Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure*

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Bibliography
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Abbreviations

COPD Chronic obstructive pulmonary disease
CPAP Continuous positive airway pressure
CRF Chronic respiratory failure
EPAP Expiratory positive airway pressure
HMV Home mechanical ventilation
LTOT Long-term oxygen therapy
NIV Non-invasive ventilation
NMD Neuromuscular disease
OHS Obesity hypoventilation syndrome
$PaCO_2$ Partial pressure of arterial carbon dioxide
PaO_2 Partial pressure of arterial oxygen
PCF Peak cough flow
PEEP Positive end-expiratory pressure
PT_cCO_2 Partial pressure of transcutaneous carbon dioxide
SaO_2 Oxygen saturation
VC Vital capacity

1 Introduction

1.1 Background

In recent years, there has been an increase in scientific publications investigating mechanical ventilation as a treatment for chronic respiratory failure (CRF). Coupled with the rapid rise in the use of home mechanical ventilation (HMV), the current political debate about the escalating financial pressures placed upon the health system, and the need to implement an appropriately-structured healthcare system, the formulation of a set of interdisciplinary, scientific guidelines containing the following objectives for HMV is urgently required:

- To determine the specific indications (including the appropriate time point) for the initiation of HMV.
- To establish the appropriate diagnostic and therapeutic approaches necessary for the implementation of a home ventilation system.
- To logistically plan the transfer of the ventilated patient from the hospital to the home environment.
- To address the technical and personnel requirements of the institutes participating in the treatment of the home-ventilated patient.
- To compile a set of criteria for quality control of HMV.

1.2 Complete Version

The following text is based on the complete, detailed version of guidelines (in German), which can be freely obtained from the websites of the German Medical Association of Pneumology and Ventilatory Support (DGP: www.pneumologie.de), the Association for Home Ventilation and Respiratory Weaning (AGH: www.heimbeatmung.de) and the Association of Scientific Medical Societies in Germany (AWMF: www.uni-duesseldorf.de/AWMF). The full version of these guidelines is also published in the journal "Pneumologie" [1] in German.

1.3 Methodology

These guidelines were developed in accordance with the AWMF system and correspond to the S2 level. A formal determination of the level of evidence was not carried out; a formal consensus was reached within the realm of two expert guideline conferences. The relevant literature was obtained through a formalized literature search in the Medline and Cochrane databases as well as through individual recommendations.

1.4 Scientific Principles

The respiratory system consists of two parts which can be independently impaired; the gas exchange system (lungs) and the ventilating system (respiratory pump). In pulmonary failure, oxygen therapy is sufficient unless it is accompanied by severe impairment of the gas exchange process, which would then additionally require the application of positive airway pressure. In contrast, dysfunction within the ventilatory system primarily requires mechanical ventilation (**Fig. 1**) [2].

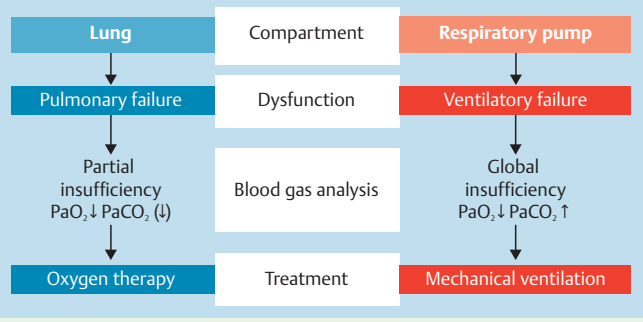

Fig. 1 The respiratory system.

The pathophysiological changes accompanying ventilatory failure comprise an increased load and/or reduced capacity of the respiratory muscles, which, as a result, become overstrained. The consequent hypoventilation most commonly manifests initially under conditions of increased activity and/or during sleep (initially in REM-sleep in particular) [3,4]. The potential causes for ventilatory failure are manifold, of which cerebral respiratory dysfunction, neuromuscular disorders (NMD), thoracic deformaties, chronic obstructive pulmonary disease (COPD) as well as obesity hypoventilation syndrome (OHS) are the primary candidates [5].

Ventilatory failure can appear suddenly and can then become coupled with respiratory acidosis. In CRF, the respiratory acidosis is in turn metabolically compensated through bicarbonate retention. Episodes of acute deterioration commonly develop on the basis of an already-present chronic disturbance, producing a conflicting blood gas profile of high bicarbonate levels and low pH values [5].

The symptoms of CRF are numerous and often combined with symptoms of the specific underlying disease, whereby dyspnea, morning headache and symptoms of sleep-disordered breathing are precedent [6,7].

Patients suffering from CRF can electively be introduced to HMV, which is most often applied intermittently; that is, nocturnal ventilation is usually alternated with intervals of spontaneous breathing during the day [6–9]. This ventilation therapy can be carried out either invasively via a tracheostoma or non-invasively via a facial mask, and aims to improve blood gases during both mechanical ventilation and the subsequent intervals of sponta-

neous breathing; the main objective is to reduce the partial pressure of arterial carbon dioxide ($PaCO_2$), with normocapnia being the best case scenario. Potential side effects and the patient's tolerance of mechanical ventilation must be taken into account during this process [6, 10, 11].

2 Technical Installation

The physicians are responsible for indicating HMV, and for choosing the type of ventilator, the ventilation mode and ventilation parameters. Uncontrolled changes to the ventilation set-up can potentially lead to life-threatening complications; therefore, any alterations to the ventilation system must only occur upon doctor's orders and be carried out preferably under clinical supervision. The following areas warrant special mention:

2.1 Ventilators

The basic requirements for ventilators were determined according to ISO-Standards, distinguishing between „Home care ventilators for ventilator-dependent patients" (ISO 10651-2: 2004) and „Home-care ventilatory support devices" (ISO 10651-6: 2004). In life-supporting ventilation, or for patients unable to remove their own face masks, a ventilation machine with an internal battery is required (ISO 10651-2: 2004). If the patient's ability to breathe spontaneously is greatly reduced (daytime ventilation time > 16 hours), an external battery pack with a capacity of at least 8 – 10 hours is required [12]. If the duration of mechanical ventilation exceeds 16 hours/day, an additional identical ventilator must be provided [12]. The replacement of the existing ventilator with a different type of machine or the adjustment of the ventilation mode must each take place under hospital conditions in a centre specialized for mechanical ventilation (also see chapter 3.5).

2.2 Tubing and Exhalation Systems

Single tube systems with exhalation systems positioned within patient proximity are commonly used. In an open so-called 'leakage' system, a series of openings in the tubing system or face mask that are close to the patient are present to aid in the elimination of expired CO_2. The prerequisite for this is the presence of continuous positive pressure during expiration (EPAP: expiratory positive airway pressure), since a significant amount of CO_2 can otherwise be rebreathed from the tubing system [13]. Pneumatically-driven exhalation vents can alternatively perform this task. A changeover of the exhalation system must take place under clinically-controlled conditions [14].

2.3 Ventilation Interfaces

Nasal masks, oronasal masks, full-face masks, mouth masks or mouth pieces are all available for home non-invasive ventilation (NIV) purposes. The choice depends on the patient's tolerance of the ventilation as well as on ventilation efficacy. Every patient should possess at least one reserve mask; for long periods of ventilation, a number of different masks may be necessary to relieve contact pressure zones [15, 16].

For home invasive ventilation, the tracheostoma must be stable, which generally corresponds to being epithelialised. In ventilation via a tracheal canulae, either blocked or unblocked canulae can be used; the use of blocked canulae requires a cuff pressure gauge [17]. In addition to the required reserve canulae of the same size, one smaller reserve canula must also be at hand to aid emergency canulation in difficult cases of canula exchange [12].

2.4 Humidifiers

Air conditioning systems (humidifiers and warmers) are fundamentally categorized as active or passive [16, 18, 19], as summarised in **Table 1**:

Table 1 Humidifiers.

