Appendix G. Guidelines

Table G.1. Guidelines for all conditions

Organization	Topic	KQ	Statement
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device initiation criteria	KQ1	Generally NIV should be commenced when there is evidence of: Daytime hypercapnia, PaCO2 ≥45mmHg and/or Evidence of nocturnal hypoventilation (in order of recommendation), such as: A rise in PaCO2 of ≥ 8mmHg between evening and morning ABGs or other accurate CO2 surrogate An acute peak rise of ≥ 8mmHg in TcCO2 or ETCO2 A rise in TcCO2 or ETCO2 > 50mmHg for more than 50% of total sleep time Whilst not ideal - when a measure of CO2 is not available - nocturnal oximetry demonstrates sustained oxygen desaturation ≤ 88% for 5 consecutive minutes or SpO2 <90% for >10% of total sleep time and Symptoms of significant sleep disordered breathing associated with nocturnal obstructive or hypopneoic events and/or Otherwise unexplained potential co-morbidity of sleep disorders, such as refractory hypertension, pulmonary hypertension, right heart failure, polycythaemia, cardiovascular disease or stroke.
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device initiation, monitoring, and candidate selection	KQ1	Hypoxia, hypercapnia, or an elevation in serum bicarbonate indicate the need for additional respiratory assessments and interventions. Polysomnography should be performed where there is a history suggestive of sleep disordered breathing or where FVC <40% predicted, base excess > +4mmols/L on arterial blood gases or erect to supine fall in VC of ≥ 25%. Consideration for polysomnography also includes symptoms of impaired sleep quality (such as daytime somnolence, waking headache or grogginess, fatigue, impaired cognition, impaired short-term memory, irritability, anxiety and depression) or symptoms of sleep-disordered breathing (such as frequent awakening, snoring, choking, gasping, waking dry mouth, waking dyspnea or witnessed apneas). Where there is no overt sign of respiratory compromise, serial VC, respiratory muscle testing, peak cough flow and oximetry should be performed to track baseline pulmonary function in suspected individuals. The minimum requirement for identifying sleep hypoventilation is overnight monitoring of oxygen saturation and, where possible, non-invasive carbon dioxide along with evening to morning arterial blood gases. When daytime indicators for NIV have already been met, a full diagnostic PSG measuring sleep quality is not an essential element in determining the need for NIV. Periodic nocturnal studies to identify unexpected problems or correct identified ones is indicated, with the frequency influenced by current response to therapy and the nature of the patient's underlying disorder. Minimum skills and level of knowledge need to be acquired by patients and / or their carers during the process of acclimatisation to NIV. Acclimatisation and education for domiciliary NIV should occur at institutions where there is a sufficient through-put of patients requiring long term NIV. The patient and/or carer are aware who to contact for medical and technical difficulties.
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device initiation, monitoring, and candidate selection	KQ1	The candidate (for home invasive or noninvasive ventilation) should be medically stable without constant or frequent monitoring, tests or treatment changes. The candidate and family must be motivated: Ventilator assisted individuals (VAIs) must express interest in transitioning/living in the community The family should express commitment to having the VAI live in the community.

Organization	Topic	KQ	Statement
			The family is willing to provide support (physical, emotional and financial). The candidate must have an adequate home setting : Identifiable home to live in, suitable to the needs of the VAI. Home is adaptable as necessary. The candidate must have sufficient caregiver support: Caregivers identified and committed to provide sufficient hours of care to meet the needs of the VAI. Available government-funded care hours identified. The candidate must have access to adequate financial resources: Sources of financial assistance identified and accessed. Sufficient financial resources available to meet projected costs The candidate must have access to equipment appropriate for the needs: Appropriate equipment selected and ordered. The candidate must have access to health care support in the community: Follow-up care available as appropriate (tracheotomy tube changes, ventilator reassessments and assessment of the ongoing effectiveness of the ventilatory support). Medical follow-up to allow for appropriate changes to the mode of ventilation (i.e., from invasive to noninvasive and vice versa, from continuous to nocturnal and vice versa). Professional services available post discharge. A government-funded ventilatory service is necessary to provide appropriate access to equipment and respiratory care.
British Thoracic Society, Guidelines for Home Oxygen Use in Adults, 2015[73]	Device initiation, monitoring, and candidate selection	KQ1	A blood gas assessment should be undertaken to exclude worsening hypercapnia and respiratory acidosis. Treatment with modalities of ventilatory support should be considered for patients who are hypercapnic. Patients with baseline hypercapnia can undergo LTOT assessment without adverse outcome but require monitoring of pH and PCO2 levels during and at the end of assessment. Patients with baseline hypercapnia should be monitored for the development of respiratory acidosis and worsening hypercapnia using ABGs after each titration of flow rate, as well as ABG sampling after oxygen titration is complete. Patients who develop a respiratory acidosis and/or a rise in PaCO2 of >1 kPa (7.5 mm Hg) during an LTOT assessment may have clinically unstable disease. These patients should undergo further medical optimization and be reassessed after 4 weeks. Patients who develop a respiratory acidosis and/or a rise in PaCO2 of >1 kPa (7.5 mm Hg) during an LTOT assessment on two repeated occasions, while apparently clinically stable, should only have domiciliary oxygen ordered in conjunction with nocturnal ventilatory support.
Clinical Indications for Noninvasive Positive Pressure Ventilation in Chronic Respiratory Failure due to Restrictive Lung Disease, COPD, and Nocturnal Hypoventilation—A Consensus Conference	Device initiation, monitoring, and candidate selection	KQ1	Certificate of Medical Necessity Should Document diagnosis Document indications Provide required settings Inspiratory parameters (such as tidal volume, pressure, inspiratory time, cycle) Expiratory pressures Rate (as clinically indicated) Supplemental oxygen (flow rate or fraction of inspired oxygen) Alarms (as clinically indicated)

Organization	Topic	KQ	Statement
Report, American Academy of Home Care Physicians, American College of Chest Physicians, American College of Physicians, American Sleep Disorders Association, American Thoracic Society, National Association for Medical Direction of Respiratory Care, 1999[74]			
Clinical Indications for Noninvasive Positive Pressure Ventilation in Chronic Respiratory Failure due to Restrictive Lung Disease, COPD, and Nocturnal Hypoventilation—A Consensus Conference Report, American Academy of Home Care Physicians, American College of Chest Physicians, American College of Physicians, American Sleep Disorders Association, American Thoracic Society, National Association for Medical Direction of Respiratory Care, 1999[74]	Device continuation, compliance, and outcomes	KQ1 and KQ2	It is expected that initial settings may be adjusted by personnel experienced and skilled in the treatment of NIPPV under the direction of the treating physician. At the 60-day reassessment, final settings must be documented Monitoring of Effectiveness Physician reassessment of patient adherence with the use of NIPPV at 30 to 60 days (documentation of machine usage average of ≥20 h/week) Ongoing monitoring and yearly recertification by physician
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device continuation, compliance, and outcomes	KQ1 and KQ2	Usage throughout all sleep periods should be recommended. Once established on therapy, regular monitoring of compliance data should be performed and compliance is deemed adequate at > 4 - 6 hours per night. Patients can be reviewed at 6 to 8 weeks following the commencement of NIV to determine the clinical response to therapy. After initiation of NIV, clinical review should occur within the first 2 to 3 months to assess symptoms, technical problems, ventilator settings, compliance and success. Further clinical reviews should be performed by a Sleep Physician / Respiratory Physician or Respiratory

Organization	Topic	KQ	Statement
			Failure clinic every 6 to 12 months, again assessing symptoms, compliance, technical problems, lung function, oximetry and further investigations (including ABGs and overnight oximetry or PSG) as required. At any time, when there are indications of unsatisfactory results like the recurrence of clinical symptoms or awake blood gases deteriorate despite clinical stability (e.g. absence of recent pulmonary infection) and adequate compliance, then inadequate ventilation must be suspected and objective evaluation during sleep must be undertaken. Outcome measures should include awake ABGs, nocturnal SpO2 and assessment of daytime sleepiness, breathlessness and health related quality of life.
German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010[75]	Device continuation, compliance, and outcomes	KQ1 and KQ2	Initialization of HMV must take place in a centre for HMV. The aim of the therapy is to eliminate hypoventilation under mechanical ventilation, as well as to reduce CO2 to the point of normocapnia during daytime spontaneous breathing. Once optimal ventilation has been achieved, criteria for supplementary oxygen supply must be assessed. The first ventilation control visit must occur in the short-term (4–8 weeks) and therapeutic success is evaluated according to subjective, clinical and technically-measurable parameters. Modifications to the ventilation system (e. g. parameters, ventilation-interface) must take place exclusively in conjunction with the centre for HMV. Identically-built machines with the same settings can be exchanged outside the hospital, whereas different machines must be exchanged under hospital conditions in the centre for HMV.
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device characteristics and titration	KQ3	Simple bilevel devices are suitable for individuals requiring nocturnal and limited daytime ventilatory support only. However, more sophisticated volume or hybrid devices are indicated for patients requiring more than 18 hours/day or where bilevel devices have proven to be inadequate. Ventilator dependent individuals should be titrated on and use ventilators which have been approved for life support and have an alternative battery source to mains power. They also should be supplied with an appropriate back-up ventilator. Machines with “mask off” or “low pressure” and “power failure” alarms are recommended for ventilator dependent patients and in disorders where there is a potential inability to arouse from an interruption to ventilation or when there is an absence of ventilatory responses when awake. Titration for long term NIV settings should occur when the patient is chronically stable (pH>7.35) and free from exacerbation. Adequate IPAP-EPAP difference is required to ameliorate hypoventilation. A Bi-level ventilation should be commenced in the spontaneous mode, unless there is specific evidence that the patient is unable to trigger the machine once baseline leak and settings have been optimized. Complete correction of sleep disordered breathing during the initial titration night is not necessary for improvement of daytime blood gases and symptoms to occur. Spontaneous-timed mode flow generator, or a ventilator, to be provided if Spontaneous mode device does not allow correction of sustained hypercapnia in the presence of central apnea or persisting hypoventilation. Ventilators using flow triggering or volume-cycled mandatory ventilation may be required for patients experiencing difficulty in triggering inspiration.
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute	Device characteristics and titration	KQ3	Pressure-targeted ventilators are the devices of choice for acute NIV. A full face mask (FFM) should usually be the first type of interface used. A range of masks and sizes is required and staff involved in delivering NIV need training in and experience of using them. NIV circuits must allow adequate clearance of exhaled air through an exhalation valve or an integral exhalation

