

Home Respiratory Management: From COPD to Neurological Diseases^{*1}

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Significance of Respiratory Management

Difficulty breathing leads directly to a life-threatening condition, representing a serious issue in clinical practice. Respiratory failure is defined as a functional disorder of the respiratory system showing an arterial oxygen tension (PaO₂) of 60 Torr or less in the absence of oxygen administration. Physical findings of respiratory failure include tachypnea, tachycardia, engorgement and edema of the jugular vein, and cyanosis. Emaciation, depression of the clavicular fossa due to hypertrophied accessory respiratory muscles, shoulder breathing, and pursed-lip breathing may also be observed as characteristic findings. If the patient lapses into CO₂ narcosis, headache or disturbed consciousness ranging from subtle personality changes to confusion or coma may occur. Respiratory management is aimed at reversal of the pathological systemic and mental states derived from respiratory failure, toward restoration of an acceptable physiological condition.

Respiratory Management and Its Current Status in Japan

Home respiratory management includes home oxygen therapy (HOT) and home mechanical ventilation (HMV). HOT comprises inhalation of oxygen at home by patients with advanced chronic respiratory failure due to cyanotic congenital heart disease or various other causes, those with pulmonary hypertension, or those

with chronic heart failure (**Table 1**).

HMV is defined as mechanical ventilation at home in patients who are in a stable disease state and who depend on continuous long-term mechanical ventilation. Patients on HMV are mainly those who have neuromuscular diseases, chronic obstructive pulmonary disease (COPD), or sequelae of pulmonary tuberculosis. HMV uses a positive pressure ventilator by which air is delivered to the airway via positive pressure or a negative/positive pressure external ventilator with a cuirass respirator.¹

Positive pressure ventilation is widely used in Japan; this procedure is divided into invasive mechanical ventilation that involves tracheostomy, i.e., tracheostomy intermittent positive pressure ventilation (TPPV), and noninvasive mechanical ventilation, i.e., positive pressure ventilation (NPPV) by which ventilation is implemented through a mask covering the nose and mouth or the entire face. According to a questionnaire survey by the Japanese Respiratory Society, patients with neuromuscular disease account for 72% of all those on TPPV, whereas patients on NPPV had COPD (26%), sequelae of pulmonary tuberculosis (23%), neuromuscular disease (18%), sleep apnea syndrome (14%), and so on.² Continuous positive airway pressure (CPAP) at home is used for patients with sleep apnea syndrome (**Table 1**). The major advantage of TPPV is that a patent airway can reliably be maintained, but the implementation rate of this method is only about 5% because of its invasiveness.

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Table 1 Methods of respiratory management and indications

	HOT	NPPV	TPPV	CPAP
Chronic Obstructive Pulmonary Disease (COPD)	⊙	⊙	○	
Sequelae of pulmonary tuberculosis	⊙	⊙	○	
Lung cancer	○			
Pulmonary fibrosis/interstitial pneumonia*	⊙	○	○	
Kyphoscoliosis		○	○	
Neuromuscular disease**	○	⊙	⊙	
Congenital heart disease	○			
Diffuse panbronchiolitis	○	○	○	
Angiogenic pulmonary hypertension	○	○	○	
Cheyne-Stokes breathing due to chronic heart failure	○			
Pulmonary thromboembolism	○			
Alveolar hypoventilation syndrome		○	○	
Sleep apnea syndrome		⊙	○	⊙

⊙ Recommended, ○ Indicated

* Interstitial pneumonia: idiopathic pulmonary fibrosis, pneumoconiosis, sarcoidosis, farmer's disease, etc.

** Neuromuscular disease: muscular dystrophy, myasthenia gravis, ALS, spinal progressive muscular atrophy, Parkinson's disease/related diseases, multiple system atrophy, sequelae of poliomyelitis, sequelae of cerebrovascular disorder, Werdnig-Hoffmann disease, phrenic nerve paralysis, etc.

Indications and Practical Aspects of HOT

HOT is indicated for patients with a PaO_2 of 55 Torr or less and those with a PaO_2 of 60 Torr or less who have marked hypoxemia during sleep or exercise. Among the beneficial effects of HOT, it was reported to alleviate subjective symptoms, reduce the burden on the cardiorespiratory system, and lead to mental stability, resulting in improved quality of life (QOL), reported as “enabling the patient to go out” or “decreasing hospitalizations.”²

Indications for and Practical Aspects of NPPV

The introduction of HMV is planned when control of respiratory failure is assumed to have become difficult. Because NPPV is noninvasive, it is common for NPPV to be introduced first. Timing of the introduction of NPPV is as follows: when the patients have subjective symptoms such as morning headache, sleeplessness, and fatigue, when signs of cor pulmonale or dyspnea on exertion occur, or when day-and-night hypoventilation symptoms or hypercapnia occur. If

compensation as evidenced by labored respiration is present, it is necessary to avoid exhaustion of respiratory muscles by early introduction of NPPV even in patients whose laboratory values are within the normal ranges.

Bilevel PAP is a method of assisting ventilation in concert with the patient's breathing employing two pressures, i.e., an inspiratory positive airway pressure (IPAP) of 8-20 cmH_2O and an expiratory positive airway pressure (EPAP) of 0-4 cmH_2O . Nose, nose-mouth, and face masks are available. For smooth introduction of this method, use of the mask should be restricted, i.e. kept within a short period of time, and then gradually prolonged in the early introductory phase, until the mask becomes usable during the night. The general initial settings are as follows: spontaneous/timed (S/T) mode, IPAP 6-8 cmH_2O , EPAP 4 cmH_2O , respiratory rate 12/min, and maximum percent inspiratory time (%IPAPmax) 30-40%. This procedure is worth conducting even in patients with progressive disease who will require respiratory management with TPPV in the future, because QOL is improved by relieving respiratory discomfort albeit temporarily and because the patient is given additional time to mentally prepare for starting TPPV.