Active		Passive
Bubble- through humidifiers	*Pass-over humidifiers*	*Heat and Moisture Exchanger (HME)*
Air flows through water	Air passes over water	Conserves patient's own humidity and airway temperature
Sterile water required	Sterile water not required	Can alter breathing mechanics Must never be used in conjunction with an active humidifying system!

Invasive ventilation always requires a humidifying system [20], in non-invasive ventilated patients a humidifier should be prescribed according to the patient's symptoms [16].

2.5 Additional Accessories

Unit-side particle filters fitted at the point of air inlet are necessary. There is insufficient evidence to suggest that filters at the point of air outlet are necessary for home use. It is recommended that filters are changed at 1 – 7 day intervals [21].

The oxygen flow rate is clinically titrated. Home monitoring via pulse oximetry is not mandatory. However, patients with NMD and cough insufficiency (see Ch. 8), as well as children (see Ch. 9), present as exceptions: in these patient groups, a drop in oxygen saturation can prematurely indicate imminent, significant secretion retention [22]. Selective measurements during invasive ventilation are also worthwhile [23].

Invasively-ventilated patients require high-performance, battery-supplied suction devices (flow rate > 25 litres/min), as well as a replacement machine and ventilation bag [12].

Recommendations

- Alterations to the ventilator or ventilator settings must occur exclusively upon doctor's orders and be carried out under clinical supervision.
- 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; every invasively-ventilated patient requires at least one reserve canula.
- 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 oxymeter is necessary.

3 Set-up, Adjustment and Control of the Ventilator

3.1 Centre for Home Mechanical Ventilation

HMV must be organized via a specialized centre; this is a clinic that has expertise in indicating, initiating and monitoring HMV. A more exact definition of structural and procedural quality will be determined in the future, while the accreditation of centres for HMV is also aspired to.

3.2 Diagnostics

In addition to medical history taking and physical examination, the following technical inspections are necessary before the initiation of ventilation:

- Basic labs and electrocardiogram (ECG)
- Blood gas analyses (day and night) during ambient air breathing and with oxygen supply, respectively, or continuous overnight transcutaneous measurement of CO_2 (PT_cCO_2).
- Pulmonary function tests; measurement of respiratory muscle function and assessment of peak cough flow (if applicable)
- X-ray of the thorax in two planes
- Overnight polygraphy/polysomnography
- Exercise test (e.g. 6 minute walking test, ergometry)
- Echocardiography if cardiac co-morbidity is suspected

Overnight oximetry alone is neither sufficient to detect nocturnal hypoventilation, nor to indicate HMV.

3.3 Launching Home Mechanical Ventilation

The objective is to improve the patient's clinical symptoms and reduce $PaCO_2$ to the point of normocapnia [6]. The indications as well as the choice of ventilator and accessories are incumbent upon the treating physician in the centre for HMV, who either directly carries out the initial set-up of the ventilation system him/herself, or delegates the task to other specially-trained medical assistants (but not to technicians working for the equipment provider). The following criteria apply:

- Daytime initialization of ventilation on a specialized general ward, in a sleep lab or on an observational ward (intermediate- or (rarely) intensive care unit).
- Initialization with heart-rate and blood pressure monitoring, blood gas analysis, oxymetry, and/or assessment of PT_cCO_2 and measurement of tidal volumes.
- Inspiratory pressure level under pressure-controlled ventilation (or hybrid mode, if applicable) might – depending on the underlying disease – exceed 30 mbar (especially in COPD) [24–26].
- $SaO_2 < 90\%$ or $PaO_2 < 55$ mmHg under optimal ventilation indicate the need for additional oxygen supply (LTOT) [27].
- Although the objective is to establish nocturnal ventilation, daytime ventilation can also be effective; if necessary, a combination of nocturnal and daytime ventilation can be applied [28,29].
- During the course of the initialization, the effectiveness of the ventilation should be assessed via $PaCO_2$, both during spontaneous breathing and ventilation, respectively, and supplemented by nocturnal measurements (polygraphy/pulse oximetry, polysomnography, PT_cCO_2, selective blood gas analyses) [30,31].

3.4 Control Visits

The first control examination with nocturnal diagnostics should take place within the first 4–8 weeks [32,33]. Any side-effects of the ventilation treatment must be duly recorded and a thorough check of the complete ventilation system is obligatory. In the case of poor adherence, it is worthwhile repeating the control procedures under hospital conditions; however, if the therapy fails to be effective due to continued poor adherence (despite optimal therapeutic set-up), ventilation should be aborted. It is recommended that further control visits are carried out 1–2 times a year, depending on the type and progression of the underlying disease, as well as on the quality of response to the therapy thus far.

3.5 Changing the Ventilator and Ventilator Interface

The exchange of identically-built machines with maintenance of all parameters can take place at home. Different machines, including those from the same manufacturer, must be exchanged under controlled conditions in the centre for HMV. The changeover to other tracheal-canula models and ventilation masks must only occur in close consultation with the centre for HMV, or, if applicable, directly in the hospital. When changing tracheal-canula models a subsequent bronchoscopic control examination should be performed [34–37].

Recommendations

- 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 CO_2 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.

4 Establishing Home Mechanical Ventilation

High-quality, individually-customized care is paramount in the management of ventilated patients. The aim is to adapt at any time the extent of care to the necessity of ventilation duration and type of ventilation interface; the inclusion of relatives in the care of ventilated patients is also paramount. This is only accomplished through close consultation and good organization between all participating professionals.

4.1 Prerequisites for Discharge from Hospital

The transition phase from the clinical to the non-clinical environment is highly vulnerable [33]. For quality of life reasons, it is preferable to accommodate the patient at home [38,39]. The correct time point for discharge is reached only when the underlying and secondary illness(es) are deemed stable [40], and when the meeting of costs as well as the provision of the necessary equipment, resources and materials have been secured. If the ventilated patient is still not at an optimal level of function and performance, (early) rehabilitative measures should be considered [39].

4.2 Out-of-hospital Care Team

The care of a home-ventilated patient entails:

- Ongoing clinical supervision (usually provided by respiratory physicians, anaesthetists, paediatricians or neurologists) in conjunction with the centre for HMV.
- At-home care team, lay helpers (including relatives).
- Technical support from equipment providers for machines and accessories.
- A team of therapists (speech-, occupational-, physio- and social therapists, teachers).

A qualified care team as well as a representative from the equipment provider should always be contactable [39,41,42].

4.3 Assistive and Professional Care

Assistive care generally entails aid from minimally-qualified helpers, while specialized professional care should only be provided by legitimate health care professionals carrying a high level of qualification. The necessary quality of care is determined by the degree of ventilation dependence, as well as the autonomy of the patient; this decision is incumbent upon the centre for HMV. For the training and qualification criteria for professional carers, please see the detailed version of guidelines.

4.4 Management of the Transition Phase

The transition-management team should consist of the following professionals:

- Team manager
- Doctor (in the clinic and at home)
- Care team (in the clinic and at home)
- Equipment provider
- Social worker, social therapist
- Specialized therapists (if necessary)
- Health insurance provider (if applicable)

Allocation of team members should be done in consultation with the patient's relatives. The team should begin planning as early as possible the discharge of the patient from the hospital [39].

