NHS Birmingham and Solihull CCG
NHS Sandwell and West Birmingham CCG

DRAFT
Policy for use of domiciliary Non-Invasive Ventilation.

Document Details:

Version:	DRAFT v2.
Ratified by (name and date of Committee):	Treatment Policy Clinical Development Group
Date issued for Public Engagement:	
Equality & Diversity Impact Assessment:	

The CCG policy has been reviewed and developed by the Treatment Policies Clinical Development Group in line with the groups guiding principles which are:

1. CCG Commissioners require clear evidence of clinical effectiveness before NHS resources are invested in the treatment;
2. CCG Commissioner require clear evidence of cost effectiveness before NHS resources are invested in the treatment;
3. The cost of the treatment for this patient and others within any anticipated cohort is a relevant factor;
4. CCG Commissioners will consider the extent to which the individual or patient group will gain a benefit from the treatment;
5. CCG Commissioners will balance the needs of each individual against the benefit which could be gained by alternative investment possibilities to meet the needs of the community
6. CCG Commissioners will consider all relevant national standards and take into account all proper and authoritative guidance;
7. Where a treatment is approved CCG Commissioners will respect patient choice as to where a treatment is delivered; AND
8. All policy decisions are considered within the wider constraints of the CCG's legally responsibility to remain fiscally responsible.

Category: Restricted

Why is Non-Invasive Ventilation (NIV) used and what is it?

When we breathe in, we take oxygen out of the air to keep us alive - this oxygen is transferred to our blood in our lungs. The body then uses the oxygen and produces a waste gas called carbon dioxide, which we breathe out. The process of this exchange is ventilation.

Some people with severe lung disease, have problems getting sufficient oxygen into the body, which results in hypoxaemia. If their oxygen level drops below a certain level, it is relatively easy to give extra oxygen for them to breathe, which is called oxygenation. However, in some severe cases of obstructive lung conditions, muscle weakness or neurological impairment, the extra effort of trying to keep the oxygen at a satisfactory level in the blood and to expel carbon dioxide results in the person tiring and leading to hypoventilation and hypercapnia causing respiratory failure.

Respiratory failure is more difficult to deal with. It is a particular problem with diseases that cause obstruction to our airways, such as chronic obstructive pulmonary disease (COPD). In COPD, the airways are narrowed, making it harder to get oxygen into the lungs and carbon dioxide out. Patients who have weak or denervated respiratory muscles in neuromuscular/neurological conditions are also unable to take in a sufficient volume of air to expel carbon dioxide. In all these conditions, a person can develop type 2 respiratory failure which cannot be corrected with oxygenation as the person needs help to ventilate to expel carbon dioxide. Type 2 respiratory failure can lead to high heart rate and cardiac complications.

The aim of using Non-Invasive ventilation (NIV) is not only to obtain satisfactory oxygen levels, but also to expire carbon dioxide. It is often first used at night when the patient is asleep and carbon dioxide levels increase, but as the patient's condition progresses, NIV may be required in the day when the patient has diurnal respiratory failure. It is also important to ease the work of breathing associated with respiratory failure as when a patient with respiratory failure becomes overly tired, this can lead to fatigue, further respiratory compromise and potential respiratory arrest. NIV also aims to take some of the effort out of breathing because the patient's chest muscles don't have to work as hard, so it helps to ease the feelings of breathlessness.

People receiving NIV need to wear a cushioned mask or use a mouthpiece, which is connected to an air pump machine. This mask fits either over the nose alone, or over both the nose and mouth; a strap holds the mask firmly in place, but it can be easily removed, to enable, for example, the patient to eat and drink.

Types of Non-Invasive Ventilation

Noninvasive ventilation (NIV) refers to the administration of ventilatory support without using an invasive artificial airway (endotracheal tube or tracheostomy tube). The use of noninvasive ventilation has markedly increased over the past three decades, and noninvasive ventilation has now become an integral tool in the management of both acute and chronic respiratory failure, in both the home setting and in critical care.

