Clinical Policy: Diaphragmatic/Phrenic Nerve Stimulation

Reference Number: WNC.CP.156 Coding Implications
Last Review Date: Revision Log

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

Note: 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.

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 WellCare of North Carolina® that diaphragmatic/phrenic nerve stimulation with the *Mark IV™ Breathing Pacemaker System or the Spirit Diaphragm Pacing Transmitter* 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 WellCare of North Carolina® that diaphragmatic/phrenic nerve stimulation with the *NeuRX RA/4* Diaphragm Pacing 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 Forced Vital Capacity (FVC)) less than 45% predicted;
c. Diaphragm movement with stimulation is visible under fluoroscopy or by other radiographic techniques such as ultrasound;;
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 or by other radiographic techniques such as ultrasound;;
   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 WellCare of North Carolina® 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 RA/4 Diaphragm Pacing System® (Synapse Biomedical, Inc.)
United States Food and Drug Administration (FDA) approval for distribution of the NeuRx DPS® (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.[1]

In 2011 the FDA approved the NeuRx RA/4 Diaphragm Pacing 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".[2(p.1)]

Avery Diaphragm Pacing System (Avery Biomedical Device, Inc.)
The *Avery Diaphragm Pacing System* includes receivers and electrodes that are surgically implanted and includes an external transmitter worn over the implanted receivers.3 The different types of Avery systems include the Mark IV Breathing Pacemaker System and the Spirit Diaphragm Pacing System.3 The Mark IV Breathing Pacemaker System is a diaphragmatic/phrenic stimulator system approved for use by the FDA in the United States. 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".[4] In 2019, the Spirit Diaphragm Pacing Transmitter received full FDA approval for the use of this system for patients who have functional lungs and diaphragm muscle and who have an intact phrenic nerve.[3,5,6]

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 2024, 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	Open implantation of neurostimulator electrode array; peripheral nerve, (excludes sacral nerve)
64580	Open implantation of neurostimulator electrode array; neuromuscular
64590	Insertion or replacement of peripheral, sacral, or gastric neurostimulator pulse generator or receiver, requiring pocket creation and connection between electrode array and pulse generator or receiver
64595	Revision or removal of peripheral, sacral, or gastric neurostimulator pulse generator or receiver, with detachable connection to electrode array.

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
L8683	Radiofrequency transmitter (external) for use with implantable neurostimulator radiofrequency receiver

HCPCS ®* Codes	Description
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

Reviews, Revisions, and Approvals	Reviewed Date	Approval Date
Original approval date	03/21	05/21
Reviewed CPT, HCPCS, and ICD-10-CM codes.	10/21	11/21
Annual Review. References reviewed, updated and reformatted. Replaced investigational verbiage, section III, with "evidence is limited in supporting safety and efficacy." Added CPT 64580 and 64590. Added HCPCS L8680 L8682 L8683 L8695 L8696	09/22	11/22
Annual review. Criteria II.A.1.c. and Criteria II.A.2.b. updated to include "or by other radiographic techniques such as ultrasound" in addition to fluoroscopy. Background updated to include U.S. Food and Drug Administration premarket approval information regarding the Avery Spirit Diaphragm Pacing Transmitter and deleted " The pacemaker is classified as a Class III neurologic therapeutic device requiring premarket approval (PMA)." ICD-10 codes removed. References reviewed and updated.	02/23	02/23
NCHC verbiage removed from NC Guidance Verbiage.	04/23	04/23
Annual review. Product name updates in criteria II. and in background with no clinical significance. References reviewed and updated.	11/23	11/23
Annual review. Criteria I. updated to include the Spirit Diaphragm Pacing Transmitter. Background updated to include information regarding full FDA approval of the Spirit Diaphragm Pacing Transmitter. Descriptions updated for 64590 64595 l8682 l8683. References reviewed and updated.		

References

1. U.S. Food and Drug Administration. Premarket Notification Database: NeuRx DPSTM 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 June 18, 2024.
2. U.S. Food and Drug Administration. Premarket Approvals for the NeuRx DPSTM 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 June 18, 2024.