A check list of the minimal requirements for patient discharge and subsequent set-up of the home ventilation station comprises the following:

- Full technical installation of the ventilatory machinery and surveillance systems
- Surveillance standards in terms of personnel (nurse attendance time)
- Time schedule and content of nursing procedures
- Type of ventilation interface and the corresponding cleaning and exchange intervals
- Detailed description of ventilator mode and associated parameters
- Duration of assisted ventilation and, if applicable, phases of spontaneous ventilation
- Oxygen flow rates during assisted and spontaneous ventilation
- Procedures for managing secretions
- Application of inhaled medication
- Planning for nutritional needs
- Psychosocial care of the patient and, if applicable, the relatives
- Additional therapeutic and educative measures
- Additional resources (e.g. rollator, therapeutic bed, communication aids)

4.5 Surveillance and Documentation of Home Mechanical Ventilation

In cases of permanent ventilation, the ventilation parameters and measured values should be continuously monitored and documented accordingly; this should be performed at least once per shift. Clinical changes (e.g. increasing spontaneous breathing times, deteriorations) require medical consultation and treatment. Conduct during emergency situations should be based on the medically-necessary aspects as well as on those declared in the patient's living will (see Ch. 10).

4.6 Equipment Provider

The machine provider is responsible for briefing all personnel and relatives involved in the care of the ventilated patient. An additional briefing on the day of hospital discharge and a functional check-up of the machinery at the final ventilation location are generally desirable; for specialized nursing care, this is obligatory. If technical problems with the ventilator and/or interface arise, a technician should be available within 24 hours to solve the issue [39].

Recommendations

- HMV must be organized in a centre for HMV, and the treating physician is responsible for the organization of home care.
- The meeting of costs and supply of equipment, resources and materials must be secured before the ventilated patient is discharged from hospital
- Professional care is more extensive than assistive care and therefore requires highly-qualified care personnel.
- The equipment provider must guarantee round-the-clock availability and ensure a prompt and customized service. An introduction to the ventilation machinery is compulsory.

5 Obstructive Airway Diseases

NIV is the primary therapeutic option for COPD patients with CRF [24,25,43–47], whereas long-term invasive ventilation via a tracheostoma is only applied nowadays under exceptional circumstances, predominantly after weaning failure.

NIV in combination with physiotherapy for cystic fibrosis can facilitate cough-up of thick, sticky mucous secretions [48].

5.1 Indications

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) (**Fig. 2**) indicate the need for HMV:

- Chronic daytime hypercapnia with $PaCO_2 \geq 50$ mmHg
- Nocturnal hypercapnia with $PaCO_2 \geq 55$ mmHg
- Stable daytime hypercapnia with 46–50 mmHg and a rise in PT_cCO_2 to ≥ 10 mmHg during sleep [49].
- Stable daytime hypercapnia with $PaCO_2$ 46–50 mmHg 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 [50].

Poor compliance with medication intake and/or LTOT are relative contraindications. Complete discontinuation of nicotine abuse should be aspired to.

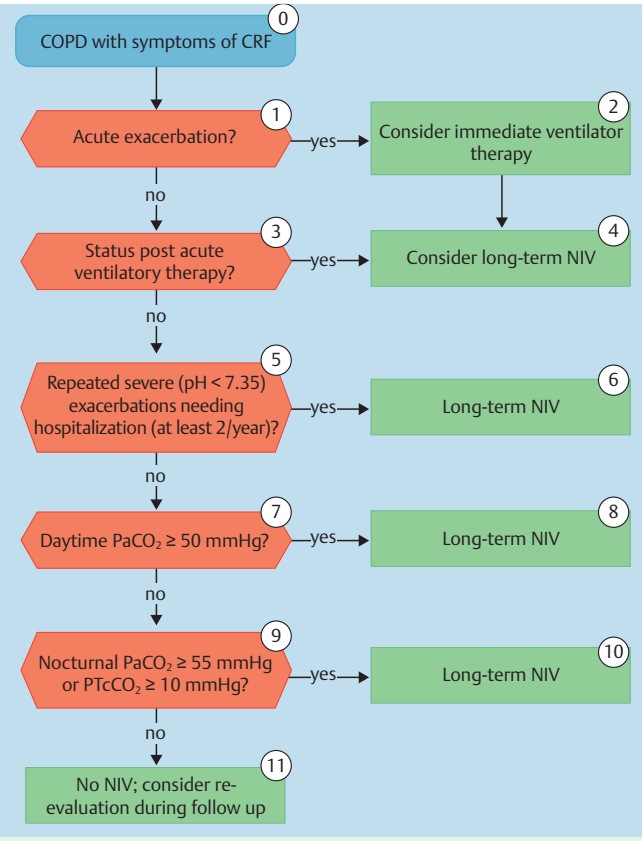

Fig. 2 Non-invasive ventilation (NIV) therapy approach in hypercapnic patients with chronic obstructive pulmonary disease (COPD).

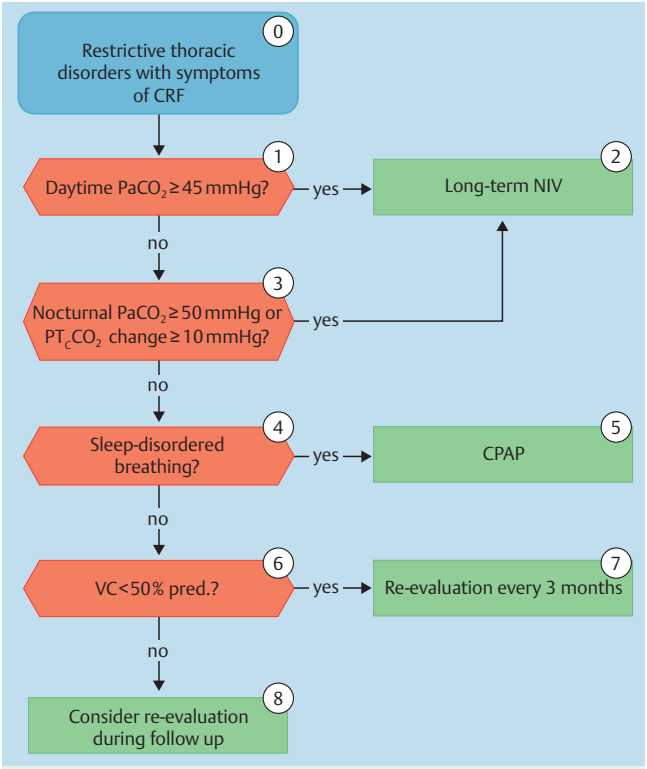

Fig. 3 Non-invasive ventilation (NIV) therapy approach in patients with hypercapnic restrictive thoracic diseases (RTD).

5.2 Procedure

- Controlled ventilation mode with ventilation pressures from 20 to 40 mbar. Pressure escalation until normocapnia or maximum tolerance is reached [11, 24 – 26, 51].
- 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 [52]
- The introduction of NIV in the hospital can take up to two weeks.

Recommendations

- 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.
- The aim of the ventilation is to normalize $PaCO_2$; sufficiently high ventilation pressures are required to achieve this.

6 Restrictive Thoracic Diseases

NIV is the primary treatment option for restrictive thoracic disease patients with CRF [8, 53 – 55]. This generally encompasses the following conditions [9, 56]:

- Scoliosis
- Kyphosis
- Pectus carinatum (pigeon chest)
- Pectus excavatum (concave chest)
- Ankolysing spondylitis
- Restrictive pleural diseases
- Post-tuberculosis syndrome
- Post-traumatic thoracic deformities
- Post-operative thoracic deformities (thoracoplastic)

6.1 Indications

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) (**Fig. 3**):

- Chronic daytime hypercapnia with $PaCO_2 \geq 45$ mmHg
- Nocturnal hypercapnia with $PaCO_2 \geq 50$ mmHg
- Daytime normocapnia with a rise in PT_cCO_2 of ≥ 10 mmHg 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.

6.2 Procedure

- NIV in pressure- and volume-limited modes is feasible [57 – 60]
- With set pressure, maximal ventilation pressure often reaches 20 – 25 mbar [56]
- Changeover from set pressure to set volume should be taken into account in order to improve ventilation [57, 61]
- EPAP is generally not necessary if bronchial obstructions are absent [56].

Recommendations

- 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.

7 Obesity Hypoventilation Syndrome

CPAP and NIV are the primary treatment options for OHS patients with CRF [62], in accordance with the following.