In its simplest terms, noninvasive ventilation differs from invasive ventilation by the interface between the patient and the ventilator. Invasive ventilatory support is provided via either an endotracheal tube or tracheostomy tube. Noninvasive ventilatory support uses a variety of interfaces, and these have continued to evolve with modifications based on patient comfort and efficacy. Many of the interfaces or masks were initially used in patients with obstructive sleep apnea before they were adapted for use in patients to provide noninvasive ventilatory support.

Nasal masks and orofacial masks were the earliest interfaces, with subsequent development and use of full-face masks, mouthpieces, nasal pillows, and helmets. Nasal masks and orofacial masks are still the most commonly used interfaces. Orofacial masks are used almost twice as frequently as nasal masks. Both have advantages and disadvantages in the application of noninvasive ventilation.

Noninvasive positive-pressure ventilation

Positive-pressure ventilation delivered through a mask, has become the predominant method of providing noninvasive ventilatory support. Early bedside physiologic studies in healthy patients and in patients with respiratory conditions document successful ventilatory support (ie, reduction in respiratory rate, increase in tidal volume, decrease in dyspnea) with reduction in diaphragmatic electromyography (EMG), transdiaphragmatic pressures, work of breathing and improvement in oxygenation with a reduction in hypercapnia.

Ventilatory support can be achieved through a variety of interfaces (mouth piece or nasal, face, or helmet mask), using a variety of ventilatory modes (eg, volume ventilation, pressure support, bilevel positive airway pressure [BiPAP], proportional-assist ventilation [PAV], continuous positive airway pressure [CPAP]) with either ventilators dedicated to noninvasive ventilation (NIV) or those capable of providing support through an endotracheal tube or mask. Older models of noninvasive ventilators required oxygen to be bled into the system, but current models incorporate oxygen blenders for precise delivery of the fraction of inspired oxygen (FIO_2).

Noninvasive negative-pressure ventilation

Negative-pressure ventilators provide ventilatory support using a device that encases the thoracic cage starting from the neck, and devices range from a whole-body tank to a cuirass shell. The general principal is the same with a vacuum device, which lowers the pressure surrounding the thorax, creating sub-atmospheric pressure and thereby passively expanding the chest wall with diaphragmatic descent, all leading to lung inflation. Exhalation occurs with passive recoil of the chest wall.

This was the predominant technology during the polio epidemics, but these devices were bulky and cumbersome to use. Upper airway obstruction was also a problem. These ventilators have been largely supplanted by the more widespread positive-pressure noninvasive ventilators; however, some patients continue to be treated with this modality. While the bulk of the experience lies in patients with chronic respiratory failure, specifically neuromuscular respiratory failure, reports described successful application in patients with acute respiratory failure.

Current use of Non-invasive Ventilation devices.

With respect to the two modes, positive-pressure ventilation has supplanted negative-pressure ventilation as the dominant mode of delivery of noninvasive ventilation. Positive-pressure ventilation is more effective than negative-pressure ventilation in unloading the respiratory muscles, at least under investigational conditions. The primary focus of this policy is domiciliary positive-pressure noninvasive ventilation, and the mention of "noninvasive ventilation" will refer to positive-pressure delivery.

Many patients who are assessed as requiring noninvasive ventilation are provided support with pressure ventilation, i.e. continuous positive airway pressure (CPAP), which is the most basic level of support. CPAP pumps a steady flow of air at constant pressure through the nose to prevent the narrowing or collapse of air passages or to help the lungs to expand. CPAP may be especially useful in patients with congestive heart failure or obstructive sleep apnea.

Bilevel positive airway pressure (BiPAP) is probably the most common mode of noninvasive positive pressure ventilation and requires provisions for inspiratory positive airway pressure (IPAP) and expiratory positive airway pressure (EPAP). The difference between IPAP and EPAP is a reflection of the amount of pressure support ventilation provided to the patient, and EPAP is synonymous with positive end-expiratory pressure (PEEP). Some noninvasive ventilation is provided using proportional-assist ventilation (PAV), which provides flow and volume assistance with each breath. Clinical trials have not demonstrated a significant difference between PAV and pressure-support ventilation with BiPAP. [5, 6] However, BiPAP is the most commonly available and more frequently used modality for noninvasive ventilation. PAV remains available on many ventilator models, but use is much less common than BiPAP.

