Clinical Policy: Diaphragmatic/Phrenic Nerve Stimulation

Reference Number: PA.CP.MP.203
Original Effective Date: 05/01/2021
Last Review Date: New PHW Policy

Coding Implications
Revision Log

Description

Diaphragmatic/phrenic nerve stimulation, also referred to as diaphragm pacing, is a treatment option used to eliminate or reduce the need for ventilator support in those with chronic ventilatory insufficiency due to bilateral paralysis or severe paresis of the diaphragm. Diaphragmatic/phrenic nerve stimulation uses the phrenic nerves to signal the diaphragm muscles to contract and produce breathing through electrical stimulation.

Policy/Criteria

I. It is the policy of PA Health & Wellness® that *diaphragmatic/phrenic nerve stimulation with the Mark IV™ Breathing Pacemaker System* is **medically necessary** when all of the following are met:

A. Stimulation is used as an alternative to mechanical ventilation for an individual with severe, chronic respiratory failure due to one of the following:
   1. Upper cervical spinal cord injury (at or above the C3 vertebral level);
   2. Central alveolar hypoventilation disorder;

B. Diaphragm movement with stimulation is visible under fluoroscopy;

C. Intact and sufficient function in the phrenic nerve, lungs, and diaphragm;

D. Stimulation of the diaphragm either directly or through the phrenic nerve results in sufficient muscle activity to accommodate independent breathing without the support of a ventilator;

E. Normal chest anatomy, a normal level of consciousness, and the ability to participate in and complete the training and rehabilitation associated with the use of the device.

II. It is the policy of PA Health & Wellness® that *diaphragmatic/phrenic nerve stimulation with the NeuRX DPS™ RA/4 Respiratory Stimulation System* is **medically necessary** when provided in accordance with the Humanitarian Device Exemption (HDE) specifications of the U.S Food and Drug Administration when the following criteria is met:

A. Stimulation is used as an alternative to mechanical ventilation for an individual with severe, chronic respiratory failure due to one of the following:
   1. Amyotrophic lateral sclerosis (ALS);
      a. Age 21 years or older;
      b. Experiencing chronic hypoventilation but not progressed to FVC (forced vital capacity) less than 45% predicted;
      c. Diaphragm movement with stimulation is visible under fluoroscopy;
      d. Intact and sufficient function in the phrenic nerve, lungs, and diaphragm.
   2. Upper cervical spinal cord injury (at or above the C3 vertebral level);
      a. Age 18 years or older;
      b. Diaphragm movement with stimulation is visible under fluoroscopy;
      c. Stimulation of the diaphragm will allow the individual to breathe without the assistance of a mechanical ventilator for at least four continuous hours a day;
      d. Intact and sufficient function in the phrenic nerve, lungs, and diaphragm.

III. It is the policy of PA Health & Wellness® that *diaphragmatic/phrenic nerve stimulation* is considered **investigational** for any other conditions, including but not limited to, *central sleep apnea*, because the evidence supporting its safety and efficacy is limited.

Background

Diaphragmatic/phrenic nerve stimulator devices are indicated for certain ventilator-dependent individuals who lack voluntary control of their diaphragm muscles to enable independent breathing without the assistance of a mechanical ventilator.

NeuRx DPS RA/4 Respiratory Stimulation System (Synapse Biomedical, Inc.)

FDA approval for distribution of the NeuRx DPS™ RA/4 Respiratory Stimulation System (Synapse Biomedical, Inc., Oberlin, OH) was granted under a Humanitarian Device Exemption (HDE) on June 17, 2008. The FDA-approved indications are: For use in patients with stable, high spinal cord injuries with stimulatable diaphragms, but lack control of their diaphragms. The device is indicated to allow the patients to breathe without the assistance of a mechanical ventilator for at least 4 continuous hours a day and is for use only in patients 18 years of age or older. This FDA approval is subject to the manufacturer developing an acceptable method of tracking device implantation to individual patient recipients.[6]

In 2011 the FDA approved the NeuRx DPS™ RA/4 Respiratory Stimulation System as a humanitarian-use device (HUD) in amyotrophic lateral sclerosis (ALS) following the submission of a humanitarian device exemption (HDE) application. The FDA approved indications are: “For use in amyotrophic lateral sclerosis (ALS) patients with a stimulatable diaphragm (both right and left portions) as demonstrated by voluntary contraction or phrenic nerve conduction studies, and who are experiencing chronic hypoventilation (CH) , but not progressed to an FVC less than 45% predicted. For use only in patients 21 years of age or older”.[7]

Mark IV™ Breathing Pacemaker System (Avery Biomedical Device, Inc.)