7.1 Indications

Due to the high prevalence of an accompanying obstructive sleep apnea syndrome (90% of cases), primary sleep diagnostics by means of polysomnography are necessary [63–68].

The indication of NIV for patients with symptomatic CRF under adequate CPAP therapy yields to the following situations (**Fig. 4**):

- A ≥ 5 minute-long increase in nocturnal PT_cCO_2 > 55 mmHg and in $PaCO_2$ ≥ 10 mmHg, respectively, in comparison to the awake state.

or

- Desaturations < 80% SaO_2 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 [69].

7.2 Procedure

- 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 [70].
- Weight loss should be part of the long-term treatment plan.

Recommendations

- 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 PT_cCO_2 > 55 mmHg and $PaCO_2$ ≥ 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.

8 Neuromuscular Diseases

Patients with neuromuscular disease (NMD) at risk of developing respiratory muscle weakness should undergo regular examinations of lung function and blood gases (every 3–12 months, depending on the underlying disease); a polygraphy is also necessary if VC < 70%. These measures are important to ensure an early diagnosis of respiratory muscle weakness, rather than first detecting it in the event of respiratory decompensation.

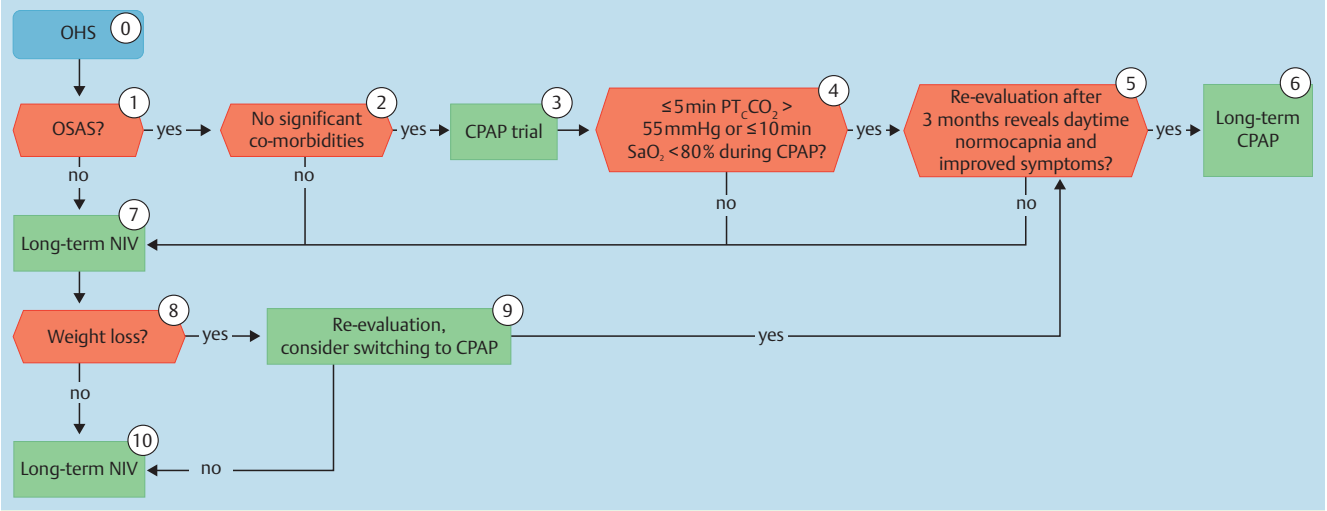

Fig. 4 Continuous positive airway pressure (CPAP) and non-invasive ventilation (NIV) therapy approach in obesity-hypoventilation syndrome patients (OHS).

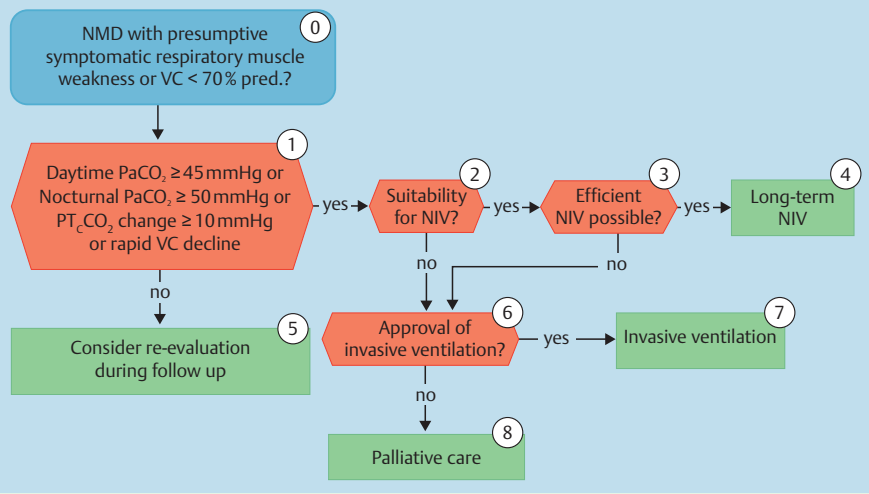

Fig. 5 Non-invasive ventilation (NIV) therapy approach in patients with neuromuscular diseases (NMD).

8.1 Indications for NIV

NIV of NMD patients with clinical signs of CRF is indicated by the following (at least 1 criterion should be fulfilled) [7, 8, 56, 71 – 81]:

- Chronic daytime hypercapnia with $PaCO_2 \geq 45$ mmHg
- Nocturnal hypercapnia with $PaCO_2 \geq 50$ mmHg
- Daytime normocapnia with a rise in PT_cCO_2 of ≥ 10 mmHg 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 [82]. NIV is also indicated prior to elective vertebral column correction surgery when VC < 60% target value and FEV_1 < 40% target value, respectively [83], or during pregnancy with restricted lung function [84], as well as palliative care of dyspnea [85].

8.2 Indications for Invasive Ventilation via Tracheostoma

There is an indication for tracheotomy in the following situations (in accordance with the thoroughly-informed patient's wishes and consent) (**Fig. 5**) [71, 73, 79, 81, 86]:

- When fitting of an appropriate NIV interface is impossible
- Intolerance of NIV
- Ineffectiveness of NIV
- Severe bulbar symptoms with recurrent aspiration
- Ineffective non-invasive management of secretions
- Failure to transfer to NIV after invasive ventilation

8.3 Procedure

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 [72, 76 – 78, 87 – 91]
- Hypersalivation; therapy with anti-cholinergics (e.g. Scopolamine patch, amitryptiline or botulinum toxin injections into the salivary glands [92])
- Coughing weakness, with the development of acute decompensation (also see Ch. 8.4)

For further special aspects that need to be considered, particularly those relating to amyotrophic lateral sclerosis, please refer to the complete version of guidelines.

8.4 Cough Impairment and Secretion Management

A reduced cough impulse (peak cough flow; PCF < 270 l/min) can lead to acute decompensations and increased incidence of aspiration pneumonia [93]. Measures to eliminate secretions should therefore be taken when SaO_2 < 95%, or a 2 – 3% drop in the patient's individual best value occurs.

Step-based secretion management (see **Fig. 6**) 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®) [81, 94 – 99].

Recommendations

- Patients with NMD should undergo clinical assessment and assessment of VC at 3 – 12 month-intervals. Polygraphy and PT_cCO_2-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.
- 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.

9 Special Considerations for Paediatric Ventilation

Most of the underlying diseases that lead to CRF in childhood (**Table 2**) are complex and often associated with multiple disabilities that must be treated in a spezialized clinic. A therapeutic master plan must anticipate both the progressive course of the underlying disease and all corresponding respiratory complications, and also include infection prevention, ventilation, treatment of cough insufficiency, sufficient nutrition and adequate management of complications and emergencies [73, 100, 101]. Burdens such as fever, airway infections or operations may necessitate earlier implementation of ventilation [73, 100, 102, 103].