National context

National Guidance for the provision of aspects of specialist non-ventilation services to patients exists for some individual patient groups e.g. Motor Neurone Disease (MND), Duchene's Muscular Dystrophy; and for broader categories of patients e.g. weaning guidance; and around specific technologies e.g. diaphragmatic pacing and tracheostomies. There are some national standards (NICE, 2010; 2016) available and some specialist society guidance (BTS/ICS 2016).

Provision of complex home ventilation services also falls within the NHS Outcomes Framework Domain 1 - preventing people from dying prematurely where Improvement Area 1a specifically identifies reducing mortality from respiratory disease, and Domain 2 – enhancing quality of life for patients with long term conditions.

Guidance supports delivery of care by respiratory specialists working within MDTs. For example, the National Institute for Health and Clinical Excellence (NICE) clinical guideline (CG) around use of NIV in MND states that "multidisciplinary teams (MDT) should coordinate and provide on-going management and treatment for patients with MND, including regular respiratory assessment and provision of non-invasive ventilation. The team should include a neurologist, a respiratory physician, an MND specialist nurse, a respiratory specialist nurse, a specialist respiratory physiotherapist, a respiratory physiologist, a specialist in palliative care and a speech and language therapist". The guidance also outlines the support and training which need to be provided to the patient and their family and carers: "support and assistance to manage non-invasive ventilation which should include training on using non-invasive ventilation and ventilator interfaces, for example emergency procedures, night-time assistance if the patient is unable to use the equipment independently (for example, emergency removal or replacement of interfaces), how to use the equipment with a wheelchair or other mobility aids if required, what to do if the equipment fails, assistance with secretion management, information on general palliative strategies, an offer of on-going emotional and psychological support for the patient and their family and carers".

Ensuring NIV is delivered by competent respiratory professionals is emphasised in NICE MND guidance and also in the National Patient Safety Agency (NPSA) alert which identified cases where problems with administering NIV were stated as causing at least moderate harm: key issues included shortage of staff skills or staff time to set up and monitor NIV.

Local context

The CCG, on the basis of strong supporting evidence for the clinical effectiveness of the intervention, will commission the use of domiciliary non-invasive ventilation in the following clinical conditions where the patient's individual clinical circumstances meet the relevant clinical eligibility criteria outlined in Sections A; B; C; D & E respectively:

- Chronic Obstructive Pulmonary Disease (Section A)
- Neuro-dependent Patients (Section B)
- Obstructive Sleep Apnoea (Section C)
- Thoracic cage deformities (Section D) – awaiting CPAG Scorecard
- Obesity-Related Rspiratory failure (Section E – awaiting CPAG Scorecard)

Commented [KD1]: To be added in once CPAG have reviewed via scorecards in August 2019

Please note the provision of non-invasive ventilation for cystic fibrosis patients is a specialised service commissioned by NHSE.

Guidance

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NIV – Section A – Chronic Obstructive Pulmonary Disease (COPD)

Chronic obstructive pulmonary disease (COPD) is the collective name for a group of lung conditions that may cause breathing difficulties.

It includes:

- **emphysema** – damage to the air sacs in the lungs
- **chronic bronchitis** – long-term inflammation of the airways

COPD is a common condition that mainly affects middle-aged or older adults who have a smoking history. The breathing problems tend to get gradually worse over time and can limit the patient's normal activities, although treatment can help keep the condition under control.

Symptoms of COPD

The main symptoms of COPD are:

- increasing breathlessness, particularly when the patient is active
- a persistent chesty cough with phlegm
- frequent chest infections
- persistent wheezing

Without treatment, the symptoms usually get slowly worse. There may also be periods when they get suddenly worse, known as a flare-up or exacerbation.

Causes of COPD

COPD occurs when the lungs become inflamed, damaged and narrowed. The main cause is smoking, although the condition can sometimes affect people who have never smoked.

The likelihood of developing COPD increases the more a patient smokes and the longer the patient has smoked. Some cases of COPD are caused by long-term exposure to harmful fumes, or dust or occur as a result of a rare genetic problem that means the lungs are more vulnerable to damage.