Organization	Topic	KQ	Statement
Hypercapnic Respiratory Failure in Adults, 2016[76]			port on the mask. As patients recover from acute hypercapnic respiratory failure, ventilator requirements change and ventilator settings should be reviewed regularly.
German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010[75]	Device characteristics and titration	KQ3	A second ventilator and an external battery pack are necessary if ventilation periods exceed 16 hours/day. Every non-invasively-ventilated patient requires at least one reserve mask A humidifier is a mandatory requirement for invasive ventilation and is also useful for non-invasive ventilation if typical symptoms are present. In NMD patients with cough insufficiency and in children, selective use of a pulse oximeter is necessary.
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Respiratory services	KQ4	Education and preventive strategies in airway clearance must precede the need for mechanical ventilation whenever possible. In the absence of contraindications, lung volume recruitment (i.e. air stacking) techniques should be introduced with the measurement of peak cough flows and maximum insufflation capacity in those with peak cough flows <270 L/min. Manually assisted coughing is recommended alone or in addition to lung volume recruitment to increase peak cough flows to >270 L/min. In the absence of contraindications, mechanical in-exsufflation should be recommended for patients unable to achieve peak cough flows >270 L/min with lung volume recruitment and/or manually assisted coughing, particularly during respiratory infection. A government-funded ventilatory service is necessary to provide appropriate access to equipment and respiratory care.

ABG: arterial blood gases, CO2: carbon dioxide, ETCO2: end tidal carbon dioxide, EPAP: expiratory positive airway pressure, FVC: Forced vital capacity, HMV: home mechanical ventilation, IPAP: inspiratory positive airway pressure, kPa: kilopascal, KQ: key question, LTOT: long term oxygen therapy, mmHg: millimeters of mercury (pressure), mmols: millimoles, NIPPV/NPPV: non-invasive positive pressure ventilation, NIV: non-invasive ventilation, NMD: neuromuscular disease, PCO2/PaCO2: partial pressure of arterial carbon dioxide, pH: potential of hydrogen, PSG: polysomnogram, SpO2: Blood oxygen saturation level, TcCO2/tCO2: transcutaneous carbon dioxide pressure, VAI: ventilator assisted individual, VC: vital capacity

Table G.2. Guidelines for COPD

Organization	Topic	KQ	Statement
German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010[75]	Device initiation criteria	KQ1	Symptoms that indicate CRF and reduced quality of life in COPD patients as well as one of the following criteria (at least 1 criterion must be fulfilled) indicate the need for HMV: Chronic daytime hypercapnia with PaCO2 ≥ 50mmHg Nocturnal hypercapnia with PaCO2 ≥ 55mmHg Stable daytime hypercapnia with 46–50mmHg and a rise in PTcCO2 to ≥ 10mmHg during sleep. Stable daytime hypercapnia with PaCO2 46–50mmHg and at least 2 acute exacerbations accompanied by respiratory acidosis that required hospitalization within the last 12 months Following an acute exacerbation needing ventilatory support, according to clinical estimation. Poor compliance with medication intake and/or LTOT are relative contraindications. Complete discontinuation of nicotine abuse should be aspired to. NIV is the primary treatment option for HMV of COPD patients with CRF. The most important criteria for the advent of long-term NIV are the presence of hypercapnia in combination with the typical symptoms of ventilatory failure, recurring exacerbations and the reduction in quality of life.
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device initiation criteria	KQ1	Nocturnal NIV is indicated in COPD with PaCO2 > 50 mmHg, where there is evidence of signs and symptoms of sleep disordered breathing, and full PSG demonstrates nocturnal hypoventilation (based on a measure of PaCO2) that is not corrected or made worse by LTOT alone.
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device initiation criteria	KQ1	The use of long-term NIPPV cannot be widely recommended in patients with stable COPD. Long-term NIPPV in COPD should only be considered on an individual basis. One subgroup of patients with COPD in which long-term NIPPV could be considered are those with severe hypercapnia (PaCO2 >55 mmHg) experiencing repeated episodes of acute hypercapnic respiratory failure that require in-hospital ventilatory support. However, definitive proof of efficacy of long-term NIPPV in these patients will need to await future studies. The overlap syndrome, and concomitant COPD and OSA syndrome, should be differentiated from chronic respiratory failure that is solely due to advanced COPD.
8th International Conference on Management and Rehabilitation of Chronic Respiratory Failure, Pescara, Italy, 2015[77]	Device initiation criteria	KQ1	The role of long-term non invasive positive pressure ventilation in improving survival in COPD patients with CRF (chronic respiratory failure) is still discussed. There is simply not enough evidence to support it. Long-term non invasive ventilation should be reserved to individual patients. Once stable hypercapnia is proven, NIPPV may improve survival and health status. Therefore, despite recent studies adding some new data, the authors cannot recommend the widespread use of this therapeutic intervention after an episode of acute-on-chronic respiratory failure in COPD. Long-term night non invasive ventilation in these patients has some physiological and clinical benefits.
Clinical Indications for Noninvasive Positive Pressure Ventilation in Chronic Respiratory Failure due to Restrictive Lung Disease, COPD, and Nocturnal Hypoventilation—A Consensus	Device initiation criteria	KQ1	Indications for usage: symptoms (such as fatigue, dyspnea, morning headache, etc.) and physiologic criteria (one of the following): PaCO2 > 55 mm Hg; PaCO2 of 50 to 54 mm Hg and nocturnal desaturation (oxygen saturation by pulse oximeter 88% for 5 continuous minutes while receiving oxygen therapy 2 L/ min); or PaCO2 of 50 to 54 mm Hg and hospitalization related to recurrent (2 in a 12- month period) episodes of hypercapnic respiratory failure

Organization	Topic	KQ	Statement
Conference Report, American Academy of Home Care Physicians, American College of Chest Physicians, American College of Physicians, American Sleep Disorders Association, American Thoracic Society, National Association for Medical Direction of Respiratory Care, 1999[74]			
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Device initiation criteria	KQ1	In acute hypercapnic respiratory failure in the hospital, NIV should be started when pH<7.35 and pCO2 >6.5 kPa persist or develop despite optimal medical therapy. In acute hypercapnic respiratory failure in the hospital, NIV can be discontinued when there has been normalization of pH and pCO2 and a general improvement in the patient's condition.
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device initiation, monitoring, and candidate selection	KQ1	Recurrent hospitalizations (2 or more in a year) for acute hypercapnic respiratory failure (especially life threatening events) or difficulty weaning from invasive ventilation are an indicator for assessment for domiciliary NIV.
Clinical Indications for Noninvasive Positive Pressure Ventilation in Chronic Respiratory Failure due to Restrictive Lung Disease, COPD, and Nocturnal Hypoventilation—A Consensus Conference Report, American Academy of Home Care Physicians, American College of Chest Physicians, American College of Physicians, American Sleep Disorders Association, American Thoracic Society, National Association for Medical Direction of Respiratory Care, 1999[74]	Device initiation, monitoring, and candidate selection	KQ1	Before considering a COPD patient for NIPPV, a physician with skills and experience in NIPPV must establish and document an appropriate diagnosis on the basis of history, physical examination, and results of diagnostic tests, and assure optimal management of COPD with such treatments as bronchodilators, oxygen when indicated, and optimal management of other underlying disorders (such as performing a multichannel sleep study to exclude associated sleep apnea if clinically indicated)
United States Department of Veterans Affairs, the Department of Defense, and the National Guideline	Device initiation, monitoring, and candidate selection	KQ1	In the absence of other contributors (e.g., sleep apnea), we suggest referral for a pulmonary consultation in patients with stable, confirmed COPD and hypercapnia.