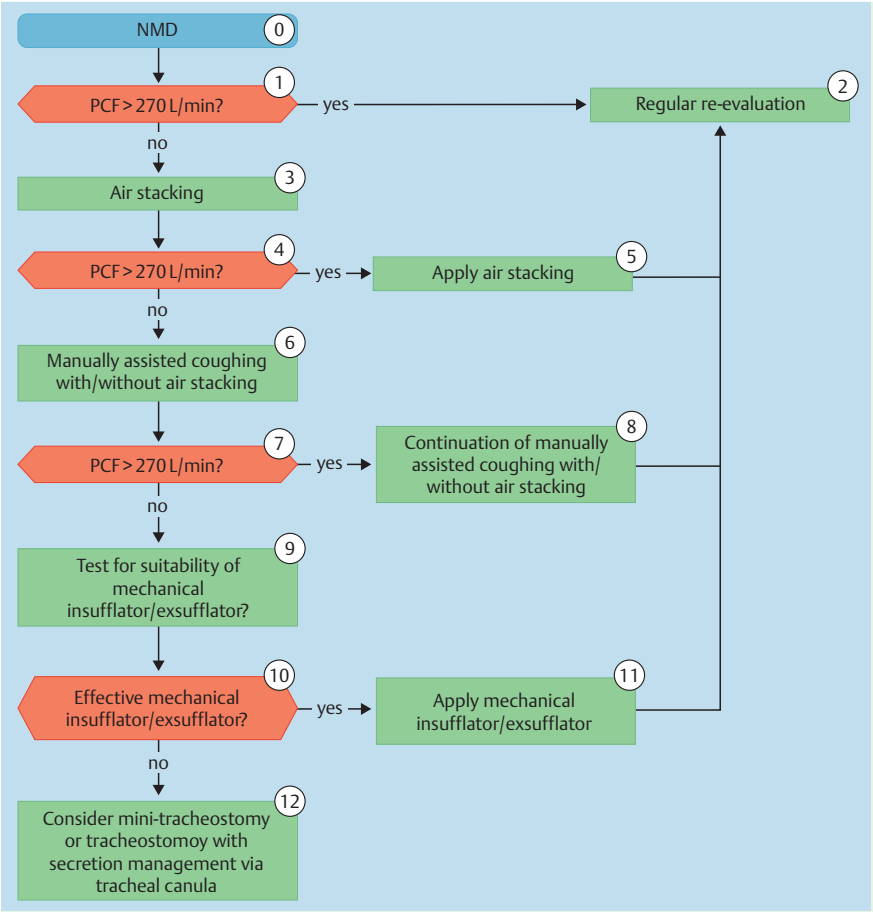

Fig. 6 Flow chart for secretion management in non-invasively ventilated patients with neuromuscular diseases (NMD).

Table 2 Paediatric diseases that are accompanied by ventilatory failure and may require ventilation therapy.

1. Lung Diseases
– Cystic Fibrosis
– Bronchopulmonary Dysplasia
2. Neuromuscular Disorders
– Duchenne's muscular dystrophy
– Spinal muscular atrophy
– Congenital muscular dystrophy
– Myotonic dystrophy
– Myopathy (congenital, mitochondrial, storage diseases)
3. Diseases und Syndromes with Primary and Secondary Thoracic Deformities
– Asphyxiating thoracic dystrophy
– Achondroplasia
– McCune-Albright Syndrome
– Cerebral palsy
– Meningomyelocele
4. Disorders of Central Respiratory Regulation
– Congenital central hypoventilation (Undine Syndrome)
– Acquired central hypoventilation after trauma, encephalitis or CNS degeneration
– Hydrocephalus with increased cranial pressure
– Arnold Chiari malformation
5. Obesity Hypoventilation Syndrome
– Morbid alimentary obesity
– Prader-Willi Syndrome
6. Diseases with primary, unrectifiable obstruction of the upper airway (when CPAP-therapy is inadequate)
– Down Syndrome
– Mitochondriopathies
– Mid-facial hypoplasias (Pierre-Robin Syndrome and others)
– Morbid alimentary obesity
– Prader-Willi Syndrome

9.1 Special Aspects in Home Mechanical Ventilation of Paediatric Patients

- Not all ventilators are licensed and appropriate for small children.
- Most children with muscle weakness are unable to independently trigger the ventilator.
- Small children have very low tidal volumes.
- Children have irregular breathing frequencies and depths.
- The ventilatory needs of children change constantly (depending on state of awakeness, stage of sleep, fever, airway infection).
- Customised masks have a relatively high amount of dead space and often don't fit children, especially infants. The risk of developing mid-facial hypoplasia is increased when using masks with high contact pressure [104, 105].
- Infants, as well as children with muscular disease and immobility, are unable to independently remove the ventilation mask in emergency situations (e.g. ventilator malfunction, power failure).

Hence, the following specific demands must be met:

- A sensitive trigger and low tidal volumes must be possible for optimal ventilation control.
- Particularly in infants, successful ventilation is usually only possible with pressure-driven equipment [80, 106 – 110].
- There is better adaptation to the breathing pattern and leakage under pressure-driven ventilation than under ventilation with preset volumes.
- Inefficacy of a conventional mask indicates replacement with an individually-customized mask. The manufacture of new masks is frequently required due to childhood growth.

9.2 Special Considerations for Paediatric Home Invasive Ventilation

In principle, there is no difference between children and adults in the indication for invasive ventilation and it should be determined in close consultation with the children, parents and treatment team.

- The danger for airway blockage with secretions increases with a decreasing inner diameter of the canula.
- Even a slight contamination of small canulae can lead to an exponential increase in airway resistance.
- The significant fluid loss that accompanies childhood tachypnoea requires sufficient conditioning of the inspired air.
- Sufficient leakage for sound production in babies and infants is necessary for speech development.
- Canula-associated emergencies occur more often in childhood than in adulthood (accidental removal of canula, aspiration).

Airway infections, fever, augmented secretions, cough, dyspnea and strenuous breathing indicate the application of a pulse oxymeter during spontaneous inhalation of ambient air (**Table 3**) [22].

Table 3 Protocol for pulse oxymeter.

SaO_2 > 95 %	No intervention required
SaO_2 between 90 % and 95 %	Intensification of ventilation with a mask and/or assisted coughing
SaO_2 < 90 % despite mechanical ventilation	Contact the centre for HMV

The care of home non-invasively- and invasively-ventilated children requires a multidisciplinary team. For a detailed account of the requirements, particularly those concerning ventilation monitoring and secretion management, please refer to the complete version of guidelines.

10 Ethical Considerations

Since the prognosis for patients with CRF is often quite poor, quality of life becomes paramount. In this light, HMV affords on the one hand the chance to relieve the extent of CRF and markedly improve the quality of life, whereas, on the other hand, holds the danger of unnecessarily prolonging the patient's suffering and preventing a dignified death after a long history of illness.

The German Federal Supreme Court decided back in 1991 that in the case of a hopeless prognosis, assistance with dying is allowed to be carried out in accordance with the declared or presumed will of the patient by way of "withholding" or "withdrawing" life-prolonging measures (such as ventilation), in order to allow a natural, dignified course of death (under analgesic medication, if applicable).

A detailed description of the "End of Life" principles relating to CRF and ventilation therapy can be found in the complete version of guidelines, and is summarized in the form of the following recommendations:

- In cases of highly-advanced or rapidly-progressive CRF, patients and their relatives should be informed well ahead of time of imminent respiratory emergencies and the therapeutic options for the end-stage of the disease.
- A partnership between the patient, physician and carer is also necessary in the final stage of the patient's life, where not only medical competence and care of duty, but also frank statements about the prognosis, particularly those concerning questions about the end of life, all remain indispensable.
- The rejection of treatment as expressed in the living will is binding for the treating physician, as long as the real, ensuing situation corresponds to the one described in the patient's will, and there is no recognisable evidence for retrospective changes of will.
- When "withholding" or "withdrawing" the ventilation therapy, the principles of palliative medicine must be applied in the form of combined, pre-emptive pharmacological and non-pharmacological treatment of dyspnea, agitation and pain.
- A separate room should be provided in which the patient is afforded a dignified death in the presence of relatives.

