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Patient selection and techniques for home mechanical ventilation

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Report of the SEPCR Rehabilitation Working Group (now European Respiratory Society Rehabilitation and Chronic Care Scientific Group. Chairman: C.F. Donner; Secretary: H.T.M. Folgering) based on a Consensus Conference held in Arona (Italy) on February 7–8, 1991, organized by the Clinica del Lavoro Foundation, Pavia, Italy.

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Home mechanical ventilation (HMV) is a method for treating stable chronic respiratory failure in the home, rather than in the hospital. Traditionally, it has been conducted through tracheostomy and intermittent positive pressure ventilation (IPPV), but the development of new non-invasive methods has widened the application. Although many of these latter ideas are in the developmental stage, it is appropriate to review the present state of the art. This document will review the goals of HMV, and will describe the new techniques in relation to traditional tracheostomy and IPPV, the selection of patients, and the outcome and support systems necessary.

Home mechanical ventilation will only be successful if the patient is in a stable clinical and psychological state. All acute disturbances must have been effectively treated, and should they worsen will usually demand re-admission to hospital.

The horizons of HMV were widened when it was realized that most patients with chronic respiratory failure require only nocturnal ventilation. After overnight correction of blood gases, clinical improvements are sufficiently sustained during the day to dispense with daytime ventilatory support. For those patients requiring more than nocturnal ventilation, the duration of daily ventilation is proportional to the severity of the disease. The greater the duration of HMV assistance, the greater the demand on the financial resources and family and/or caregiver support. For the more severe cases (ventilation >20 $h \cdot day^{-1}$), patient stay in hospital or in a specialized institution might be preferable, or unavoidable.

The goals of home mechanical ventilation

The major objectives of HMV can be summarized as: 1) improvement of survival; 2) improvement of quality of life; 3) decrease in days of hospitalization, and 4) improvement in the cost-benefit ratio of patient care and social relationships.

The outcome indicators are an improvement of blood gases, better exercise tolerance, less fatigue, improved sleep quality, longevity and quality of life.

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Methods of ventilation

Chronic respiratory failure is a terminal event in a number of respiratory disorders (table 1). Mechanical ventilation is used in attempting to relieve the consequences of disordered blood gases. Traditionally, HMV has been conducted mainly through tracheostomy and IPPV, and in a few cases with negative pressure ventilation. However, new non-invasive methods have been introduced (table 2). The efficacy of such methods remains to be demonstrated, and will inevitably find better application to some disorders than others.

Table 1. – Causes of chronic respiratory failure

Disorders of breathing control
1. Idiopathic disorders of respiratory centres (*i.e.* Ondine's course, idiopathic alveolar hypoventilation)
2. Disorders of respiratory centres secondary to other diseases (*i.e.* encephalitis, circulatory disorders, trauma)
3. Disorders of chemosensitivity or feed-back control (*i.e.* carotid body dysfunction, neuropathies, severe impairment of cardiac output)

Disorders ot the chest wall
1. Muscular disorders (*i.e.* myopathies, dystrophies, amyotrophic lateral sclerosis, Guillain-Barré syndrome, poliomyelitis)
2. Skeletal disorders (*i.e.* kyphoscoliosis, ankylosing spondylitis, thoracoplasty)
3. Pleural diseases (*i.e.* fibrothorax, malignancy, asbestosis)
4. Obesity and other structural abnormalities

Parenchymal disorders
1. Upper airway diseases (*i.e.* laryngeal and tracheal stenosis, obstructive sleep apnoea syndrome)
2. Chronic obstructive pulmonary disease, cystic fibrosis and bronchiectasis
3. Interstitial lung diseases (*i.e.* sarcoidosis, extrinsic alveolitis, occupational lung diseases)
4. Vascular disorders (*i.e.* thromboembolism, pulmonary hypertension)

Table 2. – Non-invasive methods of ventilation

Negative intermittent pressure ventilation
1. Iron lung
2. Cuirass
3. Poncho
4. Jacket suit

Positive pressure ventilation
1. IPPV administered *via*
   - facial mask
   - mouthpiece
   - nasal mask
2. CPAP
3. BiPAP

IPPV: intermittent positive pressure ventilation; CPAP: continuous positive airway pressure; BiPAP: bilevel positive airway pressure.

The present position of tracheostomy + IPPV

For many years the usual method of treating chronic respiratory failure was tracheostomy and IPPV (TIPPV). It was introduced before the days of modern oxygen therapy, so that many patients with hypoxaemia alone, as well as with hypoxaemia and hypercapnia, were treated. TIPPV is effective in treating respiratory failure and is the gold standard against which other techniques must be judged. The invasiveness of the method inevitably gave rise to complications and posed problems of maintaining effective support care systems in the home.

