Ventilatory Support for Chronic Respiratory Failure

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Ventilatory Support for Chronic Respiratory Failure

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Introduction

The concept of ventilatory support is not new. It has been reported that BC Egyptians and Greeks described the theories of respiration. The Old Testament (800 BC) tells us that the Prophet Elisha induced mouth to mouth pressure breathing in a dying child! Much later, in the sixteenth century Theophrastus Bombastus von Hohenheim, or the “Famoso Doctor” Paracelsus, also known as the “wandering spirit,” used the fire bellows of the time as devices for assisted ventilation: they were connected to the patients by a tube inserted in the mouth. In the following centuries, medical pioneers such as Vesalius, Hooke, Fathergill, and Hunter, among others, continued to advance the concept of ventilatory support, while others in the engineering field developed devices to expand its application.

However, it is the Scandinavian polio epidemics of the 1950s that gave rise to a new era of mechanical ventilation defined by the realization that many patients could benefit from it. This was coupled with the emergence of blood gas analysis and of a new medical discipline, that is, Respiratory Intensive Care. The description of the adult respiratory distress syndrome by Ashbaugh et al. in 1967, and the many studies thereafter on the treatment of this syndrome and related conditions led to an ever increasing use of mechanical ventilation with IPPV during the next 40 years, primarily to treat respiratory failure resulting from acute situations.

Meanwhile, years of productive research have demonstrated that patients with chronic respiratory insufficiency can also benefit from mechanical ventilation. As the Preface of this volume mentions “their survival as well as their health status” may be dependent on long-term ventilatory support. The ever increasing incidence and prevalence of chronic respiratory disease suggests that the use of ventilatory support will markedly increase. However, the techniques and strategies to use it, and when and where (non-intensive care unit, or home), are very different from treating the respiratory failure resulting from acute conditions and in patients with structurally near normal lungs.

This new volume of the series of monographs Lung Biology in Health and Disease edited by Nico Ambrosino and Roger Goldstein is truly a “how to” apply and monitor ventilatory support in patients with chronic respiratory failure treated in an ICU, in the hospital, or at home. It is really a “must read” for health professionals who care for such patients. The editors have called upon experts from many countries to contribute the many subjects presented in this volume. As a result, perspectives from different countries and cultures are considered.
The ultimate goal of this series of monographs is to contribute to better care of the many patients worldwide with chronic respiratory diseases: this is just the goal of this volume! I am grateful to the editors and authors for the opportunity to present such an important and timely contribution.

Claude Lenfant, M.D.
Gaithersburg, Maryland, U.S.A.
As our understanding of respiratory failure has deepened, and as the technical aspects of life support have advanced, increasing numbers of patients now survive acute critical illness but are unable to regain complete independence from ventilatory support. Patients with progressive ventilatory insufficiency due to neuromuscular disease or another chronic condition may experience both physiologic benefit and improved quality of life through the institution of part- or full-time mechanical support of ventilation, either invasive or noninvasive. For these and other patients with chronic respiratory failure, ventilatory support has become an established therapy, whose evidence base and practical application have expanded dramatically during the last 20 years. This book perfectly combines both the science and the art of long-term mechanical ventilation, bringing key research findings along with the wisdom of vast experience to the bedside in the care of patients with chronic respiratory failure. It is a remarkable achievement.

To appreciate the pathophysiology of respiratory failure, and to appropriately tailor therapy to the needs of the individual patient, the different components of the illness must be understood and assessed. The degree to which oxygenation, ventilation, airway protection, and secretion clearance are impaired, and what measures are required to manage each of them, are important determinants of where and by whom a particular patient may be cared for. They determine, for example, whether invasive or noninvasive ventilation will be more appropriate for that patient, how likely it is that the patient can be managed successfully at home, and how much external support in the form of equipment and personnel will be required.

Caring for the patient with chronic respiratory failure involves other considerations as well. General physical conditioning and peripheral muscle training, nutritional support, and attention to personal and psychological needs are only some of the important nonrespiratory areas if long-term outcomes are to be optimal. In addition, the perspectives of the patient himself or herself, as well as those of family members and others most closely involved in that person’s ongoing daily care, are vital to successful overall management.