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References

1 *Windisch W, Brambring J, Budweiser S et al.* Nichtinvasive und invasive Beatmung als Therapie der chronischen respiratorischen Insuffizienz. S2-Leitlinie herausgegeben von der Deutschen Gesellschaft fur Pneumologie und Beatmungsmedizin e.V. Pneumologie 2010; 64: 207 – 240
2 *Roussos C.* The failing ventilatory pump. Lung 1982; 160: 59 – 84
3 *Criée C, Laier-Groeneveld G.* Die Atempumpe: Atemmuskulatur und intermittierende Selbstbeatmung. 1. Aufl. New York: Thieme, 1995
4 *Kabitz H, Windisch W.* Respiratory muscle testing: state of the art. Pneumologie 2007; 61: 582 – 587
5 *Tobin MJ, Laghi F, Brochard LJ.* Role of the respiratory muscles in acute respiratory failure of COPD: lessons from weaning failure. J Appl Physiol 2009; 107: 962 – 970
6 *Mehta S, Hill NS.* Noninvasive ventilation. Am J Respir Crit Care Med 2001; 163: 540 – 577
7 *AAHCP/AARC/AACP/AAP/ASDA/ATS/NAMDRC.* Clinical indications for noninvasive positive pressure ventilation in chronic respiratory failure due to restrictive lung disease, COPD, and nocturnal hypoventilation – a consensus conference report. Chest 1999; 116: 521 – 534
8 *Simonds AK, Elliott MW.* Outcome of domiciliary nasal intermittent positive pressure ventilation in restrictive and obstructive disorders. Thorax 1995; 50: 604 – 609
9 *Simonds AK.* Home ventilation. Eur Respir J Suppl 2003; 47: 38s – 46s
10 *Schönhofer B.* Choice of ventilator types, modes, and settings for long-term ventilation. Respir Care Clin N Am 2002; 8: 419 – 445
11 *Windisch W, Dreher M, Storre JH, Sorichter S.* Nocturnal non-invasive positive pressure ventilation: physiological effects on spontaneous breathing. Respir Physiol Neurobiol 2006; 150: 251 – 260
12 *AARC clinical practice guideline.* Long-term invasive mechanical ventilation in the home-2007 revision update. Respir Care 2007; 52: 1056 – 1062
13 *Schettino GPP, Chatmongkolchart S, Hess DR, Kacmarek RM.* Position of exhalation port and mask design affect CO2 rebreathing during noninvasive positive pressure ventilation. Crit Care Med 2003; 31: 2178 – 2182
14 *Younes M, Kun J, Webster K, Roberts D.* Response of ventilator-dependent patients to delayed opening of exhalation valve. Am J Respir Crit Care Med 2002; 166: 21 – 30
15 *Storre JH, Schönhofer B.* Noninvasive mechanical ventilation in chronic respiratory failure: ventilators and interfaces. In: Muir J, Ambrosino N, Simonds AK, eds. European Respiratory Monograph. Sheffield: ERS Journals, 2008: 319 – 337
16 *Navalesi P, Frigerio P, Gregoretti C.* Interfaces und humidification in the home setting. In: Muir J, Ambrosino N, Simonds AK, eds. European Respiratory Monograph. Sheffield: ERS Journals, 2008: 338 – 349
17 *Bach JR, Alba AS.* Tracheostomy ventilation. A study of efficacy with deflated cuffs and cuffless tubes. Chest 1990; 97: 679 – 683
18 *Wenzel M, Wenzel G, Klauke M et al.* Charakteristik mehrerer Befeuchter für die CPAP- sowie invasive und nicht invasive Beatmungstherapie und Sauerstofflangzeittherapie unter standardisierten Bedingungen in einer Klimakammer. Pneumologie 2008; 62: 324 – 329
19 *Nava S, Cirio S, Fanfulla F et al.* Comparison of two humidification systems for long-term noninvasive mechanical ventilation. Eur Respir J 2008; 32: 460 – 464
20 *Nakagawa NK, Macchione M, Petrolino HM et al.* Effects of a heat and moisture exchanger and a heated humidifier on respiratory mucus in patients undergoing mechanical ventilation. Crit Care Med 2000; 28: 312 – 317
21 *Ricard JD, Le Miere E, Markowicz P et al.* Efficiency and safety of mechanical ventilation with a heat and moisture exchanger changed only once a week. Am J Respir Crit Care Med 2000; 161: 104 – 109
22 *Tzeng AC, Bach JR.* Prevention of pulmonary morbidity for patients with neuromuscular disease. Chest 2000; 118: 1390 – 1396
23 *Vargo JJ, Zuccaro GJ, Dumot JA et al.* Automated graphic assessment of respiratory activity is superior to pulse oximetry and visual assessment for the detection of early respiratory depression during therapeutic upper endoscopy. Gastrointest Endosc 2002; 55: 826 – 831
24 *Dreher M, Storre J, Schmoor C, Windisch W.* High-intensity versus low-intensity non-invasive ventilation in stable hypercapnic COPD patients: a randomized cross-over trial. Thorax 2010; 65: 303 – 308
25 *Windisch W, Haenel M, Storre JH, Dreher M.* High-intensity non-invasive positive pressure ventilation for stable hypercapnic COPD. Int J Med Sci 2009; 6: 72 – 76
26 *Windisch W, Kostic S, Dreher M et al.* Outcome of patients with stable COPD receiving controlled noninvasive positive pressure ventilation aimed at a maximal reduction of Pa(CO2). Chest 2005; 128: 657 – 662
27 *Magnussen H, Kirsten A, Köhler D et al.* Leitlinien zur Langzeit-Sauerstofftherapie. Deutsche Gesellschaft für Pneumologie und Beatmungsmedizin e.V. Pneumologie 2008; 62: 748 – 756
28 *Schönhofer B, Geibel M, Sonneborn M et al.* Daytime mechanical ventilation in chronic respiratory insufficiency. Eur Respir J 1997; 10: 2840 – 2846
29 *Toussaint M, Soudon P, Kinnear W.* Effect of non-invasive ventilation on respiratory muscle loading and endurance in patients with Duchenne muscular dystrophy. Thorax 2008; 63: 430 – 434
30 *Storre JH, Steurer B, Kabitz H et al.* Transcutaneous PCO2 monitoring during initiation of noninvasive ventilation. Chest 2007; 132: 1810 – 1816
31 *Schönhofer B, Euteneuer S, Nava S et al.* Survival of mechanically ventilated patients admitted to a specialised weaning centre. Intensive Care Med 2002; 28: 908 – 916
32 *Farre R, Lloyd-Owen SJ, Ambrosino N et al.* Quality control of equipment in home mechanical ventilation: a European survey. Eur Respir J 2005; 26: 86 – 94
33 *Farre R, Navajas D, Prats E et al.* Performance of mechanical ventilators at the patient's home: a multicentre quality control study. Thorax 2006; 61: 400 – 404
34 *Farre R, Giro E, Casolive V et al.* Quality control of mechanical ventilation at the patient's home. Intensive Care Med 2003; 29: 484 – 486
35 *Fuchs M, Bickhardt J, Morgenstern U.* Variabilität von Beatmungsparametern bei Heimbeatmungsgeräten. Biomed Tech (Berl) 2002; 47 Suppl 1 Pt 2: 845 – 848
36 *Lofaso F, Fodil R, Lorino H et al.* Inaccuracy of tidal volume delivered by home mechanical ventilators. Eur Respir J 2000; 15: 338 – 341
37 *Vitacca M, Barbano L, D'Anna S et al.* Comparison of five bilevel pressure ventilators in patients with chronic ventilatory failure: a physiologic study. Chest 2002; 122: 2105 – 2114
38 *Goldberg AI, Frownfelter D.* The Ventilator-assisted Individuals Study. Chest 1990; 98: 428 – 433
39 *Ambrosino N, Vianello A.* Where to perform long-term ventilation. Respir Care Clin N Am 2002; 8: 463 – 478
40 *Lindsay ME, Bijwadia JS, Schauer WW, Rozich JD.* Shifting care of chronic ventilator-dependent patients from the intensive care unit to the nursing home. Jt Comm J Qual Saf 2004; 30: 257 – 265
41 *Dettenmeier PA.* Planning for successful home mechanical ventilation. AACN Clin Issues Crit Care Nurs 1990; 1: 267 – 279
42 *Leger P, Laier-Groeneveld G.* Infrastructure, funding and follow-up in a programme of noninvasive ventilation. Eur Respir J 2002; 20: 1573 – 1578

