Clinical Policy: Diaphragmatic/Phrenic Nerve Stimulation

Reference Number: CP.MP.203 Coding Implications
Date of Last Revision: 12/21 Revision Log

See Important Reminder at the end of this policy for important regulatory and legal information.

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 health plans affiliated with Centene Corporation® 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 health plans affiliated with Centene Corporation® 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 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. 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 health plans affiliated with Centene Corporation® that there is insufficient evidence to support the safety and efficacy of diaphragmatic/phrenic nerve stimulation for any other conditions, including but not limited to, central sleep apnea.

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)
64580	Incision for implantation of neurostimulator electrode array; neuromuscular
64590	Insertion 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 ®* Codes	Description
C1778	Lead, neurostimulator (implantable)
C1816	Receiver and/or transmitter, neurostimulator (implantable)
L8680	Implantable neurostimulator electrode, each
L8681	Patient programmer (external) for use with implantable programmable neurostimulator pulse generator, replacement only
L8682	Implantable neurostimulator radiofrequency receiver [for phrenic nerve stimulator]
L8683	Radiofrequency transmitter (external) for use with implantable neurostimulator radiofrequency receiver [for phrenic nerve stimulator]
L8689	External recharging system for battery (internal) for use with implantable neurostimulator, replacement only
L8695	External recharging system for battery (external) for use with implantable neurostimulator, replacement only
L8696	Antenna (external) for use with implantable diaphragmatic/phrenic nerve stimulation device, replacement, each

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

ICD 10 Code	Description
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	Revision Date	Approval Date
Approved by MPC. No changes. (Original approval date 08/11)	04/16	04/16
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 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
Annual review. References reviewed, updated, and reformatted. Changed "review date" in the header to "date of last revision" and "date" in the revision log header to "revision date." Replaced investigational verbiage with "evidence is limited in supporting safety and efficacy." Added CPT 64580 and 64590 and HCPCS L8680, L8682, L8683, L8695, and L8696.	12/21	12/21

References

1. National coverage determination: phrenic nerve stimulator (160.19). Centers for Medicare and Medicaid Services Web site. http://www.cms.hhs.gov/mcd/search.asp. Accessed November 22, 2021.
2. Marion DW. Pacing the diaphragm: patient selection, evaluation, implantation and complications. UpToDate. www.uptodate.com. Published April 2, 2020. Accessed November 22, 2021.
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
4. Onders RP, Elmo M, Khansarinia S, et al. Complete worldwide operative experience in laparoscopic diaphragm pacing: results and differences in spinal cord injured patients and

amyotrophic lateral sclerosis patients. Surg Endosc. 2009;23(7):1433-1440. doi:10.1007/s00464-008-0223-3
5. McDermott CJ, Bradburn MJ, Maguire C, et al. DiPALS: Diaphragm Pacing in patients with Amyotrophic Lateral Sclerosis - a randomised controlled trial. *Health Technol Assess*. 2016;20(45):1-186. doi:10.3310/hta20450
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 23, 2021.
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 23, 2021.
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. https://www.accessdata.fda.gov/scripts/cdrh/cfdocs/cfpma/pma_template.cfm?id=p860026. Published 2019. Accessed November 23, 2021.
9. Phrenic nerve stimulation (remedē System) for central sleep apnea. Hayes. www.hayes.com. Published August 21, 2021. Accessed November 23, 2021.

Important Reminder

This clinical policy has been developed by appropriately experienced and licensed health care professionals based on a review and consideration of currently available generally accepted standards of medical practice; peer-reviewed medical literature; government agency/program approval status; evidence-based guidelines and positions of leading national health professional organizations; views of physicians practicing in relevant clinical areas affected by this clinical policy; and other available clinical information. The Health Plan makes no representations and accepts no liability with respect to the content of any external information used or relied upon in developing this clinical policy. This clinical policy is consistent with standards of medical practice current at the time that this clinical policy was approved. "Health Plan" means a health plan that has adopted this clinical policy and that is operated or administered, in whole or in part, by Centene Management Company, LLC, or any of such health plan's affiliates, as applicable.

The purpose of this clinical policy is to provide a guide to medical necessity, which is a component of the guidelines used to assist in making coverage decisions and administering benefits. It does not constitute a contract or guarantee regarding payment or results. Coverage decisions and the administration of benefits are subject to all terms, conditions, exclusions and limitations of the coverage documents (e.g., evidence of coverage, certificate of coverage, policy, contract of insurance, etc.), as well as to state and federal requirements and applicable Health Plan-level administrative policies and procedures.

This clinical policy is effective as of the date determined by the Health Plan. The date of posting may not be the effective date of this clinical policy. This clinical policy may be subject to

applicable legal and regulatory requirements relating to provider notification. If there is a discrepancy between the effective date of this clinical policy and any applicable legal or regulatory requirement, the requirements of law and regulation shall govern. The Health Plan retains the right to change, amend or withdraw this clinical policy, and additional clinical policies may be developed and adopted as needed, at any time.

This clinical policy does not constitute medical advice, medical treatment or medical care. It is not intended to dictate to providers how to practice medicine. Providers are expected to exercise professional medical judgment in providing the most appropriate care, and are solely responsible for the medical advice and treatment of member/enrollees. This clinical policy is not intended to recommend treatment for member/enrollees. Member/enrollees should consult with their treating physician in connection with diagnosis and treatment decisions.

Providers referred to in this clinical policy are independent contractors who exercise independent judgment and over whom the Health Plan has no control or right of control. Providers are not agents or employees of the Health Plan.

This clinical policy is the property of the Health Plan. Unauthorized copying, use, and distribution of this clinical policy or any information contained herein are strictly prohibited. Providers, member/enrollees and their representatives are bound to the terms and conditions expressed herein through the terms of their contracts. Where no such contract exists, providers, member/enrollees and their representatives agree to be bound by such terms and conditions by providing services to member/enrollees and/or submitting claims for payment for such services.

Note: For Medicaid members/enrollees, when state Medicaid coverage provisions conflict with the coverage provisions in this clinical policy, state Medicaid coverage provisions take precedence. Please refer to the state Medicaid manual for any coverage provisions pertaining to this clinical policy.

Note: For Medicare members/enrollees, to ensure consistency with the Medicare National Coverage Determinations (NCD) and Local Coverage Determinations (LCD), all applicable NCDs, LCDs, and Medicare Coverage Articles should be reviewed prior to applying the criteria set forth in this clinical policy. Refer to the CMS website at http://www.cms.gov for additional information.