

# Phrenic nerve pacing for congenital central hypoventilation syndrome

Interventional procedures guidance

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[www.nice.org.uk/guidance/ipg790](https://www.nice.org.uk/guidance/ipg790)

## Your responsibility

This guidance represents the view of NICE, arrived at after careful consideration of the evidence available. When exercising their judgement, healthcare professionals are expected to take this guidance fully into account, and specifically any special arrangements relating to the introduction of new interventional procedures. The guidance does not override the individual responsibility of healthcare professionals to make decisions appropriate to the circumstances of the individual patient, in consultation with the patient and/or guardian or carer.

All problems (adverse events) related to a medicine or medical device used for treatment or in a procedure should be reported to the Medicines and Healthcare products Regulatory Agency using the [Yellow Card Scheme](#).

Commissioners and/or providers have a responsibility to implement the guidance, in their local context, in light of their duties to have due regard to the need to eliminate unlawful

discrimination, advance equality of opportunity, and foster good relations. Nothing in this guidance should be interpreted in a way that would be inconsistent with compliance with those duties. Providers should ensure that governance structures are in place to review, authorise and monitor the introduction of new devices and procedures.

Commissioners and providers have a responsibility to promote an environmentally sustainable health and care system and should assess and reduce the environmental impact of implementing NICE recommendations wherever possible.

# 1 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 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 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 (a multidisciplinary approach). For respiratory insufficiency, the most common treatment includes positive pressure ventilation by tracheostomy or a mask, to assist with breathing.

### The procedure

- 2.3 Phrenic nerve pacing involves the direct stimulation of the phrenic nerve, which sends a signal to the diaphragm to contract, producing the inhalation phase of breathing. It aims to provide ventilatory support for people with intact phrenic nerves and functioning diaphragm muscles.
- 2.4 The procedure is usually done using a thoracic approach (often using a thoracoscopic technique; thoracotomy is rarely used) and under general anaesthesia. Once the phrenic nerve 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, which is powered by batteries, then sends radiofrequency signals to the device through an antenna that is worn over the receiver. The receiver translates radio waves into stimulating electrical pulses that are delivered to the phrenic nerve 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 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.

## 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 congenital central hypoventilation syndrome 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 a small number of people who have had phrenic nerve pacing for over 40 years, but 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 that there may be people with undetected daytime hypoventilation whose condition is undiagnosed and who may need pacing in adulthood.
- 3.10 The committee was informed that phrenic nerve pacing may be used in other forms of central hypoventilation.
- 3.11 The committee was informed that pacing can be used 24 hours a day if needed.

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## Endorsing organisation

This guidance has been endorsed by [Healthcare Improvement Scotland](#).