While the needs of individual patients vary, the array of resources and professional skills that must be available for optimal management is consistent regardless of the particular health care system in which those patients are cared for. Respiratory care for patients with chronic respiratory failure is a multidisciplinary,
team enterprise whose members may vary by region, practice setting, or job title, but whose purposes and needs remain the same. The management team may be under the overall direction of a pulmonologist, rehabilitation specialist, or other physician, and may include respiratory therapists, nurses, or physical therapists in varying combination; the apparatus and supplies used for ventilatory support, airway care, and monitoring may be different, provided and maintained through different systems and approaches; how ancillary services and consultation are accessed may vary. Despite these differences, however, all patients receiving ventilatory support for chronic respiratory failure need access to state-of-the-art care provided in the context of best information and up-to-date resources.

This book thoroughly covers all aspects of its subject. After reviewing the pathophysiology and manifestations of chronic respiratory failure and the available approaches to mechanical ventilation, it considers the various aspects of weaning, including how to optimize the likelihood of success and to determine whether complete liberation from ventilatory support is appropriate for a given individual. It takes the patient from the intensive care unit, through the various specialized institutional facilities that may exist in different areas, into the community and the patient’s home. The rationale, evidence, and practical application of different specialized interventions and the components of rehabilitation are covered in detail. Thorough discussion is provided about the personnel involved in long-term mechanical ventilation—their different roles, how they should be trained, and how they work together as a team in meeting the needs of the patient. Separate chapters address available devices and techniques and their optimal application in individual cases, as well as the use of pharmacological agents and the management of secretions. The important topics of quality of life, legal and ethical issues, and end-of-life care are covered in detail. In addition, separate chapters discuss the special circumstances and needs of patients with different underlying or complicating conditions, and different causes of chronic respiratory failure.

Professors Ambrosino and Goldstein have done a masterful job of bringing all this together. The author list—76 authorities in 13 countries who represent every relevant profession and specialty—is essentially a “Who’s Who” of the most respected investigators and clinicians in the field. The book has been conceived and organized so that every aspect is addressed. Clinicians involved in the care of patients with chronic respiratory failure will find here a complete, practical, accessible resource, regardless of their practice setting or the health care system in which they work.

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Preface

Chronic respiratory failure (CRF) is a global issue as, increasingly, patients with both obstructive and restrictive conditions survive longer. In parallel, the intensive care unit (ICU) has enabled major advances in the management of patients with respiratory failure, attributable to acute respiratory and non respiratory conditions. Therefore, an increasing number of patients with chronic respiratory insufficiency become dependent for their survival as well as their health status, on long term mechanical ventilation. Home mechanical ventilation (HMV) is becoming an increasingly relevant option for patients with CRF, encouraged by; the introduction of noninvasive positive pressure ventilation (NIPPV), the recognition of the many different diagnostic categories of patients who can benefit from this approach and the pressures on institutions worldwide, to reduce healthcare costs by reducing in-patient hospitalization. As the population ages, we can expect this issue to increase in importance, challenging society as well as all levels of the healthcare system.

This book, is designed to address the growing need for information on long term ventilation in CRF. It is structured in nine parts, beginning with introductory chapters on chronic respiratory failure as a global problem, broad principles of acute and chronic ventilation and the prevalence of the major diagnostic categories. The text then moves from the difficult to wean ICU patient, to the newer concept of rehabilitation in the ICU, long-term ventilation in the non ICU settings in hospital and the community to special respiratory and non respiratory considerations of this population. The last three sections provide insights into CRF among different patient groups, perspectives on long-term ventilation by the healthcare professionals, the patient and the family care givers and finally worldwide approaches, encompassing Europe, North and South America and Asia.

The editors and authors hope that this text will assist healthcare professionals interested in this area, by providing an overview of the clinical, economic and ethical challenges to the healthcare system, posed by those requiring long-term ventilation. In this way, it may assist healthcare professionals in addressing the various exciting challenges of caring for this population.

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Chronic Respiratory Failure

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I. Definition

The term respiratory failure describes a condition in which the respiratory system fails in one or both of its principal gas exchange functions: oxygenation and elimination of carbon dioxide. In clinical practice it is conventionally defined as an arterial oxygen tension (PaO₂) <60 mmHg, an arterial carbon dioxide tension (PaCO₂) >45 mmHg, or both, while breathing air. It is important to emphasize that respiratory failure is a laboratory diagnosis and that there is no absolute definition of the levels of arterial PaO₂ and PaCO₂ that indicate respiratory failure: the cutoff levels serve as a general guide, and their significance depends on the history and clinical assessment of patients.