Tracheostomy is helpful in achieving mucus clearance, reducing respiratory dead space and providing effective oxygenation. Tracheostomies are frequently undertaken for treatment of acute exacerbations, but are used thereafter for chronic care. Elective tracheostomy for long-term IPPV was performed for some cases of chronic respiratory failure, with or without alveolar hypoventilation. A number of complications could theoretically limit long-term use, particularly for chronic obstructive pulmonary disease (COPD), but their real incidence is probably low. Immediate complications are haemorrhage, aspiration, surgical emphysema, and local tissue damage. Longer term complications are tracheomalacia, severe haemorrhage, tracheal stenosis, recurrent bronchial sepsis with an increase of Gram-negative septicaemia, tracheal obstruction and tracheal disconnection, particularly at night. The size of tracheostomy cannula is quite critical and must be close-fitting. It may be cuffed or preferably cuffless. Cuffless ventilation demands higher minute volumes of 1–3 *l* to compensate for the leaks. Special attention must be given to choosing a cannula, to allow good speech quality when disconnected. Fenestrated cannulas have to be avoided where possible. Ventilatory appliances such as a humidifier or moisture exchanger are often required when nose humidification is bypassed.

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Non-invasive methods

Non-invasive methods have been introduced in an attempt to avoid tracheostomy and to simplify correction of disordered blood gases for long periods

in the home. They are more demanding of patient cooperation, and doubts remain as to the consistency of blood gas correction over long periods.

a) Negative intermittent pressure ventilation

In this form of respiratory assistance the whole body, not just the thoracic cage, is enclosed in a rigid container, where the pressure is cyclically changed from atmospheric to negative by a piston pump. A negative alveolar pressure is thus created, sucking air into the lungs.

The effectiveness is proportional to the amount of the trunk enclosed in the device. This is maximum for the iron lung, which encloses the whole body up to the neck, but is uncomfortable. Less cumbersome, but somewhat less efficient devices, are the cuirass, or a plastic shell enclosing thorax and abdomen, and the jacket suit or poncho, a nylon garment worn over a rigid shell and closed by a strap around the neck, wrists and legs.

The method demands complete relaxation by the patient. Leaks are frequent, especially around the neck, and chronic use can induce severe skin irritations and thoracic deformities. During sleep, obstructive sleep apnoeas may be induced, especially in COPD, but also in myopathies.

Since the development of non-invasive positive pressure techniques, negative intermittent pressure ventilation is likely to be considered a treatment of second choice.

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b) Nasal access for IPPV (NIPPV)

Easy non-invasive nasal access is possible, but requires the mask to be close-fitting and strapped to the face, as air leaks are the principal problem. Individual silicone-moulded masks, or special commercial nasal masks in selected sizes, are available. Comfort is essential. Ventilation is less effective when leaks occur through the mouth during sleep, and checks should always be made. The problem can usually be solved using chin straps, the references further discuss the issues when this fails.

During exacerbations, reinforcements of the ventilation by NIPPV can often delay or avoid intubation.

The principal side-effects are patient rejection, disturbance of sleep, air leakage around the mask and gastrointestinal distension. The main risk is insufficient ventilation.

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c) Mouthpiece and IPPV

Positive pressure ventilation through a mouthpiece (non-sealed during the day, sealed at night) has been successfully employed particularly in post-poliomyelitis patients.

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Different positive pressure modes

Control (totally ventilator-determined) and assist control (patient-triggered) modes are traditional methods of ventilation management. Continuous positive airway pressure (CPAP) and bilevel positive airway pressure (BiPAP) are more recent additions.

a) Continuous positive airway pressure

Continuous positive airway pressure is applied through a nasal mask, and is mainly used for the

treatment of obstructive sleep apnoea, the major disorder of sleep pathology. Positive pressure in the upper airways prevents pharyngeal collapse and increases functional residual capacity (FRC). The aim is to completely abolish apnoea and/or snoring in every sleep stage and body position. The treatment has proved effective in a large number of patients, relieving symptoms, reducing mortality and reducing the cardiovascular complications associated with the disease.

About 8% of patients complain of rhinitis or mucosal dryness, which are usually reversed by the addition of a humidifier. Other frequent side-effects are skin irritation or abrasions from the facial mask, conjuctivitis due to mask leakage, and aerophagia. Many of these problems can be avoided by specialised units.

b) Bilevel positive airway pressure

This new device is very similar to a pressure support ventilator. It permits differentiation of the pressure administered during the inspiratory (IPAP) and expiratory (EPAP) phases; the latter being effectively a positive end-expiratory pressure (PEEP).

Initiation of the cycle is through a flow trigger to a pre-set pressure, able to increase the spontaneous tidal volume (V_T) of the patient commensurate with his effort. Termination of inspiration is triggered by low inspiratory flow (approximately 30% of the maximum flow) or by timed sequence.

Leaks around the mask and through the open mouth are compensated by the system. The ventilation can be set in control mode, in assist mode, or in an intermittent mandatory position, and applied to both tracheotomy and nasal mask.

The disadvantages are the absence of alarms, and uncomfortable mucosal dryness if substantial flows are delivered through the nose when the mouth is opened. Efficacy in cases with very poor compliance of the thoracic cage remains to be demonstrated. Battery versions are not available at present.

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Clinical indications

Specific diseases of childhood and infancy

Non-invasive IPPV is not generally suitable for young children <2 yrs, and tracheostomy remains the usual method of ventilation. In older children, experiences with non-invasive IPPV is encouraging, but incomplete. Intermittent positive pressure breathing (IPPB) can be used to stretch the thoracic cage.

Congenital bronchodysplasia is a disorder of premature infants, causing respiratory failure. Intubation or tracheostomy are mandatory. Specialist units, probably no more than two or three per 50 million of the population are needed. Good improvements in ventilation can be achieved in selected patients, so that by the middle years of childhood a limited number can be progressively weaned to intermittent ventilation, supplemental oxygen therapy and finally decannulation. Home management, after the acute phase of treatment in the Intensive Care Unit (ICU), involves a number of prerequisites: no bronchospasm, constant oxygen supply, supervised nutritional care (permitting a normal development), a supportive family, and a positive educational programme.

All equipment used in the home should be duplicated. PEEP must not be used; high ventilatory pressures are required to increase FRC. Enormous effort and resources are required for a successful outcome.

Myopathies developing <2 yrs of age often pose difficulties of precise diagnosis. Bio-ethical problems occur when patients, once started on ventilation, do not progress to a stable state. Some myopathies are progressive and lead to a tragic outcome for both the patient and his family. Nevertheless, a number of patients have been successfully introduced to mechanical ventilation and have progressed to require only part time ventilation during the 24 h period. The major difficulty at the present time is to identify those likely to have a poor outcome before supportive ventilation is applied.

The requisites for home treatment are the same as those described for congenital bronchodysplasia. Common symptoms and signs of impending respiratory difficulties are a weak cough, poor chest clearance, difficulty in swallowing, indrawn thoracic cage during inspiration, blood gas abnormalities and poor vital capacity.

Spinal Muscular Amyotrophy (Werdnig Hoffmann disease) is a condition with extremely variable survival. If the onset of the disease is after 6 months of age, 40 yrs of survival may be achieved. Respiratory function during the natural history is extremely variable, but vital capacity, in the absence of airway disease, is a reasonable indicator of the severity of the restrictive disorder. Sudden death is not unusual.

Ventilation in all childhood conditions is most efficient when performed through tracheostomy and intermittent positive pressure ventilation, but the

complications, particularly when a life span of over 40 yrs is possible, have to be borne in mind.

In cystic fibrosis respiratory failure occurs in early childhood or in adolescence. The treatment of acute respiratory failure (ARF) by mechanical ventilation through tracheostomy or intubation has proved extremely difficult. NIPPV seems effective in a proportion of cases but is best used to extend survival up to the date of lung transplantation. Long-term mechanical ventilation is not a successful alternative.

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Dystrophy - Duchenne

In this disease, respiratory failure develops during childhood and is the usual cause of death after 15 yrs of age, often in association with terminal respiratory infection.

Ventilation is imperative for those in respiratory failure. After recovery, or in the case of chronic hypoventilation, HMV is possible. There can then be an assessment of the disturbed mechanical properties of the lungs and chest wall, the extent of ventilation/perfusion imbalance, and of sleep-related problems. The terminal stages are often complicated by gross discomfort from deformities, cardiac problems, particularly arrhythmias from unrelieved hypoxaemia and gastrointestinal and swallowing disorders. This progressive deterioration must be expected from the onset of HMV.

Negative pressure ventilation has been applied successfully as well as NIPPV, but there is doubt as to whether the longer term results are as good as immediate application of tracheostomy and IPPV. It may prove satisfactory to start HMV with NIPPV and to reserve tracheostomy for a later stage, but this sequence is not assured.

Trials of preventive ventilation have been conducted but, surprisingly, have had to be abandoned as the treated group showed an adverse survival profile. The reasons are quite unclear, but cautions against application of mechanical ventilation to such patients. Further research is important to better define patients selection for HMV.

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Amyotrophic lateral sclerosis (ALS)

This is a progressive disorder with an average survival of 3–4 yrs from the onset of symptoms. Respiratory failure can occur at any time and sudden death during the night is not uncommon. Sometimes ARF is consequent to inhalation of gastric content, or after an overwhelming infection, even when muscular ability is still reasonable.

Long-term mechanical ventilation can provide years of extended life for selected people with ALS. Mechanical ventilation poses great ethical difficulties. Usually respiratory support for ALS is non-elective, instituted as an emergency measure: few people elect to it in advance, but many who cannot be weaned after intensive care prefer to continue ventilatory support. The role of the doctor should probably be secondary, explaining to the patient and/or relatives the risks and benefits of HMV, and the patient should have the final choice. Maximum voluntary ventilation (MVV), maximal inspiratory pressure (MIP), maximal expiratory pressure (MEP) and vital capacity (VC) are the best indicators of respiratory muscle strength.

Preliminary results with NIPPV are encouraging, particularly when there is little bulbar involvement. IPPV *via* tracheostomy should be used when there is significant bulbar involvement (clearance of secretions, swallowing, aspiration) or when NIPPV is unsatisfactory. It is considered when hypercapnia is progressive, VC is <30% predicted, MVV <30% predicted, MIP <30 cmH_2O. Financial resources and family caregivers are important prerequisites.

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High level quadriplegia: C1-C4

High level quadriplegia, usually a consequence of an accident to a young adult, or during an adult's life, can now be managed at home.

TIPPV is the usual method of ventilation. Non-invasive methods have recently been introduced and are theoretically preferable as the patients have normal lungs, low resistances, but poor ability to manage their own bronchial toilette through an artificial airway. There are few studies to support this suggestion. Prevention of atelectasis and assisted clearance of secretions should be performed by periodic hyperinflation up to 80% of theoretical vital capacity. Pulmonary complications are often insidious and blood gas monitoring is useful.

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Poliomyelitis, kyphoscoliosis, sequelae of tuberculosis and other restrictive disorders

In this group of adult patients, some of the best results of HMV are obtained, particularly in patients with normal lungs who are ventilatory-restricted by thoracic cage distortion or impaired neuromuscular control. Survival may be extended up to 30 yrs.

Mild hypoxaemia is treated first with oxygen supplementation. TIPPV was formerly recommended when hypercapnia reaches 50–55 mmHg (6.7–7.3 kPa). Non-invasive methods are replacing tracheostomy. Negative intermittent pressure ventilation is difficult to apply to kyphoscoliosis patients because of the distorted thoracic anatomy. Specially shaped plastic ponchos have produced the best results.

NIPPV through a nasal mask seems more effective and acceptable, and recent studies have shown equal blood gas correction to tracheostomy. Studies of longer term use (>4 yrs) are not available.

Parenchymal restrictive disease

Parenchymal restrictive disease, such as occupational and non-occupational lung fibrosis, cystic and vascular disease, compose a mixed group of conditions of generally adverse prognosis, but having the potential for a useful outcome of NIPPV. Serious reduction of lung compliance makes for difficulties in effective ventilation. Few studies are available.

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COPD

Long-term domiciliary oxygen fails to arrest the decline of arterial oxygen tension (Pao_2) in COPD patients. In advanced hypercapnic respiratory failure, mechanical ventilatory assistance, either mandatory or elective, has been investigated. The outlook for tracheostomy and IPPV is little better than using supplemental oxygen alone. Ten year survivals are extremely rare, but disease severity in these retrospective studies are very different, making for difficult comparisons. Generally speaking, experienced centres see few patients who gain real benefit

from long-term tracheostomy. Quality of life improves little.

The aims of treatment are to improve blood gases, achieve muscle rest, reduce nocturnal hypoxaemia, reduce breathlessness and perhaps improve the oxygen and CO_2 sensitivity in the control of ventilation.

Tracheostomy, with or without IPPV, frequently induces complications. Tolerability and acceptability of NIPV seems poor at best. NIPPV offers the best hope of benefit, even if the increased dead space ventilation requires high tidal volumes to be delivered in a short time. The risk of an air leakage, is thereby increased, reducing the blood gas response.

The usefulness of mechanical ventilation in ARF patients with chronic bronchitis, is not matched by an improvement in the long-term. Research should try to identify special groups likely to show a worthwhile response.

Bronchiectasis

Progressive bronchial infection is associated with the decline of pulmonary function and respiratory failure. In end-stage disease, tracheostomy facilitates bronchial toilette, but survival at 2 yrs is inevitably very rare.