Indications for and Practical Aspects of TPPV

Neuromuscular disease is the most frequent reason for patients being on TPPV. In recent years, downsized turbine-type mechanical ventilators have become dominant, allowing patients to go out or travel using portable batteries if their conditions are stable. Although the mean survival time is about 6 years in patients with amyotrophic lateral sclerosis (ALS) using TPPV, some may survive for more than 10 years. Therefore, approaches to achieving better palliative care and improvements in QOL are extremely important.³

Positive end-expiratory pressure (PEEP) and fraction of inspiratory oxygen ($F_{I}O_2$) are important elements in condition setting for mechanical ventilation. The level of PEEP must be set so as to prevent pulmonary collapse, while minimizing oxygen administration. In condition setting for acute lung injury, a tidal volume of 6 mL/kg and a peak airway pressure of less than 30 cmH₂O are recommended.

Synchronized intermittent mandatory ventilation (SIMV) is a method of providing mandatory ventilation intermittently in synchronization with spontaneous breathing. In addition to a pressure support level of 10 cmH₂O and a PEEP of 4–10 cmH₂O, mechanical ventilation management should be performed by SIMV with pressure controlled ventilation (PCV) at 15–18 cmH₂O, a ventilation rate of 10 breaths/min, and an inspiratory time of 1.5 s.

The frequency of regular exchange of the tracheal cannula is usually about once a week. However, in cases with marked expectoration likely to cause airway obstruction, the cannula should be exchanged on additional occasions, and the frequency of regular exchange should be reconsidered. Because the use of disposable circuits has become more common recently, the ventilator circuit is usually exchanged once a month. It is desirable to use minimum cuff pressure of the tracheal cannula to avoid air leakage during inspiration, the risk of aspiration during body motion, ischemic injury to the lining membrane of the trachea, and so on. The instruction should be to set a target pressure that gives “the same hardness as the ear lobe,” preferably not more than 30 cmH₂O.

To treat intratracheal granulation or granulation at the tracheal orifice, adrenocortical

steroids should be used in an early stage, and external antibiotics should be applied if there is accompanying bleeding. Consultation with an otorhinolaryngology specialist is recommended for cases showing marked granulation because endoscopic observation and surgical treatment are required. When pneumothorax, pneumomediastinum, or subcutaneous emphysema has occurred, proper insertion of the cannula into the trachea should be confirmed, and the conditions of the ventilator be adjusted to properly maintain the airway pressure at 20 cmH₂O or less. Stress including communicative disorders due to prolonged respiratory management may cause gastrointestinal ulceration, and administration of antiulcer drugs such as proton pump inhibitors should thus be considered.

When the alarm of the ventilator is frequently activated, the patient’s respiratory status should be confirmed first, and the ventilator and accessories should then be examined for defects. The setting of the ventilator should, in fact, be double-checked. If there are no abnormalities, the alarm level should be adjusted. However, prudence is required in altering the alarm level because this may lead to overlooking problems in the patient or the device.

Double-lumen cannulas and speech cannulas have recently become available, and are now conducive to home TPPV management. The use of intra-cannula continuous quantitative suction devices and cough assist devices decreases the risk of aspiration and respiratory infection in patients on HMV. In view of the importance of securing batteries as recognized through the experience of the Great East Japan Earthquake Disaster, the favorable appraisal that external batteries are now covered by health insurance has been obtained.

Items Necessary for Home Respiratory Management

At the time of introducing home respiratory management, various actions such as administering drug therapy, instructions on the use of devices, respiratory rehabilitation, diet/nutrition therapy, and infection prevention measures are taken to the maximum extent feasible, necessitating adequate preparation and training of the patient and family. Home medical care aims to reduce the need for hospitalization by stabilizing

the disease state while paying attention to the aims and requests of the patient, and to support the patient in leading an adequately satisfying life by providing a good environment for treatment at home, to achieve the goal of QOL improvement for the patient and family.

Respiratory rehabilitation involves modification of the breathing pattern and flexibility training. In whole-body endurance and muscle strength training, specific and easily understandable instructions should be given to facilitate incorporation of an exercise habit into the patient's lifestyle. When the disease has progressed, programs designed for protectively and palliatively maintaining physical strength and reducing the burden of nursing care should be implemented, without causing undue strain on the patient. To enhance the effect of rehabilitation training and to suppress weight loss that progresses along with decreasing respiratory function, combined use of nutrition therapy is important. Beginning with oral care and diet therapy, artificial feeding management should be considered.⁴ Needless to say, influenza and pneumococcal vaccinations are important in the infection prevention strategy.

It is important to take advantage of regional health resources and formulate plans for the coordination of primary care doctors and home-visit nursing care stations via the regional medi-

cal network. According to a questionnaire survey conducted by the Japanese Respiratory Society, institutions that provide doctors' visits to patients receiving home TPPV account for 52%, prescription of home-visit nursing care 92%, and use of helpers or public health nurses 74%.² Prompt establishment of a home management system that includes the home-visit medical care system, rehabilitation training at home, home-visit nursing care, and telemedicine (remote medical care), based on the active use of social resources such as the Law for the Welfare of the Physically Disabled Persons (physical disability certificate) and long-term care insurance, is desirable to reduce the burdens on both patients and their family members.

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