These threshold values are empirically derived estimates of the point at which bulk transport of gases to and from the tissues may become compromised. Thus, 60 mmHg approximates to the inflection point on the normal oxyhemoglobin dissociation curve when small changes in PaO₂ produce large changes in hemoglobin saturation. Similarly, once the PaCO₂ rises above 45 mmHg for any period, the normal blood-buffering capacity will be exceeded and the pH will fall. Unlike hypoxemia, which cannot be physiologically compensated for, renal compensation for CO₂ retention is possible and occurs over a two- to three-day period during which the pH returns to normal (chronic ventilatory failure). The risks of impaired tissue oxygenation are mitigated by acute increases in cardiac output and more chronically by adaptations in the concentrations of 2,3-diphosphoglycerate, which affects the position of the dissociation curve and an increase in the hemoglobin concentration. This secondary polycythemia preserves the oxygen content of arterial blood at a cost in terms of blood viscosity and an increased tendency to thrombosis.

II. Classification

The two principal components of the respiratory system are the lung, which participates in gas exchange, and a muscular pump, which ventilates the lungs (1,2). The ventilatory pump consists of the chest wall (rib cage and abdomen), including the muscles that displace this
structure and thereby inflate and deflate the lung, together with the ventilatory control circuits in the central nervous system, and the pathways that connect controllers with respiratory muscles (spinal and peripheral nerves) provide a self-regulating feedback mechanism that maintains blood gas homeostasis.

Respiratory failure may be classified as hypoxemic (type I) or hypercapnic (type II or ventilatory failure) (3), either of which may be acute and chronic. Hypoxemic respiratory failure is due to failure of the lungs, caused by acute (cardiogenic pulmonary edema, pneumonia, acute respiratory distress syndrome) or chronic (emphysema, interstitial lung disorders) diseases (Tables 1 and 2). It is characterized by hypoxemia with normocapnia or hypocapnia. In these conditions central respiratory drive is high and there is sufficient alveolar ventilation (VA) to eliminate CO₂ and prevent hypercapnia.

Hypercapnic respiratory failure is due to failure of the ventilatory pump caused by acute (drug overdose, acute neuromuscular diseases) or chronic (chest wall abnormalities, chronic neuromuscular diseases) disorders. It is characterized by alveolar hypoventilation, which leads to hypercapnia with coexistent, usually mild, hypoxemia. The central drive may be globally reduced with the fall in PaO₂ resulting from the increase in alveolar CO₂. More commonly, the drive remains high, but the mechanical load on the respiratory system is too great or the capacity of the muscles too low to ensure efficient CO₂ elimination (Fig. 1).

In individual patients, however, both types of respiratory failure may coexist, as one respiratory problem leads to another with a cascade of interaction (3). For example, patients with cardiogenic pulmonary edema or status asthmaticus first develop hypoxemia due to lung failure; if the disease persists or progresses, pump failure and hypercapnia appear because of several mechanisms (increased work of breathing, reduced oxygen delivery, hyperinflation).

Respiratory failure can develop over minutes to hours (acute respiratory failure) or over several days or longer (chronic respiratory failure). The distinction between acute and

---

**Table 1** Causes of Chronic Hypoxemia with Normal or Low PaCO₂

<table>
<thead>
<tr>
<th>Obstructive ventilatory disorders</th>
<th>Mixed ventilatory disorders</th>
<th>Interstitial lung disorders</th>
<th>Pulmonary vascular diseases</th>
<th>Nonpulmonary diseases</th>
</tr>
</thead>
<tbody>
<tr>
<td>COPD</td>
<td>Bronchiectasis</td>
<td>Idiopathic pulmonary fibrosis</td>
<td>Pulmonary vascular hypertension</td>
<td>Severe heart failure</td>
</tr>
<tr>
<td>Chronic asthma</td>
<td>Sequelae of tuberculosis</td>
<td>Pneumoconiosis</td>
<td>Chronic pulmonary thrombosis</td>
<td>Hepatopulmonary syndrome</td>
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<tr>
<td></td>
<td></td>
<td>Sarcoïdosis</td>
<td>Arteriovenous malformations</td>
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<td></td>
<td></td>
<td>Extrinsic allergic alveolitis</td>
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<td></td>
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</tbody>
</table>

**Abbreviations:** PaCO₂, arterial carbon dioxide pressure; COPD, chronic obstructive pulmonary disease.
Table 2  Causes of Chronic Hypoxemia with Hypercapnia

Pulmonary diseases
  Obstructive ventilatory disorders
    COPD
  Mixed ventilatory disorders
    Bronchiectasis
    Sequelae of tuberculosis
  Nonpulmonary diseases
    Dysfunction of respiratory centers
    Primary alveolar hypoventilation
    Obesity hypoventilation syndrome
  Depressant drugs
    Myxoedema
    Lesion of brainstem

Neuromuscular diseases
  Poliomyelitis
  Amyotrophic lateral sclerosis
  Myasthenia gravis
  Muscular dystrophies, polymyositis

Chest wall deformities
  Kyphoscoliosis
  Ankylosing spondylitis
  Chest trauma
  Thoracoplasty

Pleural thickening

Obstruction of upper respiratory tract

Abbreviation: COPD, chronic obstructive pulmonary disease.

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Figure 1  Schematic representation of the balance of forces on the respiratory pump.
Source: Courtesy of J. Moxham.
chronic hypoxemic respiratory failure cannot readily be made on the basis of arterial blood gases. The presence of clinical markers of chronic hypoxemia (polycythemia or cor pulmonale) suggests a long-standing disorder. Acute hypercapnic respiratory failure is characterized by hypercapnia with respiratory acidosis (pH < 7.35), whereas in chronic hypercapnic respiratory failure there is time for renal compensation with increase in bicarbonate concentration. Therefore, the pH usually is normal or only slightly decreased.

III. Pathophysiology of Chronic Respiratory Failure

A. Hypoxemic Respiratory Failure

The pathophysiological mechanisms that account for the hypoxemia observed in a wide variety of diseases are ventilation/perfusion (V/Q) mismatch, shunt, diffusion impairment, and alveolar hypoventilation (4,5). In some areas of the world, living at high altitude further compromises oxygen delivery, and lesser degrees of disease severity can produce clinically alarming degrees of hypoxemia. In most cases of oxygenation failure in patients living at or close to the sea level, V/Q mismatch and varying degrees of right to left shunting are the major causes of hypoxemia. V/Q mismatch develops when there are lung regions with low ventilation relative to their perfusion (low V/Q units), as occurs in chronic obstructive pulmonary disease (COPD) and interstitial lung diseases. An intrapulmonary or intracardiac shunt causes deoxygenated mixed venous blood to bypass ventilated alveoli and results in venous admixture. This condition can occur in patients with arteriovenous malformations but is also seen in intensive care unit (ICU) practice when an acute increase in pulmonary artery pressure can lead to a patent foramen ovale’s reopening with a major effect on PaO2. Diffusion impairment contributes to hypoxemia in conditions characterized by a combination of widened alveolocapillary distance and shortened pulmonary capillary transit time, such as extensive destruction and fibrosis of pulmonary parenchyma, especially when cardiac output is high (as during exercise).

In the absence of underlying lung disease, hypoxemia due to alveolar hypoventilation is associated with normal alveolar-arterial oxygen difference; in contrast, the other three mechanisms are characterized by a widening of alveolar-arterial oxygen gradient, resulting in severe hypoxemia. Hypoxemia due to V/Q mismatch, diffusion impairment, and alveolar hypoventilation can be corrected by administering a low concentration of inspired oxygen, whereas hypoxemia due to shunt cannot be corrected even with a high concentration of inspired oxygen (4,5).

B. Hypercapnic Respiratory Failure

For a given level of CO2 production (VCO2), hypercapnic respiratory failure results only from an inadequate VA. A simple equation describes these relationships quantitatively under steady state conditions:

$$\text{PaCO}_2 = K \frac{\text{VCO}_2}{\text{VA}}$$

where $K$ is the respiratory exchange ratio.
Since VA = minute ventilation (VE) – dead-space ventilation (VD), this equation can be expressed as

\[ \text{PaCO}_2 = K \frac{\text{VCO}_2}{\text{VE} - \text{VD}} \]

or

\[ \text{PaCO}_2 = K \frac{\text{VCO}_2}{\text{VT} \text{Fr}(1 - \text{VD}/\text{VT})} \]

where VT is the tidal volume and Fr is the respiratory frequency (2).

From these equations, it follows that VA decreases and so PaCO2 increases when VE decreases. Likewise, when VE and VD remain unchanged but VT decreases and respiratory frequency (RF) increases (rapid shallow breathing), PaCO2 increases. Patients adopt a rapid shallow breathing pattern to minimize respiratory work per breath, but this form of compensatory behavior can be deleterious to gas exchange and is a major factor producing chronic hypercapnic respiratory failure in patients with COPD and neuromuscular disorders (6–10).

The function of the ventilatory pump is critically dependent on three factors: the respiratory workload, the respiratory muscle strength, and the ventilatory drive (Fig. 1). Chronic hypercapnic respiratory failure can result from one or more of these abnormalities: inadequate ventilatory drive, excessive respiratory load, and inadequate inspiratory muscle strength.

**Ventilatory Drive**

Reduction in the output of the respiratory centers to respiratory muscles leads to reduced VA and to CO2 retention. Although this is the least common of the major causes of ventilatory failure, it can contribute to exacerbation of ventilatory failure resulting from other causes. Acute failure of ventilatory drive most often results from overdoses of sedative or narcotic drugs, especially opiates and benzodiazepines. In patients with other causes of ventilatory pump failure, metabolic alkalosis or administration of excessive oxygen can contribute to reduction in VA and exacerbate hypercapnia. Myxedema due to hypothyroidism (11) and idiopathic congenital central hypoventilation syndrome, in which chemoresponsiveness is reduced or absent when asleep (12), are two conditions characterized by inadequate ventilatory drive that result in chronic hypercapnia. Much more commonly, the onset of normal sleep is accompanied by a reduction in ventilatory responsiveness and a small increase in PaCO2. This reduction in the central drive and in respiratory muscle tone is important when other causes of respiratory failure are only just being compensated for by the waking drive to breathe.

**Respiratory Load**

During spontaneous breathing the inspiratory muscles must generate sufficient force to overcome the elastic and resistive load of the respiratory system. The pressure developed by the inspiratory muscles per breath (Pi) is increased if the elastic (decreased compliance of the lungs or the chest wall) or resistive (airway obstruction) load is increased. Furthermore, in patients with hyperinflation of the chest wall (see below), a substantial effort must be
made by the inspiratory muscles to overcome intrinsic positive end-expiratory alveolar pressure (PEEPi) before any inspiratory airflow can occur (13,14). This threshold load can account for a significant proportion of the respiratory workload in patients with COPD during acute exacerbations or during the weaning process from mechanical ventilation (15). If the pressure required for breathing (PI) becomes greater than 60% of maximum inspiratory pressure (MIP), the load cannot be sustained indefinitely and inspiratory muscles are at risk of fatigue (16). Bellemare and Grassino also observed that the Pi/MIP that can be sustained indefinitely decreases when Ti/Ttot increases and that the product of Pi/MIP and Ti/Ttot (the “tension-time index”) is related to the endurance time (17). When the tension-time index becomes greater than a critical value (0.15 for the diaphragm), there is risk of inspiratory muscle fatigue and pump failure (17).

Respiratory Muscle Strength

The maximum pressure-generating capacity of inspiratory muscles can be impaired by several causes (Table 3). Like all skeletal muscles, the strength of respiratory muscles depends on the length-tension relationship (18). Hyperinflation reduces inspiratory muscle strength by shortening the inspiratory muscles, especially the diaphragm, below their optimum force-producing length (18,19). Neuromuscular disorders can affect respiratory muscles; among patients without intrinsic pulmonary or chest wall disease, chronic hypercapnic respiratory failure usually occurs when the respiratory muscle strength falls below 30% of the predicted value (20). Muscle wasting due to malnutrition does not spare respiratory muscles (21). Several metabolic factors can also reduce the strength of otherwise normal respiratory muscles. Hypercapnia and hypoxemia have been reported to reduce diaphragmatic strength (22) and the endurance of inspiratory muscles (23). Corticosteroid treatment (24), hypocalcemia (25), hypophosphatemia (26), hypokalemia, and hypomagnesemia (27) may be additional contributory factors acting in concert with malnutrition to promote generalized respiratory muscle weakness in patients with chronic respiratory diseases.

<table>
<thead>
<tr>
<th>Table 3</th>
<th>Causes of Reduced Respiratory Muscle Strength</th>
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<tbody>
<tr>
<td>Neuromuscular disorders</td>
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<tr>
<td>Hyperinflation</td>
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<tr>
<td>Malnutrition</td>
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<td>Electrolyte disorders</td>
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<td>Hypophosphatemia</td>
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<td>Hypomagnesemia</td>
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<td>Hypocalcemia</td>
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<td>Hypokalemia</td>
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<td>Hypoxemia</td>
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<td>Hypercapnia</td>
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<tr>
<td>Drugs</td>
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<td>Corticosteroids</td>
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<td>Aminoglycoside antibiotics</td>
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<tr>
<td>Disuse</td>
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<tr>
<td>Controlled mechanical ventilation</td>
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</table>
IV. Some Specific Diseases Associated with Respiratory Failure

A. COPD

Abnormalities of gas exchange are infrequent in COPD at rest before the forced expiratory volume in the first second (FEV1) has fallen to 50% of the predicted value or less. Thereafter, hypoxemia becomes increasingly more frequent, as lung mechanics worsen. The additional worsening of lung mechanics that accompanies an exacerbation can merit acute oxygen therapy, the characteristic increase in static lung volumes being accompanied by lower VA/Q units. These changes resolve slowly during recovery, but PaO2 can improve enough for domiciliary oxygen to be no longer needed. Inappropriately, severe hypoxemia in an exacerbating COPD patient raises the possibility of coincident pathology, such as cardiogenic pulmonary edema or acute pulmonary embolization, which is now more readily diagnosed with a computed tomographic (CT) pulmonary angiogram. Acute increases in low VA/Q units cause hypercapnia and worsening acidosis, which predicts both an increased mortality and the need for ventilatory support.

Although chronic hypercapnic respiratory failure is a common and important event in patients with severe COPD (28), the mechanisms leading to its occurrence are not completely understood. There is a considerable variability in the relationship of PaCO2 to indices of airway obstruction or V/Q mismatch (29,30), suggesting that factors other than lung pathology may be relevant. Neural drive assessed by measuring mouth occlusion pressure (6,7) or electromyographic activity of diaphragm (7) has been found to be higher in both eucapnic and hypercapnic patients with COPD than in normals. Given the increase in VD/VT ratio that characterizes COPD, normocapnia can be maintained only by increasing VE to a sufficiently high level. Mechanical constraint to breathing can, however, cause problems in maintaining a sufficiently high level of ventilation in patients with COPD. On the one hand, the load placed on the inspiratory muscles is increased because of high airflow resistance, reduced dynamic compliance, and presence of dynamic hyperinflation with PEEPi. On the other hand, the pressure-generating ability of respiratory muscles can be impaired because of hyperinflation, malnutrition, drug therapy, and electrolytes abnormalities (31). Bégin and Grassino (8) have shown in a large group of COPD patients that the probability of developing hypercapnia increases with the severity of airway obstruction, obesity, and inspiratory muscle weakness. When the load placed on the respiratory muscle pump becomes excessive in relation to its capacity, patients may avoid respiratory muscle fatigue and pump failure by modifying the breathing pattern. A reduction in VT could allow COPD patients to reduce PI relative to inspiratory muscle strength, thus minimizing respiratory effort and dyspnea, and avoiding fatigue (32). In line with this hypothesis a more rapid and shallower pattern of breathing has frequently been observed in hypercapnic than in eucapnic COPD patients (6,7,33–35). More recently, it has been shown that in stable COPD patients with severe airflow obstruction there was a significant association between hypercapnia and both shallow breathing and inspiratory muscle weakness, these variables explaining more than 70% of variance in PaCO2 (9). In this study VT was related directly to Ti, indicating that a small VT is primarily the consequence of alteration in respiratory timing (9).

The mechanisms leading to alteration in respiratory timing in patients with COPD have not yet been clearly defined. In line with the concept that the perception of inspiratory
effort and dyspnea is closely linked to the PI relative to inspiratory muscle strength (36), it is possible that reduction in Ti and VT involves an integrated response of the respiratory system to the perception of breathlessness. Studies showing an inverse relationship between Ti and PI relative to inspiratory muscle strength, and a significant association of the severity of dyspnea with both the increase in PI and the decrease in Ti, support the above hypothesis (9). In conclusion, it seems evident that patients with COPD alter the pattern of breathing in an attempt to optimize the performance of respiratory muscles, to reduce breathlessness, and to prevent fatigue. The rapid shallow breathing however reduces VA and increases PaCO₂.

