, Onders, RP, Kowalski, KE, et al: Dept. of Physiology, Case Western Reserve University and MetroHealth Medical Center, Cleveland Ohio. "Phrenic nerve pacing in a tetraplegic patient via intramuscular diaphragm electrodes". Am J Respir Crit Care Med 2002 Dec 15; 166(12 Pt 1): 1604-6.
4. Le Pimpec-Barthes F, Legras A, Arame A et al. Diaphragm pacing: the state of the art. Journal of Thoracic Disease 2016 Apr; 8 (Suppl 4): S376-S386.
5. Miller RG, Jackson CE, Kasarskis EJ, et al. Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: drug, nutritional, and respiratory therapies (an evidence-based review): report of the Quality Standards Subcommittee of the American Academy of Neurology. Neurology 2009 Oct 13;73(15):1218-26.
6. National Guideline Clearinghouse. Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: drug, nutritional, and respiratory therapies (an evidence-based review). Report of the Quality Standards Subcommittee of the American Academy of Neurology (2009).
7. Onders RP, Aiyar H, Mortimer JT. "Characterization of the human diaphragm muscle with respect to

the phrenic nerve motor points for diaphragmatic pacing". Am Surg. 2004 Mar; 70(3): 241-7.

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9. Shaul DB, DanielsonPD, McComb JG, et al. Thoracoscopic placement of phrenic nerve electrodes for diaphragmatic pacing in children. J Pediatr Surg. 2002 Jul; 37(7): 974-8.
10. U.S. Food and Drug Administration (FDA). Title 21 – Food and Drugs, Chapter 1 – Food and Drug Administration Department of Health and Human Services, Subchapter H – Medical Devices, Part 882 – Neurological Devices. Accessed 0610/08.
11. U.S. Food and Drug Administration (FDA) NeuRx DPS™ RA/4 Respiratory Stimulation System; Humanitarian Device Exemption (HDE) approval, September 2011.

COMMITTEE APPROVAL:

This Medical Coverage Guideline (MCG) was approved by the Florida Blue Medical Practice and Coverage Committee on 02/26/26.

GUIDELINE UPDATE INFORMATION:

06/15/04	New Medical Coverage Guideline.
06/15/05	Scheduled review; no change in coverage statement.
07/15/06	Scheduled review: no change in coverage statement; added coding information.
07/15/07	Scheduled review; reformatted guideline; updated references.
07/15/08	Reviewed; position statement revised to include indications not considered medically necessary; updated references.
06/15/09	Scheduled review; no change in position statement; references updated.
10/01/09	HCPCS 4th quarter update; deleted ICD-9 diagnosis code 348.8 was removed from the guideline.
01/01/11	Revision; related ICD-10 codes added.
01/01/12	Annual HCPCS coding update: removed 64577.
02/15/13	Revision of position statement regarding covered indications; references updated.
05/11/14	Revision: Program Exceptions section updated.
01/01/15	Annual coding update: added L8696
11/01/15	Revision: ICD-9 Codes deleted.
10/01/16	Revision; billing/coding information section updated.
01/20/17	Revised 64585, 64590 and 64595 code descriptor.
09/15/18	Review; no change in position statement. Added FDA statement to description section. Updated references.
09/15/20	Review; revised experimental or investigational position statement.
10/15/22	Review; no change in position statement.
08/21/23	Update to Program Exceptions section.
10/15/24	Review; no change in position statement. Updated references.
03/15/26	Position statements maintained.
10/01/26	Quarterly CPT/HCPCS update. Deleted L8696. Added L8697.