NHS Birmingham and Solihull CCG
NHS Sandwell and West Birmingham CCG

Policy for use of domiciliary Non-Invasive Ventilation

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

57. CCG Commissioners require clear evidence of clinical effectiveness before NHS resources are invested in the treatment;
58. CCG Commissioner require clear evidence of cost effectiveness before NHS resources are invested in the treatment;
59. The cost of the treatment for this patient and others within any anticipated cohort is a relevant factor;
60. CCG Commissioners will consider the extent to which the individual or patient group will gain a benefit from the treatment;
61. 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
62. CCG Commissioners will consider all relevant national standards and take into account all proper and authoritative guidance;
63. Where a treatment is approved CCG Commissioners will respect patient choice as to where a treatment is delivered; AND
64. 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 enough 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 apnoea 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. Hybrid 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 physiological studies in healthy patients and in patients with respiratory conditions document successful ventilatory support (i.e., reduction in respiratory rate, increase in tidal volume, decrease in dyspnoea) 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 (e.g., volume ventilation, pressure support, bilevel positive airway pressure [BiPAP], proportional-assist ventilation [PAV]) 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).

Current use of Non-invasive Ventilation devices.

Bilevel positive airway pressure (BiPAP) is probably the most common mode of noninvasive positive pressure ventilation and provides for inspiratory positive airway pressure (IPAP) and expiratory positive airway pressure (EPAP). The difference between IPAP and EPAP reflects 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 (DMD); 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,
Domain 2 – enhancing quality of life for patients with long term conditions
Domain 3 – helping patients to recover after an episode of acute illness, where post-acute admission, non-invasive ventilation has been shown to help people recover better in the community and reduce readmission rates.

Guidance supports delivery of care by respiratory specialists working within MDTs. For example, the National Institute for Health and Clinical Excellence (NICE) clinical guideline around the 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, a 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, based on 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 respectively:

- Chronic Obstructive Pulmonary Disease (Section A)
- Neuro-muscular and Neurological Weakness Patients (Section B)

Please note the provision of treatment for patients with Cystic Fibrosis and patients with Spinal Muscular Atrophy are specialised services commissioned by NHSE.

Guidance

1. Corrado A, Gorini M, Melej R, et al. Iron lung versus mask ventilation in acute exacerbation of COPD: a randomised crossover study. *Intensive Care Med*. 2009 Apr. 35(4):648-55.
2. Parke RL, McGuinness SP. Pressures delivered by nasal high flow oxygen during all phases of the respiratory cycle. *Respir Care*. 2013 Oct. 58 (10):1621-4.
3. Spoletini G, Alotaibi M, Blasi F, Hill NS. Heated Humidified High-Flow Nasal Oxygen in Adults: Mechanisms of Action and Clinical Implications. *Chest*. 2015 Jul. 148 (1):253-61.
4. Ozsancak A, Sidhom S, Liesching TN, Howard W, Hill NS. EVALUATION OF THE TOTAL FACE MASKTM FOR NONINVASIVE VENTILATION TO TREAT ACUTE RESPIRATORY FAILURE. *Chest*. 2011 Feb 17.
5. Wysocki M, Richard JC, Meshaka P. Noninvasive proportional assist ventilation compared with noninvasive pressure support ventilation in hypercapnic acute respiratory failure. *Crit Care Med*. 2002 Feb. 30 (2):323-9.
6. Fernández-Vivas M, Caturla-Such J, González de la Rosa J, Acosta-Escribano J, Alvarez-Sánchez B, Cánovas-Robles J. Noninvasive pressure support versus proportional assist ventilation in acute respiratory failure. *Intensive Care Med*. 2003 Jul. 29 (7):1126-33.
7. Hoo, G. 2018. Noninvasive Ventilation. Medscape. https://emedicine.medscape.com/article/304235-overview#a5
8. British Thoracic Society/Intensive Care Society Acute Hypercapnic Respiratory Failure Guideline Development Group. 2016. BTS/ICS Guidelines for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults. Journal of the British Thoracic Society. http://thorax.bmj.com/site/about/guidelines.xhtml#open
9. National Institute for Health and Clinical Excellence (NICE). Motor neurone disease: assessment and management**.** NICE guideline [NG42] Published date: February 2016 Last updated: July 2019
10. National Institute for Health and Clinical Excellence (NICE). *Chronic Obstructive Pulmonary Disease in Over 16s: Diagnosis and Management [CG101]*. London, England: NICE; 2010. https://www.nice.org.uk/guidance/ng42
11. Murphy, P. Rehal, S. et al. 2017. Effect of Home Noninvasive Ventilation With Oxygen Therapy vs Oxygen Therapy Alone on Hospital Readmission or Death After an Acute COPD Exacerbation: A Randomized Clinical Trial. *JAMA.* 2017;317(21):2177-2186. doi:10.1001/jama.2017.4451. https://jamanetwork.com/journals/jama/issue/317/21