Organization	Topic	KQ	Statement
Clearinghouse Clinical Practice Guideline for the Management of Chronic Obstructive Pulmonary Disease, 2014[78]			
United Kingdom National Institute for Health and Care Excellence (NICE), Chronic Obstructive Pulmonary Disease in Over 16s: Diagnosis and Management, 2010[79]	Device initiation, monitoring, and candidate selection	KQ1	Adequately treated patients with chronic hypercapnic ventilatory failure who have required assisted ventilation (whether invasive or non-invasive) during an exacerbation or who are hypercapnic or acidotic on LTOT should be referred to a specialist centre for consideration of long-term NIV. Patients with severe disease requiring interventions such as long-term non-invasive ventilation should be reviewed regularly by specialists.
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device continuation, compliance, and outcomes	KQ1 and KQ2	Changes in awake blood gases are not the best measure of effectiveness of NIV in chronic hypercapnic COPD. Changes in symptoms including exertional dyspnoea, control of nocturnal hypoventilation, reduction in hospital admissions and QoL (SF-36) are better indicators of the patient's response to therapy.
German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010[75]	Device characteristics and titration	KQ3	The aim of the ventilation is to normalize PaCO2; sufficiently high ventilation pressures are required to achieve this. Controlled ventilation mode with ventilation pressures from 20 to 40 mbar. Pressure escalation until normocapnia or maximum tolerance is reached. Rapid increase in inspiratory pressure (0.1 to 0.2 seconds) PEEP can be useful for assisted- or assisted-controlled ventilation. Minimal duration of therapy: 4.5 hours/day The introduction of NIV in the hospital can take up to two weeks.
Clinical Indications for Noninvasive Positive Pressure Ventilation in Chronic Respiratory Failure due to Restrictive Lung Disease, COPD, and Nocturnal Hypoventilation—A Consensus Conference Report, American Academy of Home Care Physicians, American College of Chest Physicians, American College of Physicians, American Sleep Disorders Association, American Thoracic Society, National Association for Medical Direction of Respiratory Care, 1999[74]	Device characteristics and titration	KQ3	NIPPV appears to be better tolerated in this patient population than negative pressure ventilation. In addition, advantages of ease of administration and portability as well as the ability to eliminate obstructive sleep apneas make NIPPV the noninvasive mode of first choice.
8th International Conference	Respiratory		Telemonitoring in ventilator dependent patients: 1) Home mechanical ventilators may be equipped with

Organization	Topic	KQ	Statement
on Management and Rehabilitation of Chronic Respiratory Failure, Pescara, Italy, 2015[77]	services		remote monitoring tools in order to improve physician supervision, with the aim to adapt settings to the needs and comfort of the patient. 2) Economic, regulatory and legal impacts of home telemonitoring will be important in its adaption by health care systems. 3) Relevant issues are prescription criteria, modalities of follow-up, team expertise, technologies, adherence, bundling of services, and outcomes

COPD: chronic obstructive pulmonary disease, CRF: chronic respiratory failure, HMV: home mechanical ventilation, kPa: kilopascal, KQ: key question, LTOT: long term oxygen therapy, NIPPV/NPPV: non-invasive positive pressure ventilation, NIV: non-invasive ventilation, OSA: obstructive sleep apnea, PCO2/PaCO2: partial pressure of arterial carbon dioxide, PEEP: positive end expiratory pressure, pH: potential of hydrogen, PSG: polysomnogram, QoL: quality of life, SF-36: Medical Outcomes Study Questionnaire Short Form, tCO2/ PTcCO2: transcutaneous carbon dioxide pressure

Table G.3. Guidelines for Neuromuscular Disease

Organization	Topic	KQ	Statement
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device initiation criteria	KQ1	The institution of NIV is recommended in patients with rapidly progressive respiratory muscle weakness associated with orthopnoea, hypercapnia or symptomatic sleep hypoventilation (sleep fragmentation/ daytime hypersomnolence/ morning headaches and cognitive dysfunction).
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Device initiation criteria	KQ1	Planned elective domiciliary NIV is preferable to crisis management in NMD and chest wall disorders. This reduces the risk of acute presentation and provides a proven alternative to invasive mechanical ventilation which risks prolonged or permanent tracheostomy ventilation. NIV should almost always be trialled in the acutely unwell patients with NMD or CWD with hypercapnia. Do not wait for acidosis to develop. In patients with NMD or CWD, NIV should be considered in acute illness when vital capacity (VC) is known to be <1 L and RR >20, even if normocapnic. In patients with NMD or CWD, nocturnal NIV should usually be continued following an episode of AHRF, pending discussion with a home ventilation service.
United Kingdom National Institute for Health and Care Excellence (NICE), Motor Neuron Disease: Assessment and Management, 2016[80]	Device initiation criteria	KQ1	If the person's SpO2 (measured at rest and breathing room air) is greater than 94%, or 92% for those with lung disease, but they have sleep-related respiratory symptoms: Consider referring them to a respiratory ventilation service for continuous nocturnal (overnight) oximetry and/or a limited sleep study and discuss both the impact of respiratory impairment and treatment options with the patient and (if the person agrees) their family and carers. If the person's arterial partial pressure of carbon dioxide (PaCO2) is greater than 6 kPa: refer them urgently to a respiratory ventilation service (to be seen within 1 week) and explain the reasons for and implications of the urgent referral to the person and (if the person agrees) their family and carers. If the person's PaCO2 is less than or equal to 6 kPa but they have any symptoms or signs of respiratory impairment, particularly orthopnoea refer them to a respiratory ventilation service for nocturnal (overnight) oximetry and/or a limited sleep study and discuss both the impact of respiratory impairment and treatment options with the person and (if the person agrees) their family and/or carers (as appropriate). If any of the results listed in box 2 is obtained, discuss with the person and (if appropriate) their family and carers: their respiratory impairment their treatment options possible referral to a respiratory ventilation service for further assessment based on discussion with the person, and their wishes.

Organization	Topic	KQ	Statement
Clinical Indications for Noninvasive Positive Pressure Ventilation in Chronic Respiratory Failure due to Restrictive Lung Disease, COPD, and Nocturnal Hypoventilation—A Consensus Conference Report, American Academy of Home Care Physicians, American College of Chest Physicians, American College of Physicians, American Sleep Disorders Association, American Thoracic Society, National Association for Medical Direction of Respiratory Care, 1999[74]	Device initiation criteria	KQ1	Indications for usage Symptoms (such as fatigue, dyspnea, morning headache, etc.) and one of the following Physiologic criteria (one of the following PaCO2 ≥ 45 mm Hg Nocturnal oximetry demonstrating oxygen saturation ≤ 88% for 5 consecutive minutes For progressive neuromuscular disease, maximal inspiratory pressures < 60 cm H2O or FVC <50% predicted
German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010[75]	Device initiation criteria	KQ1	NIV of NMD patients with clinical signs of CRF is indicated by the following (at least 1 criterion should be fulfilled): Chronic daytime hypercapnia with PaCO2 ≥ 45mmHg Nocturnal hypercapnia with PaCO2 ≥ 50mmHg Daytime normocapnia with a rise in PTcCO2 of ≥ 10mmHg during the night A rapid, significant reduction in VC At the first signs of nocturnal hypercapnia, the patient should be offered NIV therapy rather than waiting until the hypercapnia extends into the daytime period. There are no indications for prophylactic mechanical ventilation in the absence of symptoms or hypoventilation. NIV is also indicated prior to elective vertebral column correction surgery when VC < 60% target value and FEV1 < 40% target value, respectively, or during pregnancy with restricted lung function, as well as palliative care of dyspnea. Patients with NMD should undergo clinical assessment and assessment of VC at 3–12 month-intervals. Polygraphy and PTcCO2-measurement are indicated when VC is < 70%. NIV is the primary treatment option for HMV of NMD patients with CRF; in cases of inviability, failure or rejection of NIV, invasive HMV should only be established in accordance with the explicit wishes of the patient and custodian, respectively. The most important criteria for the initiation of NIV are hypercapnia in combination with the characteristic symptoms of ventilatory failure, and a reduction in quality of life.
American Academy of Neurology, Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: multidisciplinary care, symptom management, and cognitive/behavioral impairment (an evidence-based review), 2009[81]	Device initiation Criteria (ALS)	KQ1	NIV may be considered at the earliest sign of nocturnal hypoventilation or respiratory insufficiency in order to improve compliance with NIV in patients with ALS. NIV may be considered to enhance QOL in patients with ALS who have respiratory insufficiency.