3. Avery Biomedical Devices. System Information. https://averybiomedical.com/systems-information/. Accessed June 18, 2024.
4. U.S. Food and Drug Administration. Diaphragmatic Pacemaker Phrenic Nerve Stimulator - P860026. https://www.accessdata.fda.gov/scripts/cdrh/cfdocs/cfpma/pma_template.cfm?id=p860026. Published February 25, 1987. Accessed June 20, 2024.
5. U.S. Food and Drug Administration. Avery Diaphragm Pacing System - P860026. https://www.accessdata.fda.gov/scripts/cdrh/cfdocs/cfpma/pma.cfm?id=P860026S009. Published November 04, 2019. Accessed June 20, 2024.
6. Avery Biomedical Devices. https://averybiomedical.com/spirit-earns-fda-approval/. Published November 18, 2019. Accessed June 18, 2024.
7. 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 June 18, 2024.
8. Marion DW. Pacing the diaphragm: Patient selection, evaluation, implantation and complications. UpToDate. www.uptodate.com. Published November 07, 2023. Accessed June 21, 2024.
9. Health Technology Assessment. Phrenic nerve stimulation (remedē System) for central sleep apnea. Hayes. www.hayesinc.com. Published April 22, 2022 (annual review May 08, 2024). Accessed June 21, 2024.
10. 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 to S386. doi:10.21037/jtd.2016.03.97
11. 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 to 1440. doi:10.1007/s00464-008-0223-3
12. U.S. Food and Drug Administration. Approval for NeuRx Diaphragm Pacing System – P200018. https://www.fda.gov/medical-devices/recently-approved-devices/neurx-diaphragm-pacing-system-p200018. Published March 31, 2023. Accessed June 21, 2024.
13. 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 to 186. doi:10.3310/hta20450

North Carolina Guidance

Eligibility Requirements

a. An eligible beneficiary shall be enrolled in the NC Medicaid Program (Medicaid is NC Medicaid program, unless context clearly indicates otherwise);
b. Provider(s) shall verify each Medicaid beneficiary's eligibility each time a service is rendered.
c. The Medicaid beneficiary may have service restrictions due to their eligibility category that would make them ineligible for this service.

EPSDT Special Provision: Exception to Policy Limitations for a Medicaid Beneficiary under 21 Years of Age

a. 42 U.S.C. § 1396d(r) [1905(r) of the Social Security Act]

Early and Periodic Screening, Diagnostic, and Treatment (EPSDT) is a federal Medicaid requirement that requires the state Medicaid agency to cover services, products, or procedures for Medicaid beneficiary under 21 years of age if the service is medically necessary health care to correct or ameliorate a defect, physical or mental illness, or a condition [health problem] identified through a screening examination (includes any evaluation by a physician or other licensed practitioner).

This means EPSDT covers most of the medical or remedial care a child needs to improve or maintain his or her health in the best condition possible, compensate for a health problem, prevent it from worsening, or prevent the development of additional health problems.

Medically necessary services will be provided in the most economic mode, as long as the treatment made available is similarly efficacious to the service requested by the beneficiary's physician, therapist, or other licensed practitioner; the determination process does not delay the delivery of the needed service; and the determination does not limit the beneficiary's right to a free choice of providers.

EPSDT does not require the state Medicaid agency to provide any service, product or procedure:

1. that is unsafe, ineffective, or experimental or investigational.
2. that is not medical in nature or not generally recognized as an accepted method of medical practice or treatment.

Service limitations on scope, amount, duration, frequency, location of service, and other specific criteria described in clinical coverage policies may be exceeded or may not apply as long as the provider's documentation shows that the requested service is medically necessary "to correct or ameliorate a defect, physical or mental illness, or a condition" [health problem]; that is, provider documentation shows how the service, product, or procedure meets all EPSDT criteria, including to correct or improve or maintain the beneficiary's health in the best condition possible, compensate for a health problem, prevent it from worsening, or prevent the development of additional health problems.