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 with COPD still smokes, stopping smoking is the most important thing a patient can 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 hospitalisation 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%, (Suh et al. 2015).

Eligibility Criteria: Restricted

For patients with 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

4 weeks post-acute admission the patient has a paCO2 over 7 kPa.

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 pg12 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/hypopnoea index >10/hour on respiratory polysomnography or multi-channel respiratory sleep study
        - 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 12month 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 as per the Epworth Sleepiness Score)
- 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

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.

Guidance – Section A: COPD

1. Brochard L, Mancebo J,Wysocki M, et al. Noninvasive ventilation for acute exacerbations of chronic obstructive pulmonary disease. *N Engl J Med*. 1995;333(13):817-822.
2. Bott J, Carroll MP, Conway JH, et al. Randomised controlled trial of nasal ventilation in acute ventilatory failure due to chronic obstructive airways disease. *Lancet*. 1993;341(8860):1555-1557.
3. Connors AF Jr, Dawson NV, Thomas C, et al; SUPPORT Investigators (Study to Understand Prognoses and Preferences for Outcomes and Risks of Treatments). Outcomes following acute exacerbation of severe chronic obstructive lung disease. *Am J Respir Crit Care Med*. 1996;154(4 pt 1): 959-967.
4. Murray I, Paterson E, Thain G, Currie GP. Outcomes following non-invasive ventilation for hypercapnic exacerbations of chronic obstructive pulmonary disease. *Thorax*. 2011;66(9):825-826.
5. Chu CM, Chan VL, Lin AW,Wong IW, LeungWS, Lai CK. Readmission rates and life threatening events in COPD survivors treated with non-invasive ventilation for acute hypercapnic respiratory failure. *Thorax*. 2004;59(12):1020-1025.
6. Suh ES, Mandal S, Harding R, et al. Neural respiratory drive predicts clinical deterioration and safe discharge in exacerbations of COPD. *Thorax*. 2015;70(12):1123-1130.
7. Nickol AH, Hart N, Hopkinson NS, et al. Mechanisms of improvement of respiratory failure in patients with COPD treated with NIV. *Int J Chron Obstruct Pulmon Dis*. 2008;3(3):453-462.
8. Meecham Jones DJ, Paul EA, Jones PW, Wedzicha JA. Nasal pressure support ventilation plus oxygen compared with oxygen therapy alone in hypercapnic COPD. *Am J Respir Crit Care Med*. 1995;152(2):538-544.
9. Elliott MW, Mulvey DA,Moxham J, Green M, Branthwaite MA. Domiciliary nocturnal nasal intermittent positive pressure ventilation in COPD: mechanisms underlying changes in arterial blood gas tensions. *Eur Respir J*. 1991;4(9):1044-1052.
10. Lloyd-Owen SJ, Donaldson GC, Ambrosino N, et al. Patterns of home mechanical ventilation use in Europe: results from the Eurovent survey. *Eur Respir J*. 2005;25(6):1025-1031.
11. Clini E, Sturani C, Rossi A, et al; Rehabilitation and Chronic Care Study Group, Italian Association of Hospital Pulmonologists (AIPO). The Italian multicentre study on noninvasive ventilation in chronic obstructive pulmonary disease patients. *Eur Respir J*. 2002;20(3):529-538.
12. McEvoy RD, Pierce RJ, Hillman D, et al; Australian trial of non-invasive Ventilation in Chronic Airflow Limitation (AVCAL) Study Group. Nocturnal non-invasive nasal ventilation in stable hypercapnic COPD: a randomised controlled trial. *Thorax*. 2009;64(7):561-566.
13. WindischW. Noninvasive positive pressure ventilation in COPD. *Breathe*. 2011;8(2):114-123.
14. Köhnlein T, WindischW, Köhler D, et al. Non-invasive positive pressure ventilation for the treatment of severe stable chronic obstructive pulmonary disease: a prospective, multicentre, randomised, controlled clinical trial. *Lancet Respir Med*. 2014;2(9):698-705.