Organization	Topic	KQ	Statement
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device initiation Criteria (ALS)	KQ1	NIV should be offered to patients with any one of the following: Orthopnea Daytime hypercapnia Symptomatic sleep disordered breathing FVC <50% predicted SNP <40 cmH2O or PImax<40 cmH2O
American Thoracic Society, Respiratory Care of the Patient with Duchenne Muscular Dystrophy, 2004[82]	Device initiation criteria (Duchenne Muscular Dystrophy)	KQ1	Consider daytime ventilation when measured waking Pco2 exceeds 50 mm Hg or when hemoglobin saturation remains < 92% while awake.
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device initiation criteria (Duchenne Muscular Dystrophy)		Offer nocturnal NIV to patients with diurnal hypercapnia (daytime arterial PCO2 >45 mmHg), or when there is documented nocturnal hypercapnia and the presence of symptoms consistent with hypoventilation. Institution of NIV during sleep should be offered to patients demonstrating a major degree of nocturnal hypoxemia, even if asymptomatic.
German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010[75]	Device initiation, monitoring, and candidate selection	KQ1	Specific aspects in the ventilation of patients with NMD comprise: Muscle weakness in the oropharyngeal area, carrying the risk of reduced ability or complete inability to close the mouth Bulbar symptoms with the risk of recurrent aspiration Hypersalivation; therapy with anti-cholinergics (e. g. Scopolamine patch, amitryptiline or botulinum toxin injections into the salivary glands) Coughing weakness, with the development of acute decompensation
British Thoracic Society, Guidelines for Home Oxygen Use in Adults, 2015[73]	Device initiation, monitoring, and candidate selection	KQ1	Non-invasive ventilation (NIV) should be the treatment of choice for patients with NMD or chest wall disease causing type 2 respiratory failure.
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Device initiation, monitoring, and candidate selection	KQ1	In patients with NMD or CWD, senior/experienced input is needed in care planning and is essential if differences in opinion exist or develop between medical staff and patient representatives. In patients with NMD, it should be anticipated that bulbar dysfunction and communication difficulties, if present, will make NIV delivery difficult, and may make it impossible. Discussion about NIV and IMV, and patients' wishes with respect to cardiopulmonary resuscitation, should occur as part of routine care of patients with NMD or CWD. In patients with NMD or chest wall diseases, senior staff should be involved in decision-making, in conjunction with home mechanical ventilation specialists, if experience is limited, and especially when the appropriateness of invasive mechanical ventilation is questioned. Domiciliary NIV is effective in treating chronic hypercapnia, improves long-term survival and preserves a good or acceptable QoL

Organization	Topic	KQ	Statement
United Kingdom National Institute for Health and Care Excellence (NICE), Motor Neuron Disease: Assessment and Management, 2016[80]	Device initiation, monitoring, and candidate selection	KQ1	Assess and monitor the person's respiratory function and symptoms. Treat people with NMD and worsening respiratory impairment for reversible causes (for example, respiratory tract infections or secretion problems) before considering other treatments. Offer non-invasive ventilation as treatment for people with respiratory impairment. Decisions to offer non-invasive ventilation should be made by the multidisciplinary team in conjunction with the respiratory ventilation service, and the person. Consider urgent introduction of non-invasive ventilation for people with NMD who develop worsening respiratory impairment and are not already using non-invasive ventilation. As part of the initial assessment to diagnose NMD, or soon after diagnosis, a healthcare professional from the multidisciplinary team who has appropriate competencies should perform the following tests (or arrange for them to be performed) to establish the person's baseline respiratory function: oxygen saturation measured by pulse oximetry (SpO2): this should be a single measurement of SpO2 with the person at rest and breathing room air if it is not possible to perform pulse oximetry locally, refer the person to a respiratory ventilation service. Then one or both of the following: forced vital capacity (FVC) or vital capacity (VC) sniff nasal inspiratory pressure (SNIP) and/or maximal inspiratory pressure (MIP). If the person has severe bulbar impairment or severe cognitive problems that may be related to respiratory impairment: ensure that SpO2 is measured (at rest and breathing room air) do not perform the other respiratory function tests (FVC, VC, SNIP and MIP) if interfaces are not suitable for the person. A healthcare professional with appropriate competencies should perform the respiratory function tests every 2–3 months, although tests may be performed more or less often depending on: whether there are any symptoms and signs of respiratory impairment (see box 1) the rate of progression of NMD the person's preference and circumstances. [2010, amended 2016] Perform arterial or capillary blood gas analysis if the person's SpO2 (measured at rest and breathing room air): is less than or equal to 92% if they have known lung disease is less than or equal to 94% if they do not have lung disease. If it is not possible to perform arterial or capillary blood gas analysis locally, refer the person to a respiratory ventilation service.

Organization	Topic	KQ	Statement
Clinical Indications for Noninvasive Positive Pressure Ventilation in Chronic Respiratory Failure due to Restrictive Lung Disease, COPD, and Nocturnal Hypoventilation—A Consensus Conference Report, American Academy of Home Care Physicians, American College of Chest Physicians, American College of Physicians, American Sleep Disorders Association, American Thoracic Society, National Association for Medical Direction of Respiratory Care, 1999[74]	Device initiation, monitoring, and candidate selection	KQ1	Disease documentation Before considering a restrictive thoracic patient for NIPPV, a physician with skills and experience in NIPPV must establish and document an appropriate diagnosis on the basis of history, physical examination, and diagnostic tests and assure optimal treatment of other underlying disorders (such as performing a multichannel sleep study to detect associated sleep apnea if clinically indicated) The most common disorders would include sequelae of polio, spinal cord injury, neuropathies, myopathies and dystrophies, ALS, chest wall deformities, and kyphoscoliosis.
British Thoracic Society, Guidelines for Home Oxygen Use in Adults, 2015[73]	Device initiation, monitoring, and candidate selection	KQ1	Patients with neuromuscular weakness affecting respiratory muscles should not have nocturnal oxygen therapy alone ordered. It can be considered in patients with evidence of established ventilatory failure, where it should be given with NIV support.

Organization	Topic	KQ	Statement
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device initiation, monitoring, and candidate selection	KQ1	Subjects with progressive respiratory muscle weakness and other restrictive thoracic disorders should be observed regularly with lung function (VC, MIP, MEP, SNP and PCF) and oximetry. An arterial blood gas should be performed especially if VC < 40% predicted or MIP < 60 cmH2O. <u>Slowly progressive NMD</u> Hypoxia, hypercapnia, or an elevation in serum bicarbonate indicate the need for additional respiratory assessments and interventions. All subjects with DMD should be referred for clinical assessment initially to a paediatric specialist unit for assessment and then care transferred to an adult centre when age >18 years. Assessment as to the risk of development of progressive respiratory failure should be considered in all subjects with other progressive neuromuscular disorders. Referral to a specialist centre should occur if significant respiratory muscle weakness or sleep disordered breathing occurs. Patients should have access to other specialist health providers, including medical specialists and allied health professionals, preferably in a well co-ordinated multidisciplinary team. <u>Rapidly progressive NMD</u> Patients with NMD are recommended to have 3 monthly clinical evaluation to monitor for symptoms and signs of respiratory and sleep complications. Sniff nasal inspiratory pressure and overnight oximetry are the initial investigations of choice for the assessment of early respiratory muscle compromise and nocturnal hypoventilation. A diagnostic polysomnogram should be reserved for patients in whom co-existent upper airway obstruction is suspected on clinical grounds with inconclusive nocturnal oximetry. While NMD patients with significant bulbar dysfunction should still have the option to trial NIV, it should be recognized that this group of patients may have reduced tolerance to and derive less benefit from NIV. The progression to tracheostomy intermittent positive pressure ventilation (TIPPV) should be made on an individual basis, weighing the longer survival advantage with a significantly greater burden of care and cost to the patient, carer and/or community and recognizing that QoL improvements associated with the use of NIV may not be seen with TIPPV to an equivalent degree. The elective commencement of NIV is preferred over non-elective TIPPV despite the improved survival advantage. Patients with NMD should be managed in a multidisciplinary clinic as this improves survival and QoL, and facilitates earlier uptake of interventions including NIV and PEG insertion.

Organization	Topic	KQ	Statement
European Federation of Neurological Societies (EFNS) Guidelines on the Clinical Management of Amyotrophic Lateral Sclerosis, 2012[83]	Device initiation, monitoring, and candidate selection (ALS)	KQ1	Symptoms or signs of respiratory insufficiency (including symptoms of nocturnal hypoventilation) should be checked at each visit. Forced vital capacity and vital capacity are the most available and practical tests for the regular monitoring of respiratory function. Sniff nasal pressure may be used for monitoring, particularly in bulbar patients with weak lips. Percutaneous nocturnal oximetry is recommended as a screening test and for monitoring respiratory function. Symptoms or signs of respiratory insufficiency should prompt discussions with the patient and caregivers about treatment options and the terminal phase. Early discussions are needed to allow advance planning and directives. NIPPV should be considered in preference to IMV in patients with symptoms or signs of respiratory insufficiency. NIPPV can prolong survival for many months and may improve the patient's quality of life IMV has a major impact upon caregivers and should be initiated only after informed discussion. Unplanned (emergency) IMV should be avoided through an early discussion of end-of-life issues, coordination with palliative care teams and appropriate advance directives. Oxygen therapy alone should be avoided as it may exacerbate carbon dioxide retention and oral dryness. Use oxygen only if symptomatic hypoxia is present.
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device initiation, monitoring, and candidate selection (ALS)	KQ1	Regular monitoring of ALS patients is advised from the time of diagnosis every two to six months and varies with anticipated rapidity of disease progression and should include the following: Symptom review to include orthopnea, dyspnea, poor sleep, excessive daytime sleepiness, poor concentration, morning headache. Measurement of sitting FVC. Measurement of one or more of the following: supine VC, sniff nasal pressure, Pimax (MIP). Measurement of ABGs or end tidal CO2 (ETCO2) when hypercapnia is suspected. Nocturnal oximetry ± transcutaneous CO2 (tCO2) when symptomatic sleep disordered breathing is suspected. Measurement of peak cough flow. NIV should be considered the preferred option for ventilation even when ventilation is required 24 h per day. Elective tracheostomy ventilation may be considered, and is dependent on regional resources and careful discussion with the patient and caregivers. Long-term invasive ventilation can be offered after acute respiratory failure requiring invasive ventilation, if the patient and caregivers fully understand the consequences and appropriate support is available.