EPSDT and Prior Approval Requirements

1. If the service, product, or procedure requires prior approval, the fact that the beneficiary is under 21 years of age does NOT eliminate the requirement for prior approval.
2. **IMPORTANT ADDITIONAL INFORMATION** about EPSDT and prior approval is found in the *NCTracks Provider Claims and Billing Assistance Guide*, and on the EPSDT provider page. The Web addresses are specified below:

NCTracks Provider Claims and Billing Assistance Guide:
https://www.nctracks.nc.gov/content/public/providers/provider-manuals.html
EPSDT provider page: https://medicaid.ncdhhs.gov/

Provider(s) Eligible to Bill for the Procedure, Product, or Service

To be eligible to bill for the procedure, product, or service related to this policy, the provider(s) shall:

a. meet Medicaid qualifications for participation;
b. have a current and signed Department of Health and Human Services (DHHS) Provider Administrative Participation Agreement; and
c. bill only for procedures, products, and services that are within the scope of their clinical practice, as defined by the appropriate licensing entity.

Compliance

Provider(s) shall comply with the following in effect at the time the service is rendered:

a. All applicable agreements, federal, state and local laws and regulations including the Health Insurance Portability and Accountability Act (HIPAA) and record retention requirements; and
b. All NC Medicaid's clinical (medical) coverage policies, guidelines, policies, provider manuals, implementation updates, and bulletins published by the Centers for Medicare and Medicaid Services (CMS), DHHS, DHHS division(s) or fiscal contractor(s).

Claims-Related Information

Provider(s) shall comply with the NC Tracks Provider Claims and Billing Assistance Guide, Medicaid bulletins, fee schedules, NC Medicaid's clinical coverage policies and any other relevant documents for specific coverage and reimbursement for Medicaid:

a. Claim Type - as applicable to the service provided:
   Professional (CMS-1500/837P transaction)
   Institutional (UB-04/837I transaction)
   Unless directed otherwise, Institutional Claims must be billed according to the National Uniform Billing Guidelines. All claims must comply with National Coding Guidelines.
b. International Classification of Diseases and Related Health Problems, Tenth Revisions, Clinical Modification (ICD-10-CM) and Procedural Coding System (PCS) - Provider(s) shall report the ICD-10-CM and Procedural Coding System (PCS) to the highest level of specificity that supports medical necessity. Provider(s) shall use the current ICD-10 edition and any subsequent editions in effect at the time of service. Provider(s) shall refer to the applicable edition for code description, as it is no longer documented in the policy.
c. Code(s) - Provider(s) shall report the most specific billing code that accurately and completely describes the procedure, product or service provided. Provider(s) shall use the Current Procedural Terminology (CPT), Health Care Procedure Coding System (HCPCS), and UB-04 Data Specifications Manual (for a complete listing of valid revenue codes) and any subsequent editions in effect at the time of service. Provider(s) shall refer to the applicable edition for the code description, as it is no longer documented in the policy. If no such specific CPT or HCPCS code exists, then the provider(s) shall report the procedure, product or service using the appropriate unlisted procedure or service code.
   Unlisted Procedure or Service
   CPT: The provider(s) shall refer to and comply with the Instructions for Use of the CPT Codebook, Unlisted Procedure or Service, and Special Report as documented in the current CPT in effect at the time of service.

HCPCS: The provider(s) shall refer to and comply with the Instructions For Use of HCPCS National Level II codes, Unlisted Procedure or Service and Special Report as documented in the current HCPCS edition in effect at the time of service

d. Modifiers - Providers shall follow applicable modifier guidelines.
e. Billing Units - Provider(s) shall report the appropriate code(s) used which determines the billing unit(s).
f. Co-payments -
   For Medicaid refer to Medicaid State Plan:
   https://medicaid.ncdhhs.gov/get-involved/nc-health-choice-state-plan

g. Reimbursement - Provider(s) shall bill their usual and customary charges. For a schedule of rates, refer to: https://medicaid.ncdhhs.gov/.

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 members/enrollees. This clinical policy is not intended to recommend treatment for members/enrollees. Members/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, members/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, members/enrollees and their representatives agree to be bound by such terms and conditions by providing services to members/enrollees and/or submitting claims for payment for such services.