15. Struik FM, Sprooten RT, Kerstjens HA, et al. Nocturnal non-invasive ventilation in COPD patients with prolonged hypercapnia after ventilatory support for acute respiratory failure: a randomised, controlled, parallel-group study. *Thorax*. 2014;69(9):826-834.
16. Altman DG, Bland JM. Treatment allocation by minimisation. *BMJ*. 2005;330(7495):843.
17. Murphy PB, Brignall K, Moxham J, Polkey MI, Davidson AC, Hart N. High pressure versus high intensity noninvasive ventilation in stable hypercapnic chronic obstructive pulmonary disease: a randomized crossover trial. *Int J Chron Obstruct Pulmon Dis*. 2012;7:811-818.
18. 18. National Institute for Health and Clinical Excellence (NICE). Motor neurone disease: assessment and management**.** NICE guideline [NG42] Published date: February 2016 Last updated: July 2019
19. National Institute for Health and Clinical Excellence (NICE). *Chronic Obstructive Pulmonary Disease in Over 16s: Diagnosis and Management [CG101]*. London, England: NICE; 2010. https://www.nice.org.uk/guidance/ng42
20. Bestall JC, Paul EA, Garrod R, Garnham R, Jones PW,Wedzicha JA. Usefulness of the Medical Research Council (MRC) dyspnoea scale as a measure of disability in patients with chronic obstructive pulmonary disease. *Thorax*. 1999;54(7): 581-586.20. Ghosh D, Rzehak P, ElliottMW, WindischW. Validation of the English Severe Respiratory Insufficiency Questionnaire. *Eur Respir J*. 2012;40 (2):408-415.
21. Jones PW, Quirk FH, Baveystock CM. The St George's Respiratory Questionnaire. *Respir Med*. 1991;85(suppl B):25-31.
22. Jones PW. St George's Respiratory Questionnaire: MCID. *COPD*. 2005;2(1):75-79
23. Kahan BC, Morris TP. Improper analysis of trials randomised using stratified blocks or minimisation. *Stat Med*. 2012;31(4):328-340.
24. Costello R, Deegan P, Fitzpatrick M, McNicholasWT. Reversible hypercapnia in chronic obstructive pulmonary disease: a distinct pattern of respiratory failure with a favourable prognosis. *Am J Med*. 1997;102(3):239-244.
25. Seemungal TA, Donaldson GC, Paul EA, Bestall JC, Jeffries DJ,Wedzicha JA. Effect of exacerbation on quality of life in patients with chronic obstructive pulmonary disease. *Am J Respir Crit Care Med*. 1998;157(5 pt 1):1418-1422.
26. De Backer L, VosW, Dieriks B, et al. The effects of long-term non-invasive ventilation in hypercapnic COPD patients: a randomized controlled pilot study. *Int J Chron Obstruct Pulmon Dis*. 2011;6:615-624.
27. Gates KL, Howell HA, Nair A, et al. Hypercapnia impairs lung neutrophil function and increases mortality in murine pseudomonas pneumonia. *Am J Respir Cell Mol Biol*. 2013;49(5):821-828.
28. Pepperell JC, Ramdassingh-Dow S, Crosthwaite N, et al. Ambulatory blood pressure after therapeutic and subtherapeutic nasal continuous positive airway pressure for obstructive sleep apnoea: a randomised parallel trial. *Lancet*. 2002;359(9302):204-210.
29. Saatci E, Miller DM, Stell IM, Lee KC, Moxham J. Dynamic dead space in face masks used with non-invasive ventilators: a lung model study. *Eur Respir J*. 2004;23(1):129-135.
30. Rodway GW,Weaver TE, Mancini C, et al. Evaluation of sham-CPAP as a placebo in CPAP intervention studies. *Sleep*. 2010;33(2):260-266.