43 *Meecham Jones DJ, Paul EA, Jones PW et al.* Nasal pressure support ventilation plus oxygen compared with oxygen therapy alone in hypercapnic COPD. Am J Respir Crit Care Med 1995; 152: 538 – 544
44 *Clini E, Sturani C, Rossi A et al.* The Italian multicentre study on noninvasive ventilation in chronic obstructive pulmonary disease patients. Eur Respir J 2002; 20: 529 – 538
45 *Budweiser S, Jorres RA, Riedl T et al.* Predictors of Survival in COPD Patients With Chronic Hypercapnic Respiratory Failure Receiving Noninvasive Home Ventilation. Chest 2007; 131: 1650 – 1658
46 *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: 453 – 462
47 *Budweiser S, Heinemann F, Fischer W et al.* Long-term reduction of hyperinflation in stable COPD by non-invasive nocturnal home ventilation. Respir Med 2005; 99: 976 – 984
48 *Holland AE, Denehy L, Ntoumenopoulos G et al.* Non-invasive ventilation assists chest physiotherapy in adults with acute exacerbations of cystic fibrosis. Thorax 2003; 58: 880 – 884
49 *Iber C, Ancoli-Israel S, Chesson A, Quan S.* The AASM Manual for the Scoring of Sleep and Associated Events: Rules, Terminology and Technical Specifications. 1. Ed. Westchester, Illinois: American Academy of Sleep Medicine, 2007
50 *Quinnell TG, Pilsworth S, Shneerson JM, Smith IE.* Prolonged invasive ventilation following acute ventilatory failure in COPD: weaning results, survival, and the role of noninvasive ventilation. Chest 2006; 129: 133 – 139
51 *Windisch W, Vogel M, Sorichter S et al.* Normocapnia during nIPPV in chronic hypercapnic COPD reduces subsequent spontaneous PaCO2. Respir Med 2002; 96: 572 – 579
52 *McEvoy RD, Pierce RJ, Hillman D et al.* Nocturnal Non-Invasive Nasal Ventilation in Stable Hypercapnic COPD: A Randomised Controlled Trial. Thorax 2009; 64: 561 – 566
53 *Leger P, Bedicam JM, Cornette A et al.* Nasal intermittent positive pressure ventilation. Long-term follow-up in patients with severe chronic respiratory insufficiency. Chest 1994; 105: 100 – 105
54 *Midgren B, Olofson J, Harlid R et al.* Home mechanical ventilation in Sweden, with reference to Danish experiences. Swedish Society of Chest Medicine. Respir Med 2000; 94: 135 – 138
55 *Windisch W, Dreher M.* NIV and chronic respiratory failure secondary to restrictive thoracic disorders. In: Muir J, Ambrosino N, Simonds AK, Eds. European Respiratory Monograph. Sheffield: ERS Journals, 2008: 240 – 250
56 *Shneerson JM, Simonds AK.* Noninvasive ventilation for chest wall and neuromuscular disorders. Eur Respir J 2002; 20: 480 – 487
57 *Schönhofer B, Sonneborn M, Haidl P et al.* Comparison of two different modes for noninvasive mechanical ventilation in chronic respiratory failure: volume versus pressure controlled device. Eur Respir J 1997; 10: 184 – 191
58 *Restrick LJ, Fox NC, Braid G et al.* Comparison of nasal pressure support ventilation with nasal intermittent positive pressure ventilation in patients with nocturnal hypoventilation. Eur Respir J 1993; 6: 364 – 370
59 *Tejeda M, Boix JH, Alvarez F et al.* Comparison of pressure support ventilation and assist-control ventilation in the treatment of respiratory failure. Chest 1997; 111: 1322 – 1325
60 *Windisch W, Storre JH, Sorichter S, Virchow JCJ.* Comparison of volume- and pressure-limited NPPV at night: a prospective randomized cross-over trial. Respir Med 2005; 99: 52 – 59
61 *Tuggey JM, Elliott MW.* Randomised crossover study of pressure and volume non-invasive ventilation in chest wall deformity. Thorax 2005; 60: 859 – 864
62 *Janssens J, Pépin J, Guo YF.* NIV and chronic respiratory failure secondary to obesity. In: Muir J, Ambrosino N, Simonds AK, Eds. European Respiratory Monograph. Sheffield: ERS Journals, 2008: 251 – 264
63 *Olson AL, Zwillich C.* The obesity hypoventilation syndrome. Am J Med 2005; 118: 948 – 956
64 *Mokhlesi B.* Positive airway pressure titration in obesity hypoventilation syndrome: continuous positive airway pressure or bilevel positive airway pressure. Chest 2007; 131: 1624 – 1626
65 *AASM.* Sleep-related breathing disorders in adults: recommendations for syndrome definition and measurement techniques in clinical research. The Report of an American Academy of Sleep Medicine Task Force. Sleep 1999; 22: 667 – 689
66 *Banerjee D, Yee BJ, Piper AJ et al.* Obesity hypoventilation syndrome: hypoxemia during continuous positive airway pressure. Chest 2007; 131: 1678 – 1684
67 *Resta O, Guido P, Picca V et al.* Prescription of nCPAP and nBIPAP in obstructive sleep apnoea syndrome: Italian experience in 105 subjects. A prospective two centre study. Respir Med 1998; 92: 820 – 827
68 *Sullivan CE, Berthon-Jones M, Issa FG.* Remission of severe obesity-hypoventilation syndrome after short-term treatment during sleep with nasal continuous positive airway pressure. Am Rev Respir Dis 1983; 128: 177 – 181
69 *Berger KI, Ayappa I, Chatr-Amontri B et al.* Obesity hypoventilation syndrome as a spectrum of respiratory disturbances during sleep. Chest 2001; 120: 1231 – 1238
70 *Rochester DF, Enson Y.* Current concepts in the pathogenesis of the obesity-hypoventilation syndrome. Mechanical and circulatory factors. Am J Med 1974; 57: 402 – 420
71 *Cazzolli PA, Oppenheimer EA.* Home mechanical ventilation for amyotrophic lateral sclerosis: nasal compared to tracheostomy-intermittent positive pressure ventilation. J Neurol Sci 1996; 139 Suppl: 123 – 128
72 *Bourke SC, Bullock RE, Williams TL et al.* Noninvasive ventilation in ALS: indications and effect on quality of life. Neurology 2003; 61: 171 – 177
73 *Finder JD, Birnkrant D, Carl J et al.* Respiratory care of the patient with Duchenne muscular dystrophy: ATS consensus statement. Am J Respir Crit Care Med. 2004; 170: 456 – 465
74 *Barthlen GM.* Nocturnal respiratory failure as an indication of noninvasive ventilation in the patient with neuromuscular disease. Respiration 1997; 64 Suppl 1: 35 – 38
75 *Winterholler M, Claus D, Bockelbrink A et al.* Empfehlungen der bayerischen Muskelzentren in der DGM zur Heimbeatmung bei neuromuskulären Erkrankungen Erwachsener. Nervenarzt 1997; 68: 351 – 357
76 *Farrero E, Prats E, Povedano M et al.* Survival in amyotrophic lateral sclerosis with home mechanical ventilation: the impact of systematic respiratory assessment and bulbar involvement. Chest 2005; 127: 2132 – 2138
77 *Kleopa KA, Sherman M, Neal B et al.* Bipap improves survival and rate of pulmonary function decline in patients with ALS. J Neurol Sci 1999; 164: 82 – 88
78 *Lo Coco D, Marchese S, Pesco MC et al.* Noninvasive positive-pressure ventilation in ALS: predictors of tolerance and survival. Neurology 2006; 67: 761 – 765
79 *Hill NS.* Ventilator management for neuromuscular disease. Semin Respir Crit Care Med 2002; 23: 293 – 305
80 *Mellies U, Ragette R, Dohna Schwake C et al.* Long-term noninvasive ventilation in children and adolescents with neuromuscular disorders. Eur Respir J 2003; 22: 631 – 636
81 *Simonds AK.* NIV and neuromuscular disease. In: Muir J, Ambrosino N, Simonds AK, Eds. European Respiratory Monograph. Sheffield: ERS Journals, 2008: 224 – 239
82 *Raphael JC, Chevret S, Chastang C, Bouvet F.* Randomised trial of preventive nasal ventilation in Duchenne muscular dystrophy. French Multicentre Cooperative Group on Home Mechanical Ventilation Assistance in Duchenne de Boulogne Muscular Dystrophy. Lancet 1994; 343: 1600 – 1604
83 *Yuan N, Skaggs DL, Dorey F, Keens TG.* Preoperative predictors of prolonged postoperative mechanical ventilation in children following scoliosis repair. Pediatr Pulmonol 2005; 40: 414 – 419
84 *Bach JR.* Successful pregnancies for ventilator users. Am J Phys Med Rehabil 2003; 82: 226 – 229
85 *Schlamp V, Karg O, Abel A et al.* Nichtinvasive intermittierende Selbstbeatmung (ISB) als Palliativmaßnahme bei amyotropher Lateralsklerose. Nervenarzt 1998; 69: 1074 – 1082
86 *Bach JR, Bianchi C, Aufiero E.* Oximetry and indications for tracheotomy for amyotrophic lateral sclerosis. Chest 2004; 126: 1502 – 1507
87 *Bach JR, Alba AS, Bohatiuk G et al.* Mouth intermittent positive pressure ventilation in the management of postpolio respiratory insufficiency. Chest 1987; 91: 859 – 864
88 *Aboussouan LS, Khan SU, Banerjee M et al.* Objective measures of the efficacy of noninvasive positive-pressure ventilation in amyotrophic lateral sclerosis. Muscle Nerve 2001; 24: 403 – 409
89 *Bourke SC, Tomlinson M, Williams TL et al.* Effects of non-invasive ventilation on survival and quality of life in patients with amyotrophic