The damage to the lungs caused by COPD is permanent, but treatment can help slow down the progression of the condition.

Treatments include:

- smoking cessation – if a patient is diagnosed withCOPD still smokes, stopping smoking is the most important thing a patientcan do
- inhalers and medications
- pulmonary rehabilitation – a specialised programme of exercise and education
- surgery or a lung transplant –an option for a very small number of people

Chronic obstructive pulmonary disease (COPD) is characterized by recurrent exacerbations that can cause intermittent periods of severe clinical deterioration requiring hospitalization and ventilator support. Although treating patients with COPD and acute respiratory failure with non-invasive ventilation improves outcomes,persistent hypercapnia after an exacerbation is associated with excess mortality and early rehospitalization. In 2013, the 28-day COPD readmission rate was around 20%,6 (Suh et al. 2015).

Eligibility Criteria: Restricted

For patients with chronic COPD the CCG will commission the use of domiciliary non-invasive ventilation in the following clinical circumstances:

The patient has a diagnosis of COPD, identified by post bronchodilator Forced Expiratory Volume (FEV)1 / Forced Vital Capacity (FVC) <0.70

AND

- The patient must have an awake, non acute, stable $paCO_2$ level equal to or greater than 6.5kpa

  AND the patient must have ONE of the following:

A reduction in Quality of life identified by symptoms consistent with Sleep disordered breathing problems (see pg13 for definition)

  - If the patient has reduced quality of life then overnight oximetry should be undertaken to demonstrate that the patient meets ONE of the following criteria:
    - An apnoea/hyponoea index >10/hour on polysomnography
    - Four or more episodes of SpO2 <92%
    - Drops in SpO2 of at least 4% per hour of sleep)

  OR

- A co-morbidity secondary to hypoxemia
  - Pulmonary Hypertension
  - Heart Failure

If the patient has co-morbidities secondary to Hypoxemia then the patient should also meet the following criteria:

- Recurrent NIV admissions (2 or more in a 12 month period OR difficulty weaning / unable to tolerate weaning)
  AND
- Acute use of NIV has been well tolerated

N.B. Symptoms consistent with Sleep disordered breathing problems are defined as:

- Excessive daytime somnolence (a state of strong desire for sleep, or sleeping for unusually long periods)
- Headache
- Confusion
- Increased shortness of breath
- Resting tremor

Exclusion criteria:

- Inability to remove mask independently (with no waking night carer)
- Cognitive / behavioural limitation affecting ability to comply safely with NIV
- Intolerance of acute NIV
- Multiple co-morbidities limiting utility of NIV
- The patient continues to smoke

Commented [KD2]: This has been suggested by one of the reviewing clinicians. If oxygen is being delivered then smoking is dangerous, but NIV is delivery of air – we do not restrict surgery if a patient smokes, can the CCG defend restricting NIV if a patient smokes?

Funding will be provided for the following if the patient with COPD meets the above clinical criteria:

- One NIV machine
- +/- Humidifier as required
- 1-2 lengths of tubing per year
- 1-2 masks per year

This means **(for patients who DO NOT meet the above criteria)** the CCG will **only** fund the treatment if an Individual Funding Request (IFR) application proves exceptional clinical need and that is supported by the CCG.

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NIV – Section B – Neuro-Muscular Patients

A number of chronic neuromuscular disorders, for example muscular dystrophy and motor neurone disease lead to progressive respiratory muscle dysfunction, which in turn can lead to respiratory failure and death. Nocturnal and daytime Non-Invasive Ventilation (NIV) is becoming the preferred method of treatment for these disorders[1].

Non-invasive ventilation as a treatment for neuromuscular disease has several benefits. It has been shown to[1,5,6]:

- Improve blood gases
- Decrease work of breathing
- Improve symptoms of fatigue
- Reduce daytime sleepiness
- Improve morning headaches
- Improve survival in Duchenne muscular dystrophy (DMD) and amyotrophic lateral sclerosis (ALS) patients[7,8]

Patients with one of the following conditions will be considered for funding when the patient also meets the eligibility criteria outlined below.

- Motor Neurone Disease
- Spinal Muscular Atrophy
- Spinal cord injury
- Multiple Sclerosis
- Guillain-Barre Syndrome
- Post polio syndrome with respiratory impairment
- Syringomyelia
- Tuberculosis infection with respiratory impairment
- Other neuromuscular disease which is known to cause respiratory muscle weakness or upper airway functional impairment.

Commented [KD3]: Kyphoscoliosis To be moved to section D – thoracic cage deformities

Eligibility Criteria: Restricted

For patients diagnosed with a neuromuscular condition as outlined above, the patient must meet the following criteria for funding for non-invasive ventilation to be approved:

Nocturnal Ventilation

The patient must meet ONE of the following criteria:

- Signs (<50% predicted/<1l) or symptoms of hypoventilation
- MIP< 60cmH_2O
- A baseline SpO_2 <95%
- Blood or end tidal pCO2 >45mmHg whilst awake
- Four or more episodes of SpO2 <92%
- Drops in SpO2 of at least 4% per hour of sleep

Daytime Ventilation (in addition to meeting the above criteria the patient must also meet ONE of the following criteria):

- Abnormal deglutition due to dyspnea, which is relieved by ventilatory assistance
- Inability to speak in full sentences without breathlessness
- Symptoms of hypoventilation with baseline SpO2 <95%
- Blood or end tidal pCO2 >45mmHG whilst awake
- Symptoms of awake dyspnoea are present

Exclusion criteria:

- Inability to remove mask independently (with no waking night carer
- Cognitive / behavioural limitation affecting ability to comply safely with NIV
- Intolerance of acute NIV
- Multiple co-morbidities limiting utility of NIV

Funding will be provided for the following if the patient meets the above clinical criteria:

Below 14 hours of ventilation required.

- One NIV machine
- +/- Humidifier as required
- 1-2 lengths of tubing per year
- 1-2 masks per year

Above 14 hours / 24 hour period of ventilation required.

- Two NIV machine
- +/- ONE Humidifier as required
- 2-4 lengths of tubing per year
- 2-4 masks per year

This means **(for patients who DO NOT meet the above criteria)** the CCG will **only** fund the treatment if an Individual Funding Request (IFR) application proves exceptional clinical need and that is supported by the CCG.

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NIV – Section C – Obstructive Sleep Apnoea Hypopnea Syndrome (OSAHS)

Obstructive Sleep Apnoea Hypopnea Syndrome (OSAHS)

Apnoea is defined as a temporary absence or cessation of breathing. Obstructive Sleep Apnoea hypopnea syndrome (OSAHS) is a condition in which a person experiences repeated episodes of apnoea because of a narrowing or closure of the pharyngeal airway during sleep. This is caused by a decrease in the tone of the muscles supporting the airway during sleep. Complete closure (obstruction) stops airflow (apnoea) whereas partial obstruction decreases airflow (hypopnoea). OSAHS results in episodes of brief awakening from sleep to restore normal breathing.

Moderate to severe OSAHS can be diagnosed from patient history and a sleep study using oximetry or other monitoring devices carried out in the person's home. In some cases, further studies that monitor additional physiological variables in a sleep laboratory or at home may be required, especially when alternative diagnoses are being considered. The severity of OSAHS is usually assessed on the basis of both severity of symptoms (particularly the degree of sleepiness) and the sleep study, by using either the apnoea/hypopnoea index (AHI) or the oxygen desaturation index. OSAHS is considered mild when the AHI is 5–14 in a sleep study, moderate when the AHI is 15–30, and severe when the AHI is over 30. In addition to the AHI, the severity of symptoms is also important.

The symptoms of OSAHS include impaired alertness, cognitive impairment, excessive daytime sleepiness, snoring, nocturia, morning headaches and sexual dysfunction. The sleep quality of partners may also be affected. Excessive daytime sleepiness can adversely affect cognitive function, mood and quality of life. OSAHS is associated with high blood pressure, which increases the risk of cardiovascular disease and stroke. OSAHS has also been associated with an increased risk of road traffic accidents.