31. Djavadkhani Y, Marshall NS, D'Rozario AL, et al. Ethics, consent and blinding: lessons from a placebo/sham controlled CPAP crossover trial. *Thorax*. 2015;70(3):265-269.
32. Schwartz SW, Cimino CR, Anderson WM. CPAP or placebo-effect? *Sleep*. 2012;35(12):1585- 1586.
33. Murphy, P. Rehal, S. et al. 2017. Effect of Home Noninvasive Ventilation With Oxygen Therapy vs Oxygen Therapy Alone on Hospital Readmission or Death After an Acute COPD Exacerbation: A Randomized Clinical Trial. *JAMA.* 2017;317(21):2177-2186. doi:10.1001/jama.2017.4451. https://jamanetwork.com/journals/jama/issue/317/21
34. Murphy PB, Arbane G, Bourke S, Calverley P, Crooks A, Dowson L, Duffy N, Gibson GJ, Hughes SP, Hurst JR, Lewis KR**, Mukherjee R**, Nickol A, Oscroft N, Pepperell J, Rehal S, Smith I, Stradling J, Wedizcha W, Polkey MI, Elliott M, Hart N. Effect of home noninvasive ventilation with oxygen therapy vs. oxygen therapy alone on hospital readmission or death after an acute COPD exacerbation: a randomized clinical trial. *JAMA* 2017; 317(21):2177-2186. **doi:** 10.1001/jama.2017.4451 [PubMed ID: 28528348]
35. 35. Dretzke J, et al. The cost-effectiveness of domiciliary non-invasive ventilation in patients with end-stage chronic obstructive pulmonary disease: a systematic review and economic evaluation. *Health technology assessment.* 10/2015; **19**(81):1-246. **doi**: 10.3310/hta19810. [PubMed ID: 26470875 PMCID: PMC4781210]

NIV – Section B –Patients with Neuro-muscular and Neurological weakness

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 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:

- Improves lung mechanics and gas exchange
- Decrease work of breathing
- Improve symptoms of fatigue
- Reduce daytime sleepiness
- Improve survival in Duchenne Muscular Dystrophy (DMD) and Motor Neurone Disease (MND) patients.

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
- Muscular Dystrophies including Duchenne Muscular Dystrophy
- Spinal cord injury
- Multiple Sclerosis
- Guillain-Barre Syndrome
- Post polio syndrome with respiratory impairment
- Syringomyelia
- Tuberculosis infection with residual respiratory insufficiency
- Other neuromuscular impairment which is known to cause respiratory muscle weakness or upper airway functional impairment which are the commissioning responsibility of the CCG.

Eligibility Criteria: Restricted

For patients diagnosed with a neuromuscular condition as outlined above, the patient must meet the following criteria for funding f 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 dyspnoea, 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 machines
- +/- 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.

