

NICE interventional procedures consultation document, March 2024

## NATIONAL INSTITUTE FOR HEALTH AND CARE EXCELLENCE

### Interventional procedures consultation document

# Phrenic nerve pacing for congenital central hypoventilation syndrome

Congenital central hypoventilation syndrome (CCHS) is a rare genetic condition that affects the brain's ability to control breathing. People with CCHS may have difficulty breathing as well as low levels of oxygen and high levels of carbon dioxide in their blood (hypoventilation). Symptoms mostly occur during sleep, but people with severe CCHS can also have symptoms while awake. Most people with CCHS also need a mechanical ventilator to help them breathe. This may be needed all the time or only when sleeping.

In this procedure, an electrode is implanted around the phrenic nerve in the lower neck or chest. The phrenic nerve controls the diaphragm, which is the main muscle used for breathing. The electrode is connected to a receiver that is usually placed in the chest wall. An external transmitter then sends radiofrequency signals to the receiver, causing the electrode to stimulate (pacing) the phrenic nerve. The pacing makes the diaphragm contract. The aim is to help people breathe normally and to have some time without a ventilator, potentially improving their quality of life.

NICE is looking at phrenic nerve pacing for congenital central hypoventilation syndrome.

NICE's interventional procedures advisory committee met to consider the evidence and the opinions of professional experts with knowledge of the procedure.

This document contains the [draft guidance for consultation](#). Your views are welcome, particularly:

- comments on the draft recommendations
- information about factual inaccuracies
- additional relevant evidence, with references if possible.

NICE is committed to promoting equality of opportunity, eliminating unlawful discrimination and fostering good relations between people with particular protected characteristics and others.

**This is not NICE's final guidance on this procedure. The draft guidance may change after this consultation.**

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After consultation ends, the committee will:

- meet again to consider the consultation comments, review the evidence and make appropriate changes to the draft guidance
- prepare a second draft, which will go through a [resolution process](#) before the final guidance is agreed.

Please note that we reserve the right to summarise and edit comments received during consultation or not to publish them at all if, in the reasonable opinion of NICE, there are a lot of comments or if publishing the comments would be unlawful or otherwise inappropriate.

Closing date for comments: 23 April 2024

Target date for publication of guidance: August 2024

## 1 Draft recommendations

- 1.1 Use phrenic nerve pacing as an option to treat congenital central hypoventilation syndrome (CCHS) with standard arrangements in place for clinical governance, consent and audit.
- 1.2 For auditing the outcomes of this procedure, the main efficacy and safety outcomes identified in this guidance can be entered into [NICE's interventional procedure outcomes audit tool](#) (for use at local discretion).
- 1.3 Patient selection should be done by a multidisciplinary team experienced in managing the condition in specialist centres.
- 1.4 This procedure should only be done in specialist centres by clinicians with specific training and experience in the procedure. Patients should be followed up by clinicians experienced in managing the condition.

### Why the committee made these recommendations

Because CCHS is a rare genetic condition, there is limited evidence for the procedure in this population. But the available evidence shows benefits such as an increase in ventilator-free time and tracheostomy tube removal. The evidence also does not raise any major safety concerns. So, phrenic nerve pacing is recommended.

## 2 The condition, current treatments and procedure

### The condition

- 2.1 Congenital central hypoventilation syndrome (CCHS) is a rare genetic condition, with around 1,000 cases identified worldwide. CCHS affects how the autonomic nervous system manages or controls breathing. Normally, when breathing is shallow while

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asleep, the levels of carbon dioxide in the blood increase, which stimulates breathing. In CCHS, this stimulus does not happen, and breathing can stop. Common symptoms include difficulty breathing (especially during sleep), hypercapnia and hypoxemia. So, life-long ventilatory support is needed during sleep or all the time.

## **Current treatments**

2.2 There is no cure for CCHS, but the symptoms can be managed. As CCHS can affect several systems in the body, it needs to be managed by several medical teams (multidisciplinary approach). For respiratory insufficiency, the most common treatment includes positive pressure ventilation by tracheostomy or mask to assist with breathing.

## **The procedure**

2.3 Phrenic nerve pacing involves the direct stimulation of the phrenic nerve (PN) sending a signal to the diaphragm to contract, which produces the inhalation phase of breathing. It aims to provide ventilatory support for people with intact PNs and functioning diaphragm muscles.

2.4 The procedure is usually done by a thoracic approach (either an open thoracostomy or a thoracoscopic technique) and under general anaesthesia. Once the PN is identified and tested, an electrode is placed around the nerve in the chest, and then stabilised. The electrode is connected to a subcutaneous receiver usually placed in the chest wall. An external transmitter (powered by batteries) then sends radiofrequency signals to the device through an antenna which is worn over the receiver. The receiver translates radio waves into stimulating electrical pulses that are delivered to the PN by the electrode, to achieve diaphragm contraction and support breathing. The device is tested during and after the surgery to ensure that it works. This procedure is usually

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done bilaterally and can also be done unilaterally. A cervical approach can also be used and is done under general or local anaesthesia, but this is less common.

- 2.5 After implantation the person follows a diaphragm conditioning programme, which involves progressive use of the system for increasing periods of time with gradual weaning from the ventilator.

### **3 Committee considerations**

#### **The evidence**

- 3.1 NICE did a rapid review of the published literature on the efficacy and safety of this procedure. This comprised a comprehensive literature search and detailed review of the evidence from 5 sources, which was discussed by the committee. The evidence included 4 case series and 1 analysis of the Avery Biomedical Devices database. It is presented in the [summary of key evidence section in the interventional procedures overview](#). Other relevant literature is in the appendix of the overview.
- 3.2 The professional experts and the committee considered the key efficacy outcomes to be: quality of life, ventilator free hours per day, tracheostomy decannulation, survival, respiratory infections, and hospital admissions.
- 3.3 The professional experts and the committee considered the key safety outcomes to be: device failure, revision surgery, phrenic nerve palsy, and infections.
- 3.4 One patient organisation submission was received and discussed by the committee. Patient commentary was sought but none was received.

### **Committee comments**

- 3.5 The committee was pleased to hear from a patient organisation and its representative. It heard about the impact of CCHS on quality of life for people who are ventilator or mask dependent.
- 3.6 People who have this procedure should be followed up long term, with routine collection of safety and outcome data, technology failures and reoperation rates, possibly through a registry.
- 3.7 There are some people who have had phrenic nerve pacing for over 40 years, although replacement electrodes or receivers might be needed over time.
- 3.8 The committee was informed that having a backup mode of ventilation is essential, particularly in previously ventilator-dependent people.
- 3.9 The committee was informed that the condition varies in severity meaning there may be people with undetected daytime hypoventilation who are undiagnosed and may need pacing in adulthood.

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Chair, interventional procedures advisory committee

March 2024

ISBN: