Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure*
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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 homeventilated patient.
- To compile a set of criteria for quality control of HMV.
Respiratory System:

![Diagram showing the respiratory system with nodes for Lung, Compartment, and Respiratory pump. The Lung node has subnodes for Pulmonary failure, leading to Partial insufficiency (PaO₂↓, PaCO₂↓). This is further divided into Oxygen therapy. The Compartment node has a subnode for Dysfunction, leading to Blood gas analysis. The Respiratory pump node has a subnode for Ventilatory failure, leading to Global insufficiency (PaO₂↓, PaCO₂↑), and Mechanical ventilation.]

Fig. 1 The respiratory system.
Ventilatory Failure:

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)

Causes:
cerebral respiratory dysfunction, neuromuscular disorders (NMD), thoracic deformities, chronic obstructive pulmonary disease (COPD), obesity hypoventilation syndrome (OHS)

- Acute – Chronic – Acute on Chronic (ABG)
**Symptoms**: Underlying disease + dyspnea, morning headache and symptoms of sleep-disordered breathing

- Electively be introduced to HMV
- Nocturnal ventilation is usually alternated with intervals of spontaneous breathing during the day
- Invasively via a tracheostoma or non-invasively via a facial mask
- Improve blood gases during both mechanical ventilation and the subsequent intervals of spontaneous breathing; the main objective is to reduce the partial pressure of arterial carbon dioxide (PaCO2), with normocapnia being the best case scenario
Technical Installation

✓ In life-supporting ventilation, or for patients unable to remove their own face mask, a ventilation machine with an internal battery is required

✓ 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

✓ If the duration of mechanical ventilation exceeds 16 hours/day, an additional identical ventilator must be provided

Tubing and Exhalation Systems:
Single tube systems = ‘leakage’ system

Ventilation Interfaces:
✓ Nasal masks, oronasal masks, full-face masks, mouth masks or mouth pieces
✓ Every patient should possess at least one reserve mask (relieve contact pressure zones)
✓ tracheostoma must be stable, which generally corresponds to being epithelia lized. 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
<table>
<thead>
<tr>
<th>Active</th>
<th>Passive</th>
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<tbody>
<tr>
<td>Bubble-through humidifiers</td>
<td>Pass-over humidifiers</td>
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<tr>
<td>Air flows through water</td>
<td>Heat and Moisture Exchanger (HME)</td>
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<tr>
<td>Sterile water required</td>
<td>Air passes over water</td>
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<td></td>
<td>Conserves patient’s own humidity and airway temperature</td>
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<td></td>
<td>Sterile water not required</td>
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<td></td>
<td>Can alter breathing mechanics</td>
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<td></td>
<td>Must never be used in conjunction with an active humidifying system!</td>
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</table>
Accessories:

- Unit-side particle filters fitted at the point of air inlet are necessary - filters are changed at 1–7 day intervals

- Pulse oximetry is not mandatory **exceptions**: patients with NMD and cough insufficiency children. Drop in oxygen saturation can prematurely indicate imminent, significant secretion retention

- High-performance, battery-supplied suction devices (flow rate > 25 litres/min in replacement machine and ventilation bag
Set-up, Adjustment and Control of the Ventilator

- Centre for Home Mechanical Ventilation

- **Diagnostics:**
  1. History And physical examination
  2. Basic labs and electrocardiogram (ECG)
  3. Blood gas analyses (day and night) during ambient air breathing and with oxygen supply, respectively, or continuous overnight transcutaneous measurement of CO2 (PTcCO2).
  4. Pulmonary function tests; measurement of respiratory muscle function and assessment of peak cough flow (if applicable)
  5. X-ray of the thorax in two planes
  6. Overnight polygraphy/polysomnography
  7. Exercise test (e.g. 6 minute walking test, ergometry)
  8. Echocardiography if cardiac co-morbidity is suspected

- Overnight oximetry alone is neither sufficient to detect nocturnal hypoventilation, nor to indicate HMV
Launching Home Mechanical Ventilation:

- 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 PTcCO2 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).

- SaO2 < 90% or PaO2 < 55mmHg under optimal ventilation indicate the need for additional oxygen supply (LTOT).

- 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.

- During the course of the initialization, the effectiveness of the ventilation should be assessed via PaCO2, both during spontaneous breathing and ventilation, respectively, and supplemented by nocturnal measurements (polygraphy/pulse oximetry, polysomnography, PTcCO2, selective blood gas analyses).
**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 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.
Prerequisites for Discharge from Hospital:

1. only when the underlying and secondary illness(es) are deemed stable
2. provision of the necessary equipment, resources and materials have been secured

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.
COPD:

☑️ Indications: symptoms of CRF and a reduced quality of life are present (at least 1 criterion must be fulfilled):

1. Chronic daytime hypercapnia with PaCO2 ≥ 50mmHg
2. Nocturnal hypercapnia with PaCO2 ≥ 55mmHg
3. Stable daytime hypercapnia with 46–50mmHg and a rise in PTcCO2 to ≥ 10mmHg during sleep [49].
4. 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
5. Following an acute exacerbation needing ventilatory support, according to clinical estimation

☑️ Contraindication:

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).
Procedure:

1. Controlled ventilation mode with ventilation pressures from 20 to 40 mbar. Pressure escalation until normocapnia or maximum tolerance is reached.
2. Rapid increase in inspiratory pressure (0.1 to 0.2 seconds).
3. PEEP can be useful for assisted- or assisted-controlled ventilation.
4. Minimal duration of therapy: 4.5 hours/day.
5. The introduction of NIV in the hospital can take up to two weeks.
Restrictive Thoracic Diseases

NIV is the primary treatment option for restrictive thoracic disease patients with:

1. Scoliosis
2. Kyphosis
3. Pectus carinatum (pigeon chest)
4. Pectus excavatum (concave chest)
5. Ankylosing spondylitis
6. Restrictive pleural diseases
7. Post-tuberculosis syndrome
8. Post-traumatic thoracic deformities
9. Post-operative thoracic deformities (thoracoplastic)
**Indications:**

Symptoms of CRF and a reduced quality of life are present (at least 1 criterion must be fulfilled)

1. Chronic daytime hypercapnia with $\text{PaCO}_2 \geq 45\text{mmHg}$
2. Nocturnal hypercapnia with $\text{PaCO}_2 \geq 50\text{mmHg}$
3. Daytime normocapnia with a rise in $\text{PTcCO}_2$ of $\geq 10\text{mmHg}$ during the night

Patients without manifest hypercapnia but with severe, restrictive ventilatory dysfunction ($\text{VC} < 50\%$ predicted), must undergo a short-term (within 3 months) clinical control examination including polygraphy
Fig. 3  Non-invasive ventilation (NIV) therapy approach in patients with hypercapnic restrictive thoracic diseases (RTD).
Procedure:

1. NIV in pressure- and volume-limited modes is feasible
2. With set pressure, maximal ventilation pressure often reaches 20–25 mbar
3. Changeover from set pressure to set volume should be taken into account in order to improve ventilation
4. EPAP is generally not necessary if bronchial obstructions are absent
Obesity Hypoventilation Syndrome

☑ CPAP and NIV are the primary treatment options for OHS patients with CRF

☑ 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:

1. A ≥ 5 minute-long increase in nocturnal PTcCO2 > 55mmHg and in PaCO2 ≥ 10 mmHg, respectively, in comparison to the awake state.

or

2. 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.

3. If the first control visit (including polysomnography 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.
Fig. 4  Continuous positive airway pressure (CPAP) and non-invasive ventilation (NIV) therapy approach in obesity-hypoventilation syndrome patients (OHS).
Procedure:

1. Titration of CPAP pressure until hypoventilation is eliminated
2. For NIV therapy, increase EPAP until obstructions are eliminated accompanied by titration of inspiratory pressure.
3. 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 polysomnographical control
4. Weight loss should be part of the long-term treatment plan
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
**Indications for NIV:**

at least 1 criterion should be fulfilled:

1. Chronic daytime hypercapnia with $\text{PaCO}_2 \geq 45\text{mmHg}$
2. Nocturnal hypercapnia with $\text{PaCO}_2 \geq 50\text{mmHg}$
3. Daytime normocapnia with a rise in $\text{PTcCO}_2$ of $\geq 10\text{mmHg}$ during the night
4. 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 $\text{VC} < 60\%$ target value and $\text{FEV1} < 40\%$ target value, respectively or during pregnancy with restricted lung function, as well as palliative care of dyspnea.
Fig. 5  Non-invasive ventilation (NIV) therapy approach in patients with neuromuscular diseases (NMD).

0. NMD with presumptive symptomatic respiratory muscle weakness or VC < 70% pred.?

1. Daytime PaCO₂ ≥ 45 mmHg or Nocturnal PaCO₂ ≥ 50 mmHg or PTₐCO₂ change ≥ 10 mmHg or rapid VC decline
   - yes → Suitability for NIV?
   - no → Consider re-evaluation during follow up

2. Suitability for NIV?
   - yes → Efficient NIV possible?
   - no → no

3. Efficient NIV possible?
   - yes → Long-term NIV
   - no → no

4. Long-term NIV

5. Approval of invasive ventilation?
   - yes → Invasive ventilation
   - no → no

6. Invasive ventilation

7. Palliative care
**Procedure:**

Specific aspects in the ventilation of patients with NMD comprise:

1. Muscle weakness in the oropharyngeal area, carrying the risk of reduced ability or complete inability to close the mouth

2. Bulbar symptoms with the risk of recurrent aspiration

3. Hypersalivation; therapy with anti-cholinergics (e.g. Scopolamine patch, amitryptiline or botulinum toxin injections into the salivary glands)

4. Coughing weakness, with the development of acute decompensation
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):

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

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.

Measures to eliminate secretions should therefore be taken when \( \text{SaO}_2 < 95\% \), or a 2–3% drop in the patient’s individual best value occurs.
Fig. 6  Flow chart for secretion management in non-invasively ventilated patients with neuromuscular diseases (NMD).

1. PCF > 270 L/min?
   - Yes: Regular re-evaluation
   - No: Air stacking

3. Air stacking

4. PCF > 270 L/min?
   - Yes: Apply air stacking
   - No: Manually assisted coughing with/without air stacking

6. Manually assisted coughing with/without air stacking

7. PCF > 270 L/min?
   - Yes: Continuation of manually assisted coughing with/without air stacking
   - No: Test for suitability of mechanical insufflator/exsufflator?

9. Test for suitability of mechanical insufflator/exsufflator?
   - Yes: Effective mechanical insufflator/exsufflator?
   - No: Consider mini-tracheostomy or tracheostomy with secretion management via tracheal cannula

10. Effective mechanical insufflator/exsufflator?
    - Yes: Apply mechanical insufflator/exsufflator
    - No: Consider mini-tracheostomy or tracheostomy with secretion management via tracheal cannula

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

<table>
<thead>
<tr>
<th>1. Lung Diseases</th>
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<tbody>
<tr>
<td>- Cystic Fibrosis</td>
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<td>- Bronchopulmonary Dysplasia</td>
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<tr>
<th>2. Neuromuscular Disorders</th>
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<tbody>
<tr>
<td>- Duchenne’s muscular dystrophy</td>
</tr>
<tr>
<td>- Spinal muscular atrophy</td>
</tr>
<tr>
<td>- Congenital muscular dystrophy</td>
</tr>
<tr>
<td>- Myotonic dystrophy</td>
</tr>
<tr>
<td>- Myopathy (congenital, mitochondrial, storage diseases)</td>
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<thead>
<tr>
<th>3. Diseases and Syndromes with Primary and Secondary Thoracic Deformities</th>
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<tbody>
<tr>
<td>- Asphyxiating thoracic dystrophy</td>
</tr>
<tr>
<td>- Achondroplasia</td>
</tr>
<tr>
<td>- McCune-Albright Syndrome</td>
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<tr>
<td>- Cerebral palsy</td>
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<tr>
<td>- Meningomyelocele</td>
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<tr>
<th>4. Disorders of Central Respiratory Regulation</th>
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<tbody>
<tr>
<td>- Congenital central hypoventilation (Undine Syndrome)</td>
</tr>
<tr>
<td>- Acquired central hypoventilation after trauma, encephalitis or CNS degeneration</td>
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<tr>
<td>- Hydrocephalus with increased cranial pressure</td>
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<td>- Arnold Chiari malformation</td>
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<tr>
<th>5. Obesity Hypoventilation Syndrome</th>
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<tr>
<td>- Morbid alimentary obesity</td>
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<tr>
<td>- Prader-Willi Syndrome</td>
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<tr>
<th>6. Diseases with primary, unrectifiable obstruction of the upper airway (when CPAP-therapy is inadequate)</th>
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<tbody>
<tr>
<td>- Down Syndrome</td>
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<tr>
<td>- Mitochondriopathies</td>
</tr>
<tr>
<td>- Mid-facial hypoplasias (Pierre-Robin Syndrome and others)</td>
</tr>
<tr>
<td>- Morbid alimentary obesity</td>
</tr>
<tr>
<td>- Prader-Willi Syndrome</td>
</tr>
</tbody>
</table>
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 awareness, 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 (\textit{Table 3}) [22].

\begin{table}[h]
\centering
\begin{tabular}{|l|l|}
\hline
\textbf{SaO}_2 \textgreater 95\% & No intervention required \\
\hline
\textbf{SaO}_2 \text{between} 90\% \text{and} 95\% & Intensification of ventilation with a mask and/or assisted coughing \\
\hline
\textbf{SaO}_2 \text{< 90\% despite mechanical ventilation} & Contact the centre for HMV \\
\hline
\end{tabular}
\caption{Protocol for pulse oxymeter.}
\end{table}