Surprisingly, NIPPV performed at night-time has shown benefits both in sputum clearance and survival. After 22 months, half of the patients in one study survived considerably better than controls (P. Leger *et al.*, Unpublished data). Further studies are clearly required.

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Sleep-disordered breathing

Abnormalities occur as a result of primary sleep pathology and as a consequence of neuromuscular disorders or COPD.

Definitions of respiratory sleep disorders are contentious, but five groups can be identified: 1) obstructive sleep apnoea; 2) central sleep apnoea (often overestimated); 3) primary alveolar hypoventilation (a very rare disease); 4) secondary alveolar hypoventilation (a contentious group); and 5) primary snoring.

The most common disorder is obstructive sleep apnoea, a condition of recurrent pharyngeal obstruction which disrupts normal sleep rhythms. An obstruction leading to cessation of respiration for 10s or more is considered significant.

The important consequences are nocturnal desaturation, daytime sleepiness and loss of mental acuity. Criteria for defining the severity of obstructive sleep apnoea are not agreed, but generally obstructions $>20 \cdot h^{-1}$ and saturations <80% carry a high risk of cardiovascular complications, such as systemic hypertension and cardiac disrhythmia. Accidents through daytime sleepiness can occur in up to 30% of these individuals. Such patients should move straight to CPAP therapy. If the apnoea index is <20 episodes$\cdot h^{-1}$ and nocturnal desaturation between 80–90% conservative treatment (weight loss, no evening alcohol and good sleep hygiene) should be tried first.

A precise diagnosis of sleep pathology and control of CPAP treatment is possible only after formal polysomnography. In order to avoid submitting all patients with suspicious symptoms to full investigation, questionnaires, nocturnal pulse oximetry and recording of tracheal air sounds are being developed as a means of preselection. These preselection tests should not be used as a means of making a diagnosis of sleep pathology or prescribing treatment.

CPAP is the treatment of choice. Tracheostomy is an alternative treatment but is seldom used nowadays. Surgery of the pharynx and soft palate has no proven long-term benefits, and may have long-term adverse effects. It should be regarded as a research procedure. Snoring may be corrected for several years but the natural history of the sleep pathology is unchanged. Mandibular surgery can be very effective in selected patients, but here again long-term evaluation is not yet available.

Nocturnal NIPPV after failure of CPAP, the importance of nocturnal desaturation particularly in the rapid eye movement (REM) sleep phase, and the prognostic relevance in COPD and neuromuscular disorders, remain under investigation.

Primary snoring is a common condition carrying no implication of sleep pathology. If symptoms suggestive of sleep disorders are recognized then these are the signals for preselection tests and, if necessary, polysomnography. Snoring itself is not an indication for full polysomnography.

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Home Care Organization and costs

Home mechanical ventilation cannot be successfully deployed without the provision of a home care service, a supporting family or other care givers, provision of education and training for patients and family, psychosocial support and provision of the necessary finance.

Countries have different organisations for their national health services. In Europe, health care is predominantly provided by the State with a variable amount of private assistance. In the United States, the system is largely free market with a large number of interwoven bodies of a charitable, state or private nature.

Home respiratory care is a specialized technique and the general practitioner alone cannot manage it successfully. It will be most successful if co-ordinated through a national system of home respiratory care responsible for all respiratory procedures in the domestic environment. These will include occupational therapy, physiotherapy, supplemental oxygen, mechanical ventilation, psychosocial care and nutrition. These modalities are best organized through a designated home care regional or central service, responsible for co-ordinating the activities of consultant, general practitioner, home nurses and technicians undertaking installation and maintenance of th equipment. A new grade of respiratory care therapist could combine the duties of a nurse and a respiratory physiotherapist, with some technical expertise. The service should be supported by psychosocial care and a nutritional programme, particularly for infants and young children. The psychosocial service will prepare the family for home care, providing information about the disease and its natural history and hygiene, prophylaxis and help with follow-up. The use of all devices (including speaking valves for tracheostomized patients), and supplies must be fully understood by the patient.

Safety aspects

The patients on HMV and their caregivers should be trained for emergency procedures in the case of ventilatory malfunction. According to the specific needs of the single patient (totally ventilator-dependent or requiring only partial ventilatory support) the caregivers should be able to: 1) use a manual resuscitator; 2) connect the patient to a back-up ventilator; 3) contact emergency personnel (ambulance service, equipment supplier, physicians, other health personnel).

The home care service must provide minimum service or replacement periods for essential equipment, delay not usually exceeding 12 h. In the few patients disadvantaged by such periods, equipment may need to be duplicated. Emergency telephone systems are helpful for crises and for those patients with few relatives.

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