Diaphragmatic/Phrenic Nerve Stimulation and Diaphragm Pacing Systems

Policy # 00032
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Applies to all products administered or underwritten by Blue Cross and Blue Shield of Louisiana and its subsidiary, HMO Louisiana, Inc. (collectively referred to as the "Company"), unless otherwise provided in the applicable contract. Medical technology is constantly evolving, and we reserve the right to review and update Medical Policy periodically.

When Services May Be Eligible for Coverage

Coverage for eligible medical treatments or procedures, drugs, devices or biological products may be provided only if:

- *Benefits are available in the member's contract/certificate, and*
- *Medical necessity criteria and guidelines are met.*

Diaphragmatic/Phrenic Nerve Stimulation

Based on review of available data, the Company may consider the use of diaphragmatic/phrenic nerve stimulation with an FDA-approved device as an alternative to invasive mechanical ventilation for individuals who are 18 years of age or older when ALL of the following criteria are met to be **eligible for coverage**:**

Patient Selection Criteria

- The individual has ventilatory failure from stable, high spinal cord injury or ventilatory failure from central alveolar hypoventilation syndrome; **and**
- The individual cannot breathe spontaneously for 4 continuous hours or more without use of a mechanical ventilator; **and**
- Diaphragm movement with stimulation is visible under fluoroscopy; **and**
- 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 for at least 4 continuous hours a day; **and**

- Individual 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; **and**
- Bilateral clinically acceptable phrenic nerve function is demonstrated with electromyography recordings and nerve conduction times.

Diaphragmatic Stimulation
Based on review of available data, the Company may consider diaphragm stimulation with an FDA approved diaphragm pacing system as an alternative to invasive mechanical ventilation in individuals who are 18 years of age or older when ALL of the following criteria are met to be **eligible for coverage**:**

Patient Selection Criteria

- The individual has ventilatory failure from stable, high spinal cord injury or ventilatory failure from central alveolar hypoventilation syndrome or ventilatory failure from motor neuron disease, for example amyotrophic lateral sclerosis; **and**
- The individual cannot breathe spontaneously for 4 continuous hours or more without use of a mechanical ventilator; **and**
- 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 for at least 4 continuous hours a day; **and**
- Individual 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.

When Services Are Considered Not Medically Necessary

The use of diaphragmatic/phrenic nerve stimulation devices and Diaphragm Pacing Systems are considered to be **not medically necessary**** for any of the following conditions:

- The individual can breathe spontaneously for 4 continuous hours or more without use of a mechanical ventilator; **or**

- The respiratory insufficiency is temporary.

When Services Are Considered Investigational

Note: Coverage is not available for investigational medical treatments or procedures, drugs, devices or biological products.

Based on review of available data, the Company considers use of diaphragmatic/phrenic nerve stimulation and Diaphragm Pacing Systems for all other indications including, but not limited to the following to be **investigational*:**

- Underlying cardiac, pulmonary or chest wall disease is present which is significant enough to prevent spontaneous breathing off a ventilator for more than 4 hours even with the use of the phrenic nerve or diaphragm pacemaker device; **or**
- For treatment of any other condition where the phrenic nerve and diaphragm are intact (for example, chronic obstructive lung disease, restrictive lung disease, singultus [hiccups]), central sleep apnea); **or**
- For adolescents, children and infants; **or**
- When the patient selection criteria are not met.

Background/Overview

The electrophrenic pacemaker is an implanted electrode and receiver with a pocket or table-top size external transmitter. The device electrically stimulates the phrenic nerves to contract the diaphragm rhythmically, which causes breathing. Diaphragmatic/Phrenic (D/P) nerve stimulation is intended as an alternative to mechanical ventilation in selected patients with ventilatory insufficiency or failure that 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 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 patients who may be ambulatory or use a wheelchair. It also allows patients to speak and enhances their social integration.

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The NeuRx DPS RA/4 Respiratory Stimulation System is implanted through minimally invasive laparoscopic surgery and provides electrical stimulation to muscles and nerves that run through the diaphragm. This eliminates any direct contact with the phrenic nerve, allows all circuitry and electronics to remain outside the body, and provides direct, selective activation to each hemidiaphragm. According to manufacturer information, when stimulated by the NeuRx DPS, the diaphragm contracts, mimicking natural breathing and allowing air to fill the upper and lower parts of the lungs, rather than forcing air in with a mechanical ventilator. The device uses four electrodes implanted in the muscle of the diaphragm to electronically stimulate contraction; this stimulation allows the user to inhale. The DPS is lightweight and battery powered, eliminating the need for an external power source. Similar to the NeuRx DPS system, the Mark IV system is connected to the phrenic nerve by electrodes in the neck or chest area. The device consists of a surgically implanted receiver and electrodes which are connected to an external transmitter for transmitting the stimulating pulses across the skin to the implanted receiver.

The Remedē®‡ System was approved by the FDA on October 6, 2017 for the treatment of moderate to severe central sleep apnea in adult individuals. The manufacturer describes the device as:

An implantable pacemaker-like device that was designed for improving central sleep apnea (CSA) using Respidrive™‡, a Respiratory Rhythm Management™‡ algorithm. The Remedē system delivers electrical pulses via a proprietary, novel transvenous implantable lead to one of the body's two phrenic nerves. The Remedē system therapy is intended to stimulate the diaphragm to restore a more natural, less disrupted, breathing pattern.

FDA or Other Governmental Regulatory Approval

U.S. Food and Drug Administration (FDA)

On October 6, 2017, the U.S. FDA approved the Remedē System (Respicardia). This device is an implantable phrenic nerve stimulator indicated for the treatment of moderate to severe central sleep apnea (CSA) in adult patients.

On September 9, 2011, the U.S. FDA approved NeuRx DPS, Diaphragm Pacing System (Synapse Biomedical). This device is indicated for use in amyotrophic lateral sclerosis (ALS) patients with a

stimulatable diaphragm (both right and left portions) as demonstrated by voluntary contraction on phrenic nerve conduction studies, and who are experiencing chronic hypoventilation, but not progressed to forced vital capacity (FVC) less than 45% predicted. Approved for use only in patients 21 years of age or older.

On January 5, 1987, the U.S. FDA approved Diaphragmatic Pacemaker Phrenic Nerve Stimulator (Avery Biomedical Devices). The device would be marketed under the trade name mark IV and is indicated for persons who require chronic ventilator support because of upper motor neuron respiratory muscle paralysis or because of central alveolar hypoventilation and whose remaining phrenic nerve, lung, and diaphragm functions is sufficient to accommodate electrical stimulation.

Centers for Medicare and Medicaid Services (CMS)
The implantation of a phrenic nerve stimulator is covered for selected patients with partial or complete respiratory insufficiency. National Coverage Determination (NCD) for Phrenic Nerve Stimulator (160.19) pertains to phrenic nerve stimulation.

Rationale/Source

This medical policy was developed through consideration of peer-reviewed medical literature generally recognized by the relevant medical community, U.S. Food and Drug Administration approval status, nationally accepted standards of medical practice and accepted standards of medical practice in this community, technology evaluation centers, reference to federal regulations, other plan medical policies, and accredited national guidelines.

Based on humanitarian device exemptions, diaphragmatic/phrenic nerve stimulation is considered eligible for coverage for patients with high spinal cord injuries to allow freedom from mechanical ventilation for at least 4 hours daily and is indicated for patients with ALS to delay the need for mechanical ventilation.

Central sleep apnea (CSA) is characterized by sleep-disordered breathing due to diminished or absent respiratory effort. CSA may be idiopathic or secondary (associated with a medical condition, drugs, or high altitude breathing). The use of positive airway pressure devices is currently the most

common form of therapy for CSA. An implantable device that stimulates the phrenic nerve in the chest is a potential alternative treatment. The battery-powered device sends signals to the diaphragm in order to stimulate breathing and normalize sleep-related breathing patterns.

For individuals with CSA who receive phrenic nerve stimulation, the evidence includes 1 randomized controlled trial (RCT) and observational studies. Relevant outcomes are change in disease status, functional outcomes, and quality of life. The RCT compared the use of phrenic nerve stimulation to no treatment among patients with CSA of various etiologies. All patients received implantation of the phrenic nerve stimulation system, with activation of the system after 1 month in the intervention group and activation after 6 months in the control group. Activation is delayed 1 month after implantation to allow for 4 lead healing. At 6 months follow-up, the patients with the activated device experienced significant improvements in several sleep metrics and quality of life measures. At 12 months follow-up, patients in the activated device arm showed sustained significant improvements from baseline in sleep metrics and quality of life. A subgroup analysis of patients with heart failure combined 6- and 12-month data from patients in the intervention group and 12- and 18-month data from the control group. Results from this subgroup analysis showed significant improvements in sleep metrics and quality of life at 12 months compared with baseline. Results from observational studies supported the results of the RCT. An invasive procedure would typically be considered only if non-surgical treatments had failed, but there is limited data in which phrenic nerve stimulation was evaluated in patients who had failed the current standard of care, positive airway pressure, or respiratory stimulant medication. The evidence is insufficient to determine that the technology results in an improvement in the net health outcome.

References

1. Blue Cross and Blue Shield of Massachusetts. “Diaphragmatic/Phrenic Nerve Stimulation and Diaphragm Pacing Systems. 593, 06:2022.
2. Blue Cross and Blue Shield of Massachusetts.” Phrenic Nerve Stimulation for Central Sleep Apnea” 955, 01:2024.

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Policy History

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05/07/2004	Medical Director review
05/18/2004	Medical Policy Committee review
06/28/2004	Managed Care Advisory Council approval
03/01/2005	Medical Director review
03/15/2005	Medical Policy Committee review
04/04/2005	Managed Care Advisory Council approval
03/14/2007	Medical Director review
03/21/2007	Medical Policy Committee approval. Coverage eligibility unchanged.
03/12/2008	Medical Director review
03/19/2008	Medical Policy Committee approval. No change to coverage eligibility.
03/04/2009	Medical Director review
03/18/2009	Medical Policy Committee approval. No change to coverage eligibility.
06/03/2010	Medical Policy Committee review
06/16/2010	Medical Policy Implementation Committee approval. No change to coverage. eligibility.
05/05/2011	Medical Policy Committee review
05/18/2011	Medical Policy Implementation Committee approval. No change to coverage. eligibility.
05/03/2012	Medical Policy Committee review. Recommend archiving policy.
05/16/2012	Medical Policy Implementation Committee approval. Archived medical policy.
03/07/2019	Medical Policy Committee review.
03/20/2019	Medical Policy Implementation Committee approval. Brought back to active status. Title and coverage changed.
03/05/2020	Medical Policy Committee review.
03/11/2020	Medical Policy Implementation Committee approval. No change to coverage.
03/04/2021	Medical Policy Committee review.
03/10/2021	Medical Policy Implementation Committee approval. No change to coverage.
03/03/2022	Medical Policy Committee review.

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03/09/2022 Medical Policy Implementation Committee approval. No change to coverage.
03/02/2023 Medical Policy Committee review.
03/08/2023 Medical Policy Implementation Committee approval. No change to coverage.
12/13/2023 Coding update
03/07/2024 Medical Policy Committee review.
03/13/2024 Medical Policy Implementation Committee approval. No change to coverage.
Next Scheduled Review Date: 03/2025

Coding

The five character codes included in the Blue Cross Blue Shield of Louisiana Medical Policy Coverage Guidelines are obtained from Current Procedural Terminology (CPT®)‡, copyright 2023 by the American Medical Association (AMA). CPT is developed by the AMA as a listing of descriptive terms and five character identifying codes and modifiers for reporting medical services and procedures performed by physician.

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Codes used to identify services associated with this policy may include (but may not be limited to) the following:

Code Type	Code
CPT	33276, 33277, 33278, 33279, 33280, 33281, 33287, 33288, 64575, 64585, 64590, 64595, 93150, 93151, 93152, 93153, 95972
HCPCS	C1816, C1883, L8680, L8681
ICD-10 Diagnosis	All related diagnoses

*Investigational – A medical treatment, procedure, drug, device, or biological product is Investigational if the effectiveness has not been clearly tested and it has not been incorporated into standard medical practice. Any determination we make that a medical treatment, procedure, drug, device, or biological product is Investigational will be based on a consideration of the following:

A. Whether the medical treatment, procedure, drug, device, or biological product can be lawfully marketed without approval of the U.S. Food and Drug Administration (FDA) and whether such approval has been granted at the time the medical treatment, procedure, drug, device, or biological product is sought to be furnished; or
B. Whether the medical treatment, procedure, drug, device, or biological product requires further studies or clinical trials to determine its maximum tolerated dose, toxicity, safety, effectiveness, or effectiveness as compared with the standard means of treatment or diagnosis, must improve health outcomes, according to the consensus of opinion among experts as shown by reliable evidence, including:
    1. Consultation with technology evaluation center(s);
    2. Credible scientific evidence published in peer-reviewed medical literature generally recognized by the relevant medical community; or
    3. Reference to federal regulations.

**Medically Necessary (or "Medical Necessity") - Health care services, treatment, procedures, equipment, drugs, devices, items or supplies that a Provider, exercising prudent clinical judgment, would provide to a patient for the purpose of preventing, evaluating, diagnosing or treating an illness, injury, disease or its symptoms, and that are:

A. In accordance with nationally accepted standards of medical practice;

B. Clinically appropriate, in terms of type, frequency, extent, level of care, site and duration, and considered effective for the patient's illness, injury or disease; and
C. Not primarily for the personal comfort or convenience of the patient, physician or other health care provider, and not more costly than an alternative service or sequence of services at least as likely to produce equivalent therapeutic or diagnostic results as to the diagnosis or treatment of that patient's illness, injury or disease.

For these purposes, "nationally accepted standards of medical practice" means standards that are based on credible scientific evidence published in peer-reviewed medical literature generally recognized by the relevant medical community, Physician Specialty Society recommendations and the views of Physicians practicing in relevant clinical areas and any other relevant factors.

‡ Indicated trademarks are the registered trademarks of their respective owners.

NOTICE: If the Patient's health insurance contract contains language that differs from the BCBSLA Medical Policy definition noted above, the definition in the health insurance contract will be relied upon for specific coverage determinations.

NOTICE: Medical Policies are scientific based opinions, provided solely for coverage and informational purposes. Medical Policies should not be construed to suggest that the Company recommends, advocates, requires, encourages, or discourages any particular treatment, procedure, or service, or any particular course of treatment, procedure, or service.

NOTICE: Federal and State law, as well as contract language, including definitions and specific contract provisions/exclusions, take precedence over Medical Policy and must be considered first in determining eligibility for coverage.