lateral sclerosis: a randomised controlled trial. Lancet Neurol 2006; 5: 140 – 147
90 *Peysson S, Vandenberghe N, Philit F et al.* Factors predicting survival following noninvasive ventilation in amyotrophic lateral sclerosis. Eur Neurol 2008; 59: 164 – 171
91 *Pinto AC, Evangelista T, Carvalho M et al.* Respiratory assistance with a non-invasive ventilator (Bipap) in MND/ALS patients: survival rates in a controlled trial. J Neurol Sci 1995; 129 Suppl : 19 – 26
92 *Winterholler M.* Behandlung der Sialorrhoe bei beatmeten Patienten. Pneumologie 2008; 62 Suppl 1: S39 – S42
93 *Bach JR, Ishikawa Y, Kim H.* Prevention of pulmonary morbidity for patients with Duchenne muscular dystrophy. Chest 1997; 112: 1024 – 1028
94 *Bach JR, Baird JS, Plosky D et al.* Spinal muscular atrophy type 1: management and outcomes. Pediatr Pulmonol 2002; 34: 16 – 22
95 *Bach JR, Bianchi C, Vidigal-Lopes M et al.* Lung inflation by glossopharyngeal breathing and 'air stacking' in Duchenne muscular dystrophy. Am J Phys Med Rehabil 2007; 86: 295 – 300
96 *Bach JR.* Update and perspective on noninvasive respiratory muscle aids. Part 2: The expiratory aids. Chest 1994; 105: 1538 – 1544
97 *Gomez-Merino E, Bach JR.* Duchenne muscular dystrophy: prolongation of life by noninvasive ventilation and mechanically assisted coughing. Am J Phys Med Rehabil 2002; 81: 411 – 415
98 *Mustfa N, Aiello M, Lyall RA et al.* Cough augmentation in amyotrophic lateral sclerosis. Neurology 2003; 61: 1285 – 1287
99 *Sancho J, Servera E, Marin J et al.* Effect of lung mechanics on mechanically assisted flows and volumes. Am J Phys Med Rehabil 2004; 83: 698 – 703
100 *Wallgren-Pettersson C, Bushby K, Mellies U, Simonds A.* 117th ENMC workshop: ventilatory support in congenital neuromuscular disorders – congenital myopathies, congenital muscular dystrophies, congenital myotonic dystrophy and SMA (II) 4 – 6 April 2003, Naarden, The Netherlands. Neuromuscul Disord 2004; 14: 56 – 69
101 *Fauroux B, Aubertin G, Lofaso F.* NIV and chronic respiratory failure in children. In: Muir J, Ambrosino N, Simonds AK, Eds. European Respiratory Monograph. Sheffield: ERS Journals, 2008: 272 – 286
102 *Mellies U, Dohna-Schwake C.* Pediatric Pulmonary Function Testing – Neuromuscular Disorders. In: Hammer J, Ebner E, eds. Prog Respir Res. Basel, Freiburg: Karger, 2005: 233 – 246
103 *Mellies U, Dohna-Schwake C, Voit T.* Respiratory function assessment and intervention in neuromuscular disorders. Curr Opin Neurol 2005; 18: 543 – 547
104 *Li KK, Riley RW, Guilleminault C.* An unreported risk in the use of home nasal continuous positive airway pressure and home nasal ventilation in children: mid-face hypoplasia. Chest 2000; 117: 916 – 918
105 *Villa MP, Pagani J, Ambrosio R et al.* Mid-face hypoplasia after long-term nasal ventilation. Am J Respir Crit Care Med 2002; 166: 1142 – 1143
106 *Simonds AK, Ward S, Heather S et al.* Outcome of paediatric domiciliary mask ventilation in neuromuscular and skeletal disease. Eur Respir J 2000; 16: 476 – 481
107 *Ward S, Chatwin M, Heather S, Simonds AK.* Randomised controlled trial of non-invasive ventilation (NIV) for nocturnal hypoventilation in neuromuscular and chest wall disease patients with daytime normocapnia. Thorax 2005; 60: 1019 – 1024
108 *Dohna-Schwake C, Podlewski P, Voit T, Mellies U.* Non-invasive ventilation reduces respiratory tract infections in children with neuromuscular disorders. Pediatr Pulmonol 2008; 43: 67 – 71
109 *Eagle M, Baudouin SV, Chandler C et al.* Survival in Duchenne muscular dystrophy: improvements in life expectancy since 1967 and the impact of home nocturnal ventilation. Neuromuscul Disord 2002; 12: 926 – 929
110 *Mellies U, Dohna-Schwake C, Stehling F, Voit T.* Sleep disordered breathing in spinal muscular atrophy. Neuromuscul Disord 2004; 14: 797 – 803