**B. Restrictive Disorders**

**Neuromuscular Diseases**

Ventilatory failure, often in association with pneumonia, is a frequent cause of death in many neuromuscular disorders. Severe weakness of the respiratory muscles produces a restrictive pattern with decrease in vital capacity and total lung capacity, whereas functional residual capacity generally tends to be low and the residual volume is within normal limits (37,38). Hypercapnia is likely when respiratory muscle strength falls to 30% of the predicted value (20). Chronic respiratory failure in patients with subacute or chronic neuromuscular diseases is not simply due to the direct effect of weakness of respiratory muscles leading to inability to inflate the lungs and alveolar hypoventilation. A variety of additional factors play a role, including alteration in the mechanical properties of the lung (37,38) and the chest wall (39), respiratory abnormalities during sleep (40–45), and inability to cough (46). Abnormalities during sleep, including frequent arousals, decreased rapid eye movement sleep, hypoventilation, and hypoxemia, are common in patients with neuromuscular diseases (40–45), particularly in those with severe diaphragmatic weakness (42,45). These abnormalities usually precede and probably contribute to daytime ventilatory failure (40,43,44). The effectiveness of cough is reduced in patients with neuromuscular diseases because of both inspiratory and expiratory muscle weakness. Inspiratory muscle weakness affects the inspiratory phase of cough and expiratory muscle weakness reduces the cough-induced dynamic compression and hence the linear velocity of airflow through the large intrathoracic airways (46). As a result the clearance of secretions is defective in these patients, thus contributing to the high prevalence of bronchopulmonary infections. Finally, in patients with neuromuscular diseases, chronic hypercapnic respiratory failure is associated with rapid shallow breathing leading to alveolar hypoventilation (10), probably as a result of afferent signals in weakened respiratory muscles, intrapulmonary receptors, or both (10,47).

**Thoracic Deformity**

In patients with kyphoscoliosis the severity is quantified by measuring the angle between the upper and lower portions of the spinal curve (Cobb angle). When this angle exceeds 100° (severe scoliosis), the vital capacity falls below 50% of the predicted value (48). A major factor in the pathophysiology of chronic respiratory failure in patients with kyphoscoliosis is the decrease in the compliance of the chest wall and lungs (49,50). In severe scoliosis the compliance of the chest wall may be about 25% of the predicted value.
Furthermore, the spinal deformity causes inefficient coupling between the respiratory muscles and the chest wall, with reduction in the maximum pressure-generating capacity of inspiratory and expiratory muscles (49,51). The imbalance between the increased elastic load and the reduced capacity of respiratory muscles elicits a rapid and shallow pattern of breathing (52). Although this breathing pattern does have the advantage of minimizing the work of breathing, it causes alveolar hypoventilation. Like patients with neuromuscular disorders, patients with kyphoscoliosis develop sleep-disordered breathing, especially during rapid eye movement sleep (53).

Obesity Hypoventilation Syndrome

The obesity hypoventilation syndrome (OHS) was originally described in 1955 in subjects with severe obesity, chronic hypercapnic respiratory failure, polycythemia, hypersomnolence, and right ventricular failure (54). The pathogenesis of OHS is certainly multifactorial in nature and not fully understood. Abnormalities of chest wall mechanics with increased work of breathing, reduction in inspiratory muscle strength, hypoventilation during sleep, and abnormalities in ventilatory control with blunting of both hypercapnic and hypoxic ventilatory responsiveness could explain chronic hypoventilation in these patients (55). Clinically, it is important to distinguish this condition from obstructive sleep apnea (OSA) with overlap. OSA is a common condition, often a result of obesity, and when it coexists with significant airflow obstruction or chronic heart failure, hypoxemia with CO₂ retention can occur. Marked daytime somnolence is characteristic of these patients, and they do well with continuous positive airway pressure rather than ventilatory support, often correcting their gas exchange abnormality as their hypersomnolence resolves.