Guidance for NMD

1. Joshua O. Benditt1 and Louis J. Boitano. Pulmonary Issues in Patients with Chronic Neuromuscular Disease. Am J Respir Crit Care Med Vol 187, Iss. 10, pp 1046–1055, May 15, 2013
2. Miller RG, Jackson CE, Kasarskis EJ, England JD, Forshew D, Johnston W, Kalra S, Katz JS, Mitsumoto H, Rosenfeld J, et al.; Quality Standards Subcommittee of the American Academy of Neurology. Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: drug, nutritional, and respiratory therapies (an evidence-based review): report of the Quality Standards Subcommittee of the American Academy of Neurology. Neurology 2009;73: 1218–1226.
3. Bourke SC, Gibson GJ. Sleep and breathing in neuromuscular disease. Eur Respir J 2002;19:1194–1201.
4. Birnkrant DJ, Panitch HB, Benditt JO, Boitano LJ, Carter ER, Cwik VA, Finder JD, Iannaccone ST, Jacobson LE, Kohn GL, et al. American College of Chest Physicians consensus statement on the respiratory and related management of patients with Duchenne muscular dystrophy undergoing anesthesia or sedation. Chest 2007; 132:1977–1986.
5. Mehta S, Hill NS. Noninvasive ventilation. Am J Respir Crit Care Med 2001;163:540–577.
6. Eagle M, Baudouin SV, Chandler C, Giddings DR, Bullock R, Bushby K. Survival in Duchenne muscular dystrophy: improvements in life expectancy since 1967 and the impact of home nocturnal ventilation. Neuromuscul Disord 2002;12:926–929.
7. Toussaint M, Steens M, Wasteels G, Soudon P. Diurnal ventilation via mouthpiece: survival in end-stage Duchenne patients. Eur Respir J 2006; 28:549–555
8. Radunovic A, Annane D, Jewitt K, Mustfa N. Mechanical ventilation for amyotrophic lateral sclerosis/motor neuron disease. Cochrane Database Syst Rev 2009;4:CD004427. Update in: Cochrane Database Syst Rev 2013;3:CD004427.
9. Motor neurone disease: assessment and management (2016) - https://www.nice.org.uk/guidance/ng42/chapter/Recommendations#non-invasive-ventilation
10. Outcome of non-invasive positive pressure ventilation in paediatric neuromuscular disease (2004) - https://www.ncbi.nlm.nih.gov/pubmed/14736624
11. Effects of non-invasive ventilation on survival and quality of life in patients with amyotrophic lateral sclerosis: a randomised controlled trial (2006) - https://www.ncbi.nlm.nih.gov/pubmed/16426990

12. Non-invasive ventilation in amyotrophic lateral sclerosis: a 10 year population based study (2012) - https://www.ncbi.nlm.nih.gov/pubmed/22013242
13. Mechanical ventilation for amyotrophic lateral sclerosis/motor neuron disease (2017) - https://www.ncbi.nlm.nih.gov/pubmed/28982219
14. Outcome of non-invasive positive pressure ventilation in paediatric neuromuscular disease (2004) - https://www.ncbi.nlm.nih.gov/pubmed/14736624
15. Polysomnographic evaluation of non-invasive ventilation in children with neuromuscular disease (2014) - https://www.ncbi.nlm.nih.gov/pubmed/24033486
16. Mechanical ventilation for amyotrophic lateral sclerosis/motor neuron disease (2017) - https://www.ncbi.nlm.nih.gov/pubmed/28982219
17. Sleep disordered breathing in motor neurone disease (2018) - http://jtd.amegroups.com/article/view/18178/14526
18. The use of non-invasive ventilation at end of life in patients with motor neurone disease: a qualitative exploration of family carer and health professional experiences (2013) - https://www.ncbi.nlm.nih.gov/pubmed/23462702
19. Sheehan, D. Birnkrant, J. et al. 2018. Respiratory Management of the Patient With Duchenne Muscular Dystrophy Paediatrics, October 2018, vol 142 https://pediatrics.aappublications.org/content/142/Supplement_2/S62
20. Hamada et al. 2011. Indicators for ventilator use in Duchenne muscular dystrophy. Respiratory Medicine. Volume 105, Issue 4, April 2011, Pages 625-629. https://www.sciencedirect.com/science/article/pii/S0954611110005317#tbl1
21. Birnkrant et al 2010. The Respiratory Management of Patients With Duchenne Muscular Dystrophy: A DMD Care Considerations Working Group Specialty Article. Pediatric Pulmonology. DOI 10.1002/ppul.21254. http://muscle.ca/wp-content/uploads/2012/11/Respirarory_Management_DMD_Publication_2010.pdf