Organization	Topic	KQ	Statement
American Thoracic Society, Respiratory Care of the Patient with Duchenne Muscular Dystrophy, 2004[82]	Device initiation, monitoring, and candidate selection (Duchenne Muscular Dystrophy)	KQ1	In centers with appropriate expertise, consider mouthpiece intermittent positive pressure ventilation or other forms of noninvasive daytime ventilation. Consider tracheostomy when contraindications or patient aversion to noninvasive ventilation are present. Patients receiving noninvasive ventilation should have regular (at least annual) noninvasive monitoring of gas exchange, including oxygen saturation and end-tidal Pco2 levels. Discussions regarding ventilatory support for each patient should involve the patient, caregivers, and medical team. Where CO2 monitoring is not available, overnight pulse oximetry can be used to detect nighttime oxyhemoglobin desaturation. Simple oximetry provides, at best, only indirect information on ventilation, and should be used to assess need for ventilatory support only when better alternatives are unavailable. Schedule periodic reassessment as appropriate to stage of disease. Follow-up visits should include monitoring for the development of daytime hypoventilation, which may necessitate around-the-clock ventilation. Use nasal intermittent positive pressure ventilation to treat sleep-related upper airway obstruction and chronic respiratory insufficiency in patients with DMD. Negative-pressure ventilators should be used with caution in patients with DMD due to the risk of precipitating upper airway obstruction and hypoxemia. Do not use oxygen to treat sleep-related hypoventilation without ventilatory assistance. Objective evaluation at each clinic visit should include: oxyhemoglobin saturation by pulse oximetry, spirometric measurements of FVC, FEV1, and maximal mid-expiratory flow rate, maximum inspiratory and expiratory pressures, and peak cough flow. Awake carbon dioxide tension should be evaluated at least annually in conjunction with spirometry. Where available, capnography is ideal for this purpose. Arterial blood gas analysis is not necessary for routine follow-up of patients with DMD. If capnography is not available, then a venous or capillary blood sample should be obtained to assess for the presence of alveolar hypoventilation. Additional measures of pulmonary function and gas exchange may be useful, including lung volumes, assisted cough peak flow, and maximum insufflation capacity. Carefully evaluate patients for evidence of other respiratory disorders, such as obstructive sleep apnea, oropharyngeal aspiration, gastroesophageal reflux, and asthma. Annual laboratory studies in patients requiring a wheelchair for ambulation should include a complete blood count, serum bicarbonate concentration, and a chest radiograph.
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device initiation, monitoring, and candidate selection (Duchenne Muscular Dystrophy)	KQ1	Carefully question and educate patients to report symptoms consistent with hypoventilation, including disturbed sleep, excessive daytime sleepiness, morning headache and weight loss. -Measure VC, MIP, maximal expiratory pressure, peak cough flow and awake oxyhemoglobin saturation by pulse oximetry at least yearly; if VC <40% predicted, also monitor awake CO2 tension by noninvasive methods or ABG analysis. Perform an evaluation of ventilation during sleep if there are symptoms consistent with nocturnal hypoventilation or other forms of sleep disordered breathing. In the absence of such symptoms, periodic screening for sleep disordered breathing should also be considered once FEV1 or FVC is <40% predicted.

Organization	Topic	KQ	Statement
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device initiation, monitoring, and candidate selection (Other myopathies)	KQ1	Obtain periodic clinical assessment and spirometry at six- to 12-month intervals, including sitting (plus supine if diaphragmatic weakness is suspected) spirometric testing. Consider monitoring for sleep disordered breathing in patients with VC <60%. Consider ABGs or nocturnal measure of CO2 in patients with VC <40% to exclude hypercapnia. NIV should be offered when there is daytime hypercapnia or symptomatic nocturnal hypoventilation. Assess airway clearance ability with peak cough flows and implement cough-assistance strategies
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device initiation, monitoring, and candidate selection (Myotonic dystrophy)	KQ1	Obtain six to 12 monthly clinical assessment of symptoms of daytime or nocturnal hypoventilation. Obtain yearly VC and consider daytime *Pa*CO2 measurement, even with mild reductions of VC when patients exhibit symptoms of hypoventilation. Consider overnight oximetry or polysomnography when there are symptoms of nocturnal hypoventilation. Long-term NIV should be offered to patients with daytime hypercapnia or symptomatic nocturnal hypoventilation as for other NMDs Carefully assess motivation and ability to adhere to treatment with patients and their caregivers before initiating long-term ventilatory support. Reassess every six months to verify treatment adherence and provide extra help and motivation as needed.
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device initiation, monitoring, and candidate selection (Post-polio syndrome)	KQ1	Yearly assessment of VC is recommended from the time of presentation of post polio syndrome. If VC >50% with symptoms of hypoventilation, perform measurements of daytime ABGs, overnight oximetry and consider polysomnography. When VC <50%, perform ABG analysis and/or nocturnal oximetry yearly. With confirmation of the presence of chronic hypoventilation, offer NIV.
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device initiation, monitoring, and candidate selection (Spinal cord injury)	KQ1	Each patient must be individually evaluated for the need for long-term ventilation either acutely or in follow-up. Noninvasive support is preferable to invasive ventilation. Phrenic nerve pacing is recommended in selected individuals as an alternative to positive pressure ventilation alone. In the long term, individuals with SCI require regular monitoring to identify the development of sleep disordered breathing or respiratory failure and evaluate the need for NIV.
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device initiation, monitoring, and candidate selection (Spinal cord injury)	KQ1	NIV is indicated when there is intractable or refractory sputum retention, atelectasis, respiratory tract infection or type-I respiratory failure (PaO2 < 80 mmHg, SpO2 <95%). NIV is indicated when there is intolerance of CPAP for treatment of OSA, especially in cases of SCI at C6 or above. Use of an abdominal binder may be considered as the initial intervention in cases of mild hypoventilation, or as an adjunct to the use of NIV. The implementation of NIV should occur in a specialised centre where there is access to a spinal unit, accredited pulmonary function and sleep laboratory, physician experienced in the use of NIV, NIV service and physiotherapy service trained in secretion removal in patients with spinal cord injury.

Organization	Topic	KQ	Statement
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device characteristics and titration (Duchenne Muscular Dystrophy)	KQ3	When bilevel ventilation is used, backup respiratory rates are recommended during sleep while on NIV to reduce the work of breathing associated with breath initiation. Individualize the decision about the transition from nocturnal NIV to daytime ventilation by carefully evaluating patient factors (symptoms, bulbar involvement, patient preference, etc.) and available resources. In patients requiring daytime ventilation, strongly consider mouthpiece ventilation as an alternative to invasive tracheostomy.
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Device characteristics and titration	KQ3	In patients with NMD or CWD, consider controlled ventilation as triggering may be ineffective.
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device characteristics and titration (ALS)	KQ3	Ventilator settings should be adjusted for optimal patient comfort and improvement of symptoms. ABGs and/or nocturnal oximetry and/or polysomnography are not required, but may be helpful in some circumstances. When bilevel pressure ventilators are used for NIV, a backup rate is recommended.
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Respiratory services	KQ4	Ability to generate PCF of at least 160 L/min is necessary for non-invasive management of pulmonary secretions. Baseline assisted PCF <270 L/min are likely to decrease to <160 L/min during chest infections, increasing the likelihood of pneumonia and respiratory failure. Patients with a baseline PCF < 270 L/min should have access to equipment which can provide insufflation and a mechanical cough in-exsufflation. Training of insufflation should commence when VC < 2L or 50% predicted. As manual assisted coughing techniques (e.g. abdominal thrust) further enhance PCF, they should be incorporated with insufflation or mechanical in-exsufflation techniques, where possible. For patients with VC < 1 to 1.5L, insufflations should precede manual assisted coughing techniques (e.g. abdominal thrusts). In adults, mechanical in-exsufflation settings of +40 cmH2O and – 40 cmH2O appear to safely provide adequate PCF for the majority of patients with neuromuscular disease. Mechanical in-exsufflation can be ineffective in patients with very poor bulbar dysfunction with insufflation capacity >1L, where dynamic airway collapse occurs. Techniques of insufflation, manual assisted coughing and mechanical in-exsufflation require substantial acclimatisation and should be trained when the patient is well and ideally prior to an acute infective requirement.
German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010[75]	Respiratory services	KQ4	A reduced cough impulse (peak cough flow; PCF < 270 l/min) can lead to acute decompensations and increased incidence of aspiration pneumonia. Measures to eliminate secretions should therefore be taken when SaO2< 95%, or a 2–3% drop in the patient's individual best value occurs. Step-based secretion management consists of measures to increase intrapulmonary volume via air stacking, frog breathing or manual hyperinflation, as well as assisted coughing techniques or mechanical cough assistants (CoughAssist ®, Pegaso Cough®) The measurement of coughing capacity in NMD patients is obligatory. Coughing weakness (PCF < 270 l/min) indicates the need for the initiation of secretion management.