The Avery Breathing Pacemaker System (i.e., the Mark IV™ Avery Biomedical Device, Inc., Commack, NY) is the only other diaphragmatic/phrenic stimulator system approved for use by the FDA in the United States. The pacemaker is classified as a Class III neurologic therapeutic device requiring premarket approval (PMA). The device is approved "For persons who require chronic ventilatory support because of upper motor neuron respiratory muscle paralysis (RMP) or because of central alveolar hypoventilation (CAH) and whose remaining phrenic nerve, lung, and diaphragm function is sufficient to accommodate electrical stimulation".[8]

Coding Implications

This clinical policy references Current Procedural Terminology (CPT®). CPT® is a registered trademark of the American Medical Association. All CPT codes and descriptions are copyrighted 2020, American Medical Association. All rights reserved. CPT codes and CPT descriptions are from the current manuals and those included herein are not intended to be all-inclusive and are included for informational purposes only. Codes referenced in this clinical policy are for informational purposes only. Inclusion or exclusion of any codes does not guarantee coverage. Providers should reference the most up-to-date sources of professional coding guidance prior to the submission of claims for reimbursement of covered services.

CPT®* Codes	Description
64575	Incision for implantation of neurostimulator electrode array; peripheral nerve, (excludes sacral nerve)
64595	Revision or removal of peripheral or gastric neurostimulator pulse generator or receiver

HCPCS ®* Codes	Description
C1778	Lead, neurostimulator (implantable)
C1816	Receiver and/or transmitter, neurostimulator (implantable)
L8681	Patient programmer (external) for use with implantable programmable neurostimulator pulse generator, replacement only
L8689	External recharging system for battery (internal) for use with implantable neurostimulator, replacement only

ICD-10-CM Diagnosis Codes that Support Coverage Criteria
+ Indicates a code(s) requiring an additional character

ICD-10 Code	Description
G12.20	Motor neuron disease, unspecified
G12.21	Amyotrophic lateral sclerosis
G12.22	Progressive bulbar palsy
G12.23	Primary lateral sclerosis
G12.24	Familial motor neuron disease
G12.25	Progressive spinal muscle atrophy
G12.29	Other motor neuron disease
G47.35	Congenital central alveolar hypoventilation syndrome
G82.50	Quadriplegia, unspecified
G82.51	Quadriplegia, C1-C4 complete
G82.52	Quadriplegia, C1-C4 incomplete
G83.89	Paralytic syndrome, unspecified
J96.10	Chronic respiratory failure, unspecified whether with hypoxia or hypercapnia
J96.11	Chronic respiratory failure with hypoxia
J96.12	Chronic respiratory failure with hypercapnia
J96.20	Acute and chronic respiratory failure, unspecified whether with hypoxia or hypercapnia
J96.21	Acute and chronic respiratory failure with hypoxia
J96.22	Acute and chronic respiratory failure with hypercapnia
R06.81	Apnea, not elsewhere classified
Z99.11	Dependence on respirator [ventilator] status

Reviews, Revisions, and Approvals	Date	Approval Date
Approved by MPC. No changes. (Original approval date 08/11)	04/16	04/16

Reviews, Revisions, and Approvals	Date	Approval Date
Approved by MPC. No changes.	04/17	04/17
Approved by MPC. No changes.	03/18	03/18
Approved by MPC. No changes.	03/19	03/19
Approved by MPC. No changes.	04/20	04/20
Integrated diaphragmatic pacing criteria from PA.CP.MP.107 DME and Legacy WellCare Diaphragmatic Phrenic Nerve Stimulation HS-185 policy. Removed ICD-10-PCS codes and replaced with ICD-10-CM codes. Seperated criteria by FDA approved device. Added medical necessity criteria for amyotrophic lateral sclerosis (ALS), additional verbiage changes made with no clinical significance. Specialist reviewed. Background and references reviewed and updated. Replaced "member" with "member/enrollee" in all instances.	11/20	12/20
New Policy to PHW	4/2021	

References

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2. Marion DW. Pacing the diaphragm: Patient selection, evaluation, implantation and complications. In: UpToDate, Shefner JM (Ed), UpToDate, Waltham, MA. Accessed October 30, 2020.
3. Le Pimpec-Barthes F, Legras A, Arame A, et al. Diaphragm pacing: the state of the art. J Thorac Dis. 2016;8(Suppl 4):S376-S386. doi:10.21037/jtd.2016.03.97
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6. Premarket Notification Database: NeuRx DPS™ RA/4 Respiratory Stimulation System. Summary of Safety and Probable Benefit. U.S. Food and Drug Administration Center for Devices and Radiological Health Web site. https://www.accessdata.fda.gov/cdrh_docs/pdf7/H070003B.pdf Published June 17, 2008. Accessed November 2, 2020.
7. Premarket Approvals for the NeuRx DPS™ Diaphragm Pacing System. Summary of Safety and Probable Benefit. U.S. Food and Drug Administration Center for Devices and Radiological Health Website. https://www.accessdata.fda.gov/cdrh_docs/pdf10/H100006b.pdf Published September 28, 2011. Accessed November 2, 2020.
8. Premarket Approvals for the Avery Breathing Pacemaker System Mark IV™ (Avery Biomedical Device, Inc., Commack, NY). Summary of Safety and Effectiveness. U.S. Food and Drug Administration Center for Devices and Radiological Health Web site.

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