References


References

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Part IX: Worldwide Approaches to Long-Term Ventilation

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Table 3 Recent Innovations in Mechanical Ventilation

<table>
<thead>
<tr>
<th>Innovation</th>
<th>Clinical Applicability</th>
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<tbody>
<tr>
<td><strong>New Modes</strong></td>
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<tr>
<td>Proportional assist ventilation</td>
<td>Interactive mode that adjusts pressure and flow according to spontaneous flow</td>
</tr>
<tr>
<td>Adaptive support ventilation</td>
<td>Feedback mode that adjusts minute ventilation to patient mechanics</td>
</tr>
<tr>
<td>Neurally adjusted ventilatory</td>
<td></td>
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<tr>
<td>assist</td>
<td>Adjusts interactive support to diaphragm EMG signal</td>
</tr>
<tr>
<td>Computerized pressure support</td>
<td>Adjusts pressure support to the ventilatory pattern and exhaled CO₂</td>
</tr>
<tr>
<td><strong>New Adjusters for Interactive Breaths</strong></td>
<td></td>
</tr>
<tr>
<td>Automatic tube compensation</td>
<td>Adjusts airway pressure to compensate for endotracheal tube resistance</td>
</tr>
<tr>
<td>Pressure slope/rise time</td>
<td>Adjusts rate of pressure rise for synchrony</td>
</tr>
<tr>
<td>Pressure support cycle adjust</td>
<td>Adjusts pressure support flow cycle criteria for synchrony</td>
</tr>
<tr>
<td><strong>New monitors</strong></td>
<td></td>
</tr>
<tr>
<td>Esophageal pressure</td>
<td>Approximates pleural pressure</td>
</tr>
</tbody>
</table>
Trend monitors Allows for data storage

Remote systems Allows for central monitoring

Pressure volume plots Allows for selection of settings that avoid overdistention and collapse/reopening

Spontaneous breathing trials Allows for assessment of discontinuation potential

Abbreviations: EMG, electromyogram; CO₂, carbon dioxide.


17. Kacmarek RM, Pierson DJ, eds. AARC Conference on positive end expiratory pressure. Respir Care 1988; 33:419-527.


30. Banner MJ, Kirby RR, MacIntyre NR. Patient and ventilator work of breathing and ventilatory muscle loads at different levels of pressure support ventilation. Chest 1991; 100:531-533.


32. Marini JJ. Exertion during ventilator support: how much and how important? Respir Care 1986; 31:385-387.


37. Stroetz RW, Hubmayr RD. Patient-ventilator


49. Mead J, Takishima T, Leith D. Stress distribution in


22. Buyse B, Messerman W, Demedts M. Treatment of chronic respiratory failure in kyphoscoliosis: oxygen or


34. Fogarty A, Hubbard R, Britton J. International


4 Chapter 4. Size of the Problem, What Constitutes Prolonged Mechanical Ventilation, Natural History, Epidemiology


82. Epstein SK. Predicting extubation failure: is it in (on) the cards? Chest 2001; 120(4): 1061-1063.

83. MacIntyre NR, Cook DJ, Ely EW Jr., et al, Evidence-based guidelines for weaning and discontinuing ventilatory support: a collective task force facilitated by the American College of Chest Physicians; the American Association for Respiratory Care; and the American College of Critical Care Medicine. Chest 2001; 120(6 suppl):375S-395S.


Chapter 5. Causes of Difficult Weaning: Which Mechanisms Are Associated with Long-Term Ventilator Dependence?


45. Cooper KR, Phillips BA. Effect of short-term sleep loss


123. Holliday JE, Myers TM. The reduction of weaning time


Chapter 6. Weaning Protocols, Including Noninvasive Ventilation


Chapter 7A. Weaning Units: The U.S. Perspective


12. Gracey DR, Naessens JM, Viggiano RW, et al. Outcome of


Chapter 7B. Weaning in a Specialized Facility


34. Indihar FJ. A 10-year report of patients in a prolonged


Chapter 8. Organization of Rehabilitation in the ICU


Figure 10 Patients requiring >21 days MV in 3 separate ICUs across the SWCCN during 2005.

Abbreviation: MV = mechanical ventilation.


11. Pronovost P, Berenholtz S, Ngo K, et al. Developing and


Once Sarah was placed into the long-term phase of the rehabilitation framework, all active
weaning from MV was stopped and the nurse consultant, the lead doctor, and the PT made a comprehensive and holistic assessment. Further advice was sought from the dietician, the speech and language therapist, and the pharmacist. An overall plan was then developed, which included the following: 1. Providing full support for respiratory muscles during and between exercise sessions. 2. Increasing PS, PEEP, and Fi O 2 during exercises. 3. Decreasing ventilatory support only after Sarah demonstrated significant improvement in exercise tolerance, as measured by maximum inspiratory measure (P imax ), modified Borg scale, and functional independence measure. 4. Setting daily goals with Sarah, team, and family. 5. Sarah, dressing in her normal clothes during the day. 6. Not stopping therