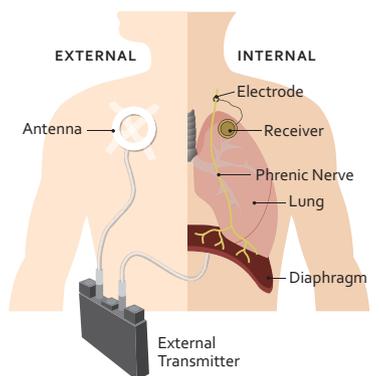


Diaphragm Pacing by Phrenic Nerve Stimulation

What is diaphragm pacing?

Diaphragm pacing is a way to help support people who cannot breathe on their own. It can be used in place of a mechanical ventilator at times. It is a treatment option for some people diagnosed with congenital central hypoventilation syndrome (CCHS) as well as those who have suffered a high cervical spinal cord injury.



The diaphragms are large muscles found under each lung that are the major muscles used in breathing. The phrenic nerves send a signal to the diaphragms stimulating them to breathe. People who have problems with the brain or spinal cord at times do not send the signals well to breathe. Diaphragm pacing can use the phrenic nerves to send the signals to a person's diaphragm muscles to contract and take a breath in. Often management of diaphragm pacers is done by special medical care centers, some of whom have had experience with this for decades.

How does it work?

Diaphragm pacing uses the person's own diaphragms as the "ventilator." A diaphragm pacer system involves 4 components:

- electrodes that are surgically attached to the phrenic nerves on each side of the neck or in the chest,
- receivers that are surgically implanted under the skin in the abdomen or chest,
- antennae which are taped on the chest over the receivers during pacing,
- portable external transmitter machine.

When turned on, the transmitter sends a signal to the receivers through the antennae. The external transmitter generates electrical energy similar to radio wave signals. The receiver converts this energy signal to electrical current that is conducted to the phrenic nerves. The nerve stimulation causes the diaphragm muscles to contract and the person takes a breath in. This cycle repeats for the number of breaths needed each minute. A respiratory rate is set for the transmitter for how often to trigger a breath.

What are the advantages of diaphragm pacing?

Diaphragm pacing is an attractive treatment option for two reasons. First, it allows for the possibility of removal of the tracheostomy in people who require mechanical ventilator support only during sleep. Second, for people who need support full time (both day and night), it permits some freedom from the ventilator that may allow easier speech or participation in certain types of activities.

Who is a candidate for diaphragm pacing?

For a person to be considered for diaphragm pacing, one must be sure that:

- there is normal diaphragm muscle function,
- the phrenic nerves are intact and able to send a signal when stimulated,
- The person has relatively mild or no lung disease. This system does not work well if the lungs don't work well.

Testing can be done to check whether the phrenic nerves and diaphragms work normally. A lung specialist will evaluate the person to see how healthy his or her lungs are and whether this is an option to consider. People who are very obese may not be good candidates for pacing. There may be too much fat tissue between the antenna and receiver that can limit getting a good signal to the phrenic nerves. With obesity, it may not be possible to find a consistent diaphragm pacer setting to achieve adequate ventilation.

Diaphragm pacing has also been successful in patients with cardiac pacemakers.

What happens during surgery?

Surgery is done with general anesthesia so a person is asleep for the whole procedure. This surgery is

delicate and usually is performed in specialized centers experienced with diaphragm pacing. Both the phrenic nerve electrodes and diaphragm pacer receivers are put in place. Usually these are placed on both the right and left side with small incisions (thoracoscopic surgery). Phrenic nerve electrodes are attached to the nerves in the lower sides of the chest or upper chest near the neck. The phrenic nerve electrodes are connected inside by lead wires to the receivers, which are implanted on each side just under the skin, either on the upper abdomen or in the upper chest. The diaphragm pacer equipment is tested during the surgery to be sure that it is functional.

When is diaphragm pacing started?

Diaphragm pacing is not used right after surgery. It is best to wait for the incisions to heal, and for scar tissue formation around the nerve electrodes to stabilize them. Diaphragm pacing is generally started about 6-8 weeks after surgery. Most of the time, it is first turned on with observation in the hospital. Often, pacing can only be done for 1-1.5 hours per night at first before the diaphragms get tired (fatigue). The time on pacers is gradually increased. This gradual process is necessary even in people who breathe on their own while awake.

It is thought that the diaphragm signals that the phrenic nerves send with the pacer is different from the natural phrenic nerve impulses. This takes some getting used to. So, it is necessary to train the diaphragm to accept longer periods of pacing without fatigue. The time pacing is gradually increased and usually, patients are pacing up to 8-12 hours at a time by about 3 months. This is the usual maximum time pacers are used. Twenty- four hour diaphragm pacing is not recommended because of diaphragm fatigue. Therefore, for people who depend on full time assisted ventilation, there is a need to have another method of ventilatory support such as home mechanical ventilation by tracheostomy or noninvasive positive pressure ventilation for use when not pacing. Your specialist can help you decide how much time you could use pacers and whether they are the right choice to help support your breathing.

People using diaphragm pacers should have a pulse oximeter to check their oxygen saturation levels (see ATS Patient Information Series "Pulse Oximetry"). Ideally, one should also have a machine to measure the carbon dioxide (CO₂) level, which is what we breathe out to rid the body of extra acid. This can be done using an exhaled CO₂ monitor. This equipment requires prescription and insurance approvals that will need to be done when planning for pacer placement.

What are the risks of diaphragm pacing?

Diaphragm pacing is generally very well tolerated. The

main risks of diaphragm pacing include risk of injury to the phrenic nerve during surgery, infection of implanted components, and failure of the equipment. Changing body positions, which can compress or increase the distance between the antenna and receiver, can result in decreased or increased diaphragm contractions. It is important to get the right settings for the position that the person will use the pacers. If a person has a respiratory infection, the settings may need to be adjusted during the illness or at times another form of ventilator support used.

Obstructive sleep apnea may occur in those who are diaphragm pacing without tracheostomy. Because a diaphragm pacer breath is generated by direct stimulation of the phrenic nerve, there is no synchronous contraction of upper airway skeletal muscles with each inspiration. This can result in upper airway collapse and obstructive apnea. Usually, this can be improved by adjusting the diaphragm pacer setting.

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Adapted from the ATS Clinical Statement on Congenital Central Hypoventilation Syndrome. <http://www.thoracic.org/newsroom/press-releases/resources/cchs-statement.pdf>

Rx Action Steps

- ✓ Talk to your lung specialist to see if you are a candidate for diaphragm pacing.
- ✓ Discuss if tests need to be done to check on your phrenic nerve and diaphragm function.
- ✓ Ask to be referred to a center specializing in diaphragm pacing if you are a possible candidate.

Healthcare Provider's Contact Number:

Other Resources:

**Introduction to Diaphragmatic Pacing—
Children's Hospital of Los Angeles**

<https://www.youtube.com/watch?v=ZbhPbcd4yrl>

**Diaphragmatic Pacing- Trouble shooting Tips—
Children's Hospital of Los Angeles**

<https://www.youtube.com/watch?v=g174Yv2yUs>

ATS Patient Information Series

www.thoracic.org/patients

- Congenital Central Hypoventilation Syndrome (CCHS)
- Mechanical Ventilation
- Pulse Oximetry

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