Major risk factors for developing OSAHS are increasing age, obesity and being male. OSAHS is also associated with certain specific craniofacial characteristics (such as retrognathia), enlarged tonsils and enlarged tongue. Use of alcohol or sedatives can also increase the risk or severity of the condition. OSAHS has been reported to affect up to 4% of middle-aged men and 2% of middle-aged women in the UK. It is estimated that 1% of men in the UK may have severe OSAHS.

The use of Non-Invasive Ventilation in OSAHS.

Treatment for OSAHS aims to reduce daytime sleepiness by reducing the number of episodes of apnoea/hypopnoea experienced during sleep. The type of non-invasive ventilation most commonly used in the clinical management of sleep apnoea is continuous positive airway pressure (CPAP).

The alternatives to CPAP are:

- lifestyle management,
- dental devices
- surgery.

Lifestyle management involves helping people to lose weight, stop smoking and/or decrease alcohol consumption.

Dental devices are designed to keep the upper airway open during sleep. The efficacy of dental devices has been established in clinical trials, but these devices are traditionally viewed as a treatment option only for mild and moderate OSAHS.

Surgery involves resection of the uvula and redundant retrolingual soft tissue. However, there is a lack of evidence of clinical effectiveness, and surgery is not routinely used in clinical practice.

A CPAP device consists of a unit that generates airflow, which is directed to the airway via a mask. Positive pressure is generated by the airflow, which prevents upper airway collapse. For CPAP treatment to be effective the patient must always wear their device when they go to sleep.

Reasons for not adhering to CPAP treatment include poor mask fit, pressure intolerance and, more commonly, upper airway symptoms such as nasal dryness, nasal bleeding and throat irritation. Humidification devices are now commonly used in conjunction with CPAP devices in order to reduce these side effects. Masks should be replaced at least annually, and long-term follow-up of patients is critical to ensure adherence.

There are two types of CPAP devices. Fixed CPAP devices deliver air at constant pressure throughout the night, and the person will continue to receive this pressure until a further titration study is performed to determine whether the set pressure is still appropriate. Auto-titrating CPAP devices continually adjust the pressure delivered throughout the night, with the aim of improving comfort and thus adherence.

Eligibility Criteria: Restricted

1. Continuous positive airway pressure (CPAP) is commissioned as a treatment option for adults with moderate or severe symptomatic obstructive sleep apnoea/hypopnoea syndrome (OSAHS).

2. CPAP is only recommended as a treatment option for adults with mild OSAHS if:

    a. The OSAHS is causing severe functional impairment, which is impacting on the patient's ability to carry out activities of daily living

AND

    b. lifestyle advice and any other relevant treatment options have been unsuccessful or are considered inappropriate

The diagnosis and treatment of OSAHS, and the monitoring of the response, should always be carried out by a specialist service with appropriately trained medical and support staff.

N.B. The definition of OSAHS following a sleep study is as follows:
Mild OSAHS= Apnoea–Hypopnoea Index (AHI) 5–14.
Moderate OSAHS = AHI is 15–30.
Severe OSAHS = AHI is over 30.

Functional impairment is defined as preventing activities of daily living to be undertaken independently, i.e. sleeping; eating; walking, driving.

Exclusion criteria:

- Inability to remove mask independently (with no waking night carer)
- Cognitive / behavioural limitation affecting ability to comply safely with NIV
- Intolerance of acute NIV
- Multiple co-morbidities limiting utility of NIV

Funding will be provided for the following if the patient meets the above clinical criteria:

- One CPAP machine
- 1-2 lengths of tubing per year
- 1-2 masks per year

In a small proportion of OSA patients, CPAP proves insufficient to control apnoeas and it becomes necessary to use bi-level NIV. If a patient has failed treatment with CPAP, but continues to meet the eligibility criteria outlined above, a further funding application will be considered for:

- One Bi-level NIV machine
- 1-2 lengths of tubing per year
- 1-2 masks per year

This means **(for patients who DO NOT meet the above criteria)** the CCG will **only** fund the treatment if an Individual Funding Request (IFR) application proves exceptional clinical need and that is supported by the CCG.

Guidance - OSA

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NIV – Section D – Thoracic Cage Deformities

NIV – Section E – Obesity Related Respiratory Failure