Organization	Topic	KQ	Statement
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Respiratory services	KQ4	In patients with neuromuscular disease (NMD), mechanical insufflation and exsufflation should be used, in addition to standard physiotherapy techniques, when cough is ineffective and there is sputum retention.
American Academy of Neurology, Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: multidisciplinary care, symptom management, and cognitive/behavioral impairment (an evidence-based review), 2009[81]	Respiratory services	KQ4	Mechanical insufflation/exsufflation) may be considered to clear secretions in patients with ALS who have reduced peak cough flow, particularly during an acute chest infection. There are insufficient data to support or refute high frequency chest wall oscillation for clearing airway secretions in patients with ALS.
United Kingdom National Institute for Health and Care Excellence (NICE), Motor Neuron Disease: Assessment and Management, 2016[80]	Respiratory services	KQ4	Offer cough augmentation techniques such as manual assisted cough to people with NMD who cannot cough effectively. Consider unassisted breath stacking and/or manual assisted cough as the first-line treatment for people with NMD who have an ineffective cough For patients with bulbar dysfunction, or whose cough is ineffective with unassisted breath stacking, consider assisted breath stacking (for example, using a lung volume recruitment bag). Consider a mechanical cough assist device if assisted breath stacking is not effective, and/or during a respiratory tract infection. Consider opioids as an option to relieve symptoms of breathlessness. Take into account the route of administration and acquisition cost of medicines. Consider benzodiazepines to manage breathlessness that is exacerbated by anxiety. Take into account the route of administration and acquisition cost of medicines.
Canadian Thoracic Society 2011[72]	Respiratory services (ALS)	KQ4	Lung volume recruitment maneuvers should be introduced with declining VC. Methods to assist secretion clearance should be initiated when PCF is <4.25 L/s or the Norris bulbar core is <29.

Organization	Topic	KQ	Statement
European Federation of Neurological Societies (EFNS) Guidelines on the Clinical Management of Amyotrophic Lateral Sclerosis, 2012[83]	Respiratory services (ALS)	KQ4	Active management of secretions and provision of cough-assist devices can increase the effectiveness of assisted ventilation in ALS. For bronchial secretions: A mucolytic including N-acetylcysteine, 200–400 mg three times daily, may be beneficial. Beta-receptor antagonists and a nebulizer with saline and/or an anticholinergic bronchodilator and/or a mucolytic and/or furosemide may be used in combination. Mucolytics should only be used if sufficient cough flow is present. The patient and carer should be taught the technique of assisting expiratory movements using a manual-assisted cough (can also be performed by a physical therapist). The use of a mechanical insufflator–exsufflator may be helpful, particularly in the setting of an acute respiratory infection. A portable home suction device and a room humidifier may be of use. The medical treatment of intermittent dyspnoea should involve: a for short dyspnoeic bouts: relieve anxiety and give lorazepam 0.5–2.5 mg sublingually; b for longer phases of dyspnoea (>30 minutes): give morphine 2.5 mg orally or s.c. For the medical treatment of chronic dyspnoea, start with morphine 2.5 mg orally four to six times daily. For severe dyspnoea, give morphine s.c. or as an i.v. infusion. Start with 0.5 mg/h and titrate. If needed, add midazolam (2.5–5 mg) or diazepam for nocturnal symptom control and to relieve anxiety.
American Thoracic Society, Respiratory Care of the Patient with Duchenne Muscular Dystrophy, 2004[82]	Respiratory services (Duchenne Muscular Dystrophy)	KQ4	Patients with DMD should be taught strategies to improve airway clearance and how to employ those techniques early and aggressively. Use assisted cough technologies in patients whose clinical history suggests difficulty in airway clearance, or whose peak cough flow is less than 270 L/minute and/or whose maximal expiratory pressures are less than 60 cm H2O. The committee strongly supports use of mechanical insufflation-exsufflation in patients with DMD and also recommends further studies of this modality. Home pulse oximetry is useful to monitor the effectiveness of airway clearance during respiratory illnesses and to identify patients with DMD needing hospitalization Individuals who require mechanically assisted airway clearance therapy or mechanically assisted ventilation should see a pulmonologist every 3 to 6 months or as indicated for routine follow-up.
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Respiratory services (Duchenne Muscular Dystrophy)	KQ4	Lung volume recruitment maneuvers should be introduced with declining VC. -Methods to assist secretion clearance should be initiated when PCF <270 L/min.
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Respiratory services (All NMD except ALS and Duchenne Muscular Dystrophy)	KQ4	Assess airway clearance ability with peak cough flows and implement cough-assistance strategies

Organization	Topic	KQ	Statement
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Respiratory services (Spinal cord injury)	KQ4	Regular airway clearance techniques (lung volume recruitment, manually assisted coughing, and mechanical in-exsufflation), clinical assessment and ongoing monitoring of pulmonary function is recommended to ensure adequate airway clearance.

ABG: arterial blood gases, AHRF: acute hypercapnic respiratory failure, ALS: amyotrophic lateral sclerosis, cmH2O: centimeters of water (pressure), CO2: carbon dioxide, CPAP: continuous positive airway pressure, CRF: chronic respiratory failure, CWD: chest wall deformity, DMD: Duchenne muscular dystrophy, ETCO2: end tidal carbon dioxide, FEV1: Forced expiratory volume in one second, FVC: Forced vital capacity, HMV: home mechanical ventilation, IMV: invasive mechanical ventilation, kPa: kilopascal, KQ: key question, MEP: maximal expiratory pressure, mg: milligram, MIP: maximal inspiratory pressure, mmHg: millimeters of mercury (pressure), NIPPV/NPPV: non-invasive positive pressure ventilation, NIV: non-invasive ventilation, NMD: neuromuscular disease, OSA: obstructive sleep apnea, PaO2: partial pressure of arterial oxygen, PCO2/PaCO2: partial pressure of arterial carbon dioxide, PCF: peak cough flow, PEG: Polyethylene glycol, PImax: Maximal inspiratory mouth pressures, QOL: quality of life, RR: respiratory rate, s.c: subcutaneous, SCI: spinal cord injury, SNP/SNIP: sniff nasal inspiratory pressure, SpO2: Blood oxygen saturation level, tCO2/PTcCO2: transcutaneous carbon dioxide, TIPPV: tracheostomy intermittent positive pressure ventilation, VC: vital capacity

Table G.4. Guidelines for Thoracic Restrictive Disorders

Organization	Topic	KQ	Statement
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Device initiation criteria	KQ1	Planned elective domiciliary NIV is preferable to crisis management in NMD and chest wall disorders. This reduces the risk of acute presentation and provides a proven alternative to invasive mechanical ventilation which risks prolonged or permanent tracheostomy ventilation. NIV should almost always be trialled in the acutely unwell patients with NMD or CWD with hypercapnia. Do not wait for acidosis to develop. In patients with NMD or CWD, NIV should be considered in acute illness when vital capacity (VC) is known to be <1 L and RR >20, even if normocapnic. In patients with NMD or CWD, nocturnal NIV should usually be continued following an episode of AHRF, pending discussion with a home ventilation service.
Clinical Indications for Noninvasive Positive Pressure Ventilation in Chronic Respiratory Failure due to Restrictive Lung Disease, COPD, and Nocturnal Hypoventilation—A Consensus Conference Report, American Academy of Home Care Physicians, American College of Chest Physicians, American College of Physicians, American Sleep Disorders Association, American Thoracic Society, National Association for Medical Direction of Respiratory Care, 1999[74]	Device initiation criteria	KQ1	Indications for usage Symptoms (such as fatigue, dyspnea, morning headache, etc.) and one of the following Physiologic criteria (one of the following PaCO2 ≥ 45 mm Hg Nocturnal oximetry demonstrating oxygen saturation ≤ 88% for 5 consecutive minutes For progressive neuromuscular disease, maximal inspiratory pressures < 60 cmH2O or FVC <50% predicted
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device initiation criteria	KQ1	Patients with kyphoscoliosis should undergo periodical spirometry testing, and if FVC is <50%, ongoing review, assessing for evidence of hypercapnic respiratory failure should be instituted. Long-term nocturnal NIV should be offered to all patients with kyphoscoliosis who have developed chronic hypercapnic respiratory failure. Patients with hypoxemia but without hypercapnia may be managed cautiously with oxygen therapy alone while monitoring for development of hypercapnia.
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device initiation criteria	KQ1	NIV in patients with respiratory insufficiency from chest wall disease provides greater physiological and symptomatic relief over oxygen alone. NIV should be trialled in all patients with chest wall disorders with evidence of nocturnal hypoventilation.

Organization	Topic	KQ	Statement
German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010[75]	Device initiation criteria	KQ1	The following indication criteria are valid when symptoms of CRF and a reduced quality of life are present (at least 1 criterion must be fulfilled): Chronic daytime hypercapnia with $PaCO_2 \geq 45$mmHg Nocturnal hypercapnia with $PaCO_2 \geq 50$mmHg Daytime normocapnia with a rise in $PTcCO_2$ of ≥ 10mmHg during the night Patients without manifest hypercapnia but with severe, restrictive ventilatory dysfunction (VC < 50% predicted), must undergo a short-term (within 3 months) clinical control examination including polygraphy. NIV is the primary treatment option for HMV of restrictive thoracic disease patients with CRF. The most important criteria for the advent of long-term NIV are hypercapnia in combination with the typical symptoms of ventilatory insufficiency, and the reduction in quality of life. For symptoms of hypoventilation in the absence of hypercapnia, a somnological examination should take place. Patients with severe, restrictive ventilatory dysfunction in the absence of manifest hypercapnia must be closely monitored.
British Thoracic Society, Guidelines for Home Oxygen Use in Adults, 2015[73]	Device initiation, monitoring, and candidate selection	KQ1	Non-invasive ventilation (NIV) should be the treatment of choice for patients with NMD or chest wall disease causing type 2 respiratory failure.
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Device initiation, monitoring, and candidate selection	KQ1	In patients with NMD or CWD, senior/experienced input is needed in care planning and is essential if differences in opinion exist or develop between medical staff and patient representatives. In patients with NMD, it should be anticipated that bulbar dysfunction and communication difficulties, if present, will make NIV delivery difficult, and may make it impossible. Discussion about NIV and IMV, and patients' wishes with respect to cardiopulmonary resuscitation, should occur as part of routine care of patients with NMD or CWD. In patients with NMD or chest wall diseases, senior staff should be involved in decision-making, in conjunction with home mechanical ventilation specialists, if experience is limited, and especially when the appropriateness of invasive mechanical ventilation is questioned. Domiciliary NIV is effective in treating chronic hypercapnia, improves long-term survival and preserves a good or acceptable QoL

Organization	Topic	KQ	Statement
Clinical Indications for Noninvasive Positive Pressure Ventilation in Chronic Respiratory Failure due to Restrictive Lung Disease, COPD, and Nocturnal Hypoventilation—A Consensus Conference Report, American Academy of Home Care Physicians, American College of Chest Physicians, American College of Physicians, American Sleep Disorders Association, American Thoracic Society, National Association for Medical Direction of Respiratory Care, 1999[74]	Device initiation, monitoring, and candidate selection	KQ1	Disease documentation Before considering a restrictive thoracic patient for NIPPV, a physician with skills and experience in NIPPV must establish and document an appropriate diagnosis on the basis of history, physical examination, and diagnostic tests and assure optimal treatment of other underlying disorders (such as performing a multichannel sleep study to detect associated sleep apnea if clinically indicated) The most common disorders would include sequelae of polio, spinal cord injury, neuropathies, myopathies and dystrophies, ALS, chest wall deformities, and kyphoscoliosis.
British Thoracic Society, Guidelines for Home Oxygen Use in Adults, 2015[73]	Device initiation, monitoring, and candidate selection	KQ1	NIV should be the treatment of choice for patients with chest wall or neuromuscular disease causing type 2 respiratory failure. Additional LTOT (long term oxygen therapy) may be required in case of hypoxaemia not corrected with NIV.
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Device characteristics and titration	KQ3	In patients with NMD or CWD, consider controlled ventilation as triggering may be ineffective.
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device characteristics and titration	KQ3	Both pressure and volume preset ventilation is likely to be equally effective in chest wall disease, but there is a subset of patients which may demonstrate the need for volume ventilation if adequately titrated pressure preset fails to significantly improve diurnal hypercapnia.
German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010[75]	Device characteristics and titration	KQ3	NIV in pressure- and volume-limited modes is feasible. With set pressure, maximal ventilation pressure often reaches 20–25 mbar. Changeover from set pressure to set volume should be taken into account in order to improve ventilation. EPAP is generally not necessary if bronchial obstructions are absent.

Organization	Topic	KQ	Statement
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Respiratory services	KQ4	In patients with neuromuscular disease (NMD), mechanical insufflation and exsufflation should be used, in addition to standard physiotherapy techniques, when cough is ineffective and there is sputum retention.
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Respiratory services	KQ4	Methods to assist secretion clearance should be initiated when peak cough flow is <270 L/min

AHRF: acute hypercapnic respiratory failure, ALS: amyotrophic lateral sclerosis, cmH2O: centimeters of water (pressure), CRF: chronic respiratory failure, CWD: chest wall deformity, EPAP: expiratory positive airway pressure, FVC: Forced vital capacity, IMV: invasive mechanical ventilation, KQ: key question, LTOT: long term oxygen therapy, mbar: megabar (pressure), mmHg: millimeters of mercury (pressure), NIPPV/NPPV: non-invasive positive pressure ventilation, NIV: non-invasive ventilation, NMD: neuromuscular disease, PCO2/PaCO2: partial pressure of arterial carbon dioxide, QOL: quality of life, RR: respiratory rate, tCO2/PTcCO2: transcutaneous carbon dioxide pressure, VC: vital capacity

Table G.5. Guidelines for Obesity Hypoventilation Syndrome

Organization	Topic	KQ	Statement
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Device initiation criteria	KQ1	In patients with OHS, NIV should be started in acute hypercapnic respiratory failure using the same criteria as in AECOPD (pH<7.35 and pCO2 >6.5 kPa persist or develop despite optimal medical therapy). Many patients with acute hypercapnic respiratory failure secondary to OHS will require long-term domiciliary support (CPAP or NIV). Following an episode of acute hypercapnic respiratory failure referral to a home ventilation service is recommended. Patients with OSA, OHS or overlap syndrome should not have nocturnal oxygen therapy alone ordered. It can be considered in patients with evidence of established ventilatory failure, where it should be given with NIV support
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device initiation criteria	KQ1	NIV is the treatment of choice for OHS. In patients with OHS who have a minor degree of nocturnal desaturation and no nocturnal rise in PaCO2, CPAP is a reasonable initial therapy provided that follow-up is arranged within one to three months to evaluate response to therapy. Polysomnography is useful for titrating and confirming efficacy of bilevel pressures. Under circumstances when access to more than one device (bilevel PAP or CPAP) is limited, bilevel therapy is recommended. In patients with OHS who experience significant nocturnal desaturation or a nocturnal increase in PaCO2, bilevel PAP remains the therapy of choice.
Clinical Indications for Noninvasive Positive Pressure Ventilation in Chronic Respiratory Failure due to Restrictive Lung Disease, COPD, and Nocturnal Hypoventilation—A Consensus Conference Report, American Academy of Home Care Physicians, American College of Chest Physicians, American College of Physicians, American Sleep Disorders Association, American Thoracic Society, National Association for Medical Direction of Respiratory Care, 1999[74]	Device initiation criteria	KQ1	Before considering NIPPV for a patient with nocturnal hypoventilation from causes other than COPD or neuromuscular disease (criteria as outlined in part 1 and 2), a physician with demonstrated skills and experience in NIPPV must establish and document an appropriate diagnosis from this category on the basis of history and physical examination. A polysomnogram (PSG) is required for diagnosis of sleep apnea. A CPAP trial is recommended if OSA is documented unless a previous CPAP trial was unsuccessful or there is significant hypoventilation that is believed to be unlikely to respond to CPAP alone. Indications for usage of NIPPV PSG criteria for OSA not responsive to CPAP PSG criteria for mixed sleep apnea not responsive to CPAP Central sleep apnea Other forms of nocturnal hypoventilation
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device initiation criteria	KQ1	Indications for NIV in OHS include an awake PaCO2 >45mmHg and failure of CPAP therapy as evidence by either sustained oxygen desaturation during sleep or an increase in nocturnal daytime or nocturnal CO2 >8mmHg.
German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for	Device initiation criteria	KQ1	Due to the high prevalence of an accompanying obstructive sleep apnea syndrome (90% of cases), primary sleep diagnostics by means of polysomnography are necessary. The indication of NIV for patients with symptomatic CRF under adequate CPAP therapy yields to the following situations: A ≥ 5 minute-long increase in nocturnal PTcCO2 > 55mmHg and in PaCO2 ≥ 10 mmHg, respectively, in

Organization	Topic	KQ	Statement
Treatment of Chronic Respiratory Failure, 2010[75]			comparison to the awake state or Desaturations < 80% SaO2 over ≥ 10 minutes In the case of severe hypercapnia or symptomatic, severe co-morbidity, primary NIV can be implemented according to the physician's assessment. If the first control visit (including poly(somno)graphy under CPAP therapy) reveals no improvement in the characteristic symptoms of chronic hypoventilation or the absence of daytime normocapnia ("non-responder"), transfer of the patient to NIV is indicated. CPAP or NIV are the primary treatment options for HMV of patients with OHS. An accompanying loss of weight should also be aimed for. An initial attempt at CPAP treatment under polysomnographical conditions should take place in patients without significant co-morbidities. In the presence of significant co-morbidities, however, primary NIV therapy can be indicated. Persistent hypoventilation under CPAP (≥ 5 minute-long increase in PTcCO2 > 55mmHg and PaCO2 ≥ 10 mmHg, respectively, in comparison to normocapnia during the awake state, or desaturation < 80% over ≥ 10 minutes) is an indication for NIV. Significant weight loss can enable a change from NIV to CPAP therapy, or even an attempt at resting the treatment.
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device initiation, monitoring, and candidate selection	KQ1	Simple spirometry, SpO2 and serum bicarbonate should be performed in all patients referred for SDB assessment when BMI is greater than 35kg/m2. Arterial blood gases should be obtained in those individuals where SpO2 is ≤ 92% or where the serum bicarbonate is >27mmol/L to confirm the presence and severity of hypoventilation. Thyroid function should also be assessed and any airflow limitation treated appropriately. Positive airway pressure is first line therapy in patients with OHS, although adjunctive oxygen therapy is likely to be required, at least initially, for a significant number of patients. Autotitrating and home studies are not appropriate for this patient group. A full PSG should be performed during manual titration in order to identify the nature of the sleep disordered breathing and response to CPAP pressure. Many individuals will respond to initial intervention with CPAP. Titration should commence in CPAP mode to document the patient's response to abolition of upper airway obstruction alone.
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device continuation, compliance, and outcomes	KQ1 and KQ2	Individuals initially using bilevel support should be reviewed again after 3 months on therapy and CPAP retried, since a significant number may be switched to CPAP without clinical deterioration. In patients placed on CPAP in whom awake PaCO2 at baseline was 45-55mmHg, a clinical review at one month with repeat blood gases should be performed. Bilevel support should be used as initial therapy in patients presenting with acute decompensated respiratory failure. After 3 months, a CPAP titration should be undertaken to determine long term therapy. The need for and type of nocturnal PAP therapy should be reassessed if significant weight loss occurs.
German Society for Pneumology), Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure, 2010[75]	Device characteristics and titration	KQ3	Titration of CPAP pressure until hypoventilation is eliminated For NIV therapy, increase EPAP until obstructions are eliminated accompanied by titration of inspiratory pressure. In the case of considerable weight loss, a repeated attempt at CPAP, a change from NIV to CPAP, or a rest in treatment are all possible under poly(somno)graphical control. Weight loss should be part of the long-term treatment plan.

Organization	Topic	KQ	Statement
Canadian Thoracic Society, Home Mechanical Ventilation Clinical Practice Guideline, 2011[72]	Device characteristics and titration (Central hypoventilation syndrome)	KQ3	CHS patients who require only nocturnal ventilator support may be managed by NIV with a backup rate or diaphragmatic pacing. Severe CHS, mainly seen in congenital CHS, requires continuous invasive ventilator support, but daytime diaphragmatic pacing can markedly improve mobility and, as the child matures, NIV may suffice.
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Device characteristics and titration	KQ3	High inspiratory positive airway pressure (IPAP) and expiratory positive airway pressure (EPAP) settings are commonly required in patients with OHS (e.g., IPAP>30, EPAP>8). Volume control (or volume assured) modes of providing NIV may be more effective when high inflation pressures are required.

AECOPD: acute exacerbation of chronic obstructive pulmonary disease, BMI: body mass index, CHS: central hypoventilation syndrome, CO2: carbon dioxide, COPD: chronic obstructive pulmonary disease, CPAP: continuous positive airway pressure, CRF: chronic respiratory failure, EPAP: expiratory positive airway pressure, HMV: home mechanical ventilation, IPAP: inspiratory positive airway pressure, kPa: kilopascal, KQ: key question, mmHg: millimeters of mercury (pressure), mmol: millimole, NIPPV/NPPV: non-invasive positive pressure ventilation, NIV: non-invasive ventilation, OHS: Obesity hypoventilation syndrome, OSA: obstructive sleep apnea, PAP: positive airway pressure, PCO2/PaCO2: partial pressure of arterial carbon dioxide, pH: potential of hydrogen, PSG: polysomnogram, SaO2: arterial blood oxygen saturation, SDB: sleep disordered breathing, SpO2: Blood oxygen saturation level, tCO2/PTcCO2: transcutaneous carbon dioxide

Table G.6. Guidelines for Other Respiratory Diseases

Organization	Topic	KQ	Statement
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Device initiation, monitoring, and candidate selection (Asthma)	KQ1	Acute (or acute on chronic) episodes of hypercapnia may complicate chronic asthma. This condition closely resembles COPD and should be managed as such.
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Device initiation, monitoring, and candidate selection (Bronchiectasis)	KQ1	In patients with non-CF bronchiectasis, NIV should be started in acute hypercapnic respiratory failure using the same criteria as in AECOPD (pH<7.35 and pCO2 >6.5 kPa persist or develop despite optimal medical therapy).
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Device initiation, monitoring, and candidate selection (Cystic fibrosis)	KQ1	In patients with cystic fibrosis, NIV is the treatment of choice when ventilatory support is needed.
British Thoracic Society, Guidelines for Home Oxygen Use in Adults, 2015[73]	Device initiation, monitoring, and candidate selection (Cystic fibrosis)	KQ1	Nocturnal oxygen therapy should not be given to CF patients with nocturnal hypoxaemia alone who do not fulfil LTOT criteria. It can be considered in patients with evidence of established ventilator failure, where it should be given with NIV support.
Agency for Clinical Innovation, Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	Device initiation, monitoring, and candidate selection (Cystic fibrosis)	KQ1	Individuals with awake SpO2<94% or spirometry (FEV1<65% predicted) are at risk of nocturnal oxygen desaturation. Overnight oximetry should be undertaken in individuals meeting these criteria. Non-invasive ventilation is indicated if daytime CO2>45mmHg and nocturnal gas exchange shows SpO2<90% for >5% of TST and/or a rise in TcCO2 / ETCO2 from NREM to REM >5mmHg during room air breathing occurs. Nocturnal NIV is more effective than oxygen therapy in controlling nocturnal hypoventilation in patients with hypercapnic CF lung disease. Bilevel ventilation should be trialled initially. Volume ventilation may offer additional benefits in some individuals especially if work of breathing is high. NIV does not appear to increase the incidence of pneumothorax, but this is a relatively common occurrence in this population. Therefore, patients need to be educated regarding the symptoms of pneumothorax and should seek immediate medical attention should these symptoms arise. Changes in awake blood gases are not the best measure of the effectiveness of NIV in CF. Changes in symptoms, exertional dyspnoea and exercise tolerance, and control of nocturnal hypoventilation are better indicators of the patient's response to therapy. NIV may be used in patients unsuitable for transplant to relieve symptoms and improve sleep quality. However, alternative methods of symptom relief need to be introduced at the appropriate time.
Agency for Clinical Innovation,	Device	KQ1	Awake PaCO2 > 45 mmHg in the absence of lung and chest wall abnormalities, skeletal malformations and

Organization	Topic	KQ	Statement
Australia, Domicilary Non-Invasive Ventilation in Adult Patients, 2012[71]	initiation, monitoring, and candidate selection (Hypercapnic central sleep apnea)		neuromuscular disorders, in combination with symptoms consistent with sleep disordered breathing warrant a full polysomnogram. In patients with isolated sleep hypoventilation, titrate NIV settings in a spontaneous-timed mode, during a full polysomnogram. Where hypercapnic central apnoea is caused from pharmacological intake (e.g. opioid based derivatives), referrals to chronic pain team or relevant prescribing body should be made with the aim of reducing medication intake in order to improve central events and stabilise oxygen saturations. Overall patient management should be performed by specialised teams. Any signs of chest infection should be reviewed and managed promptly, especially in the case of CCHS where a lack of dyspnoea in response to pneumonia may mask severe respiratory compromise.
British Thoracic Society/Intensive Care Society, Guideline for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults, 2016[76]	Respiratory services (Cystic fibrosis)	KQ4	In patients with cystic fibrosis, specialised physiotherapy is needed to aid sputum clearance.

CCHS: congenital central hypoventilation syndrome, CF: cystic fibrosis, CO2: carbon dioxide, COPD: chronic obstructive pulmonary disease, ETCO2: end tidal carbon dioxide, FEV1: Forced expiratory volume in one second, kPa: kilopascal, KQ: key question, LTOT: long term oxygen therapy, mmHg: millimeters of mercury (pressure), NIV: non-invasive ventilation, NREM: non-rapid eye movement, PCO2/PaCO2: partial pressure of arterial carbon dioxide, pH: potential of hydrogen, REM: rapid eye movement, SpO2: Blood oxygen saturation level, tCO2/TcCO2: transcutaneous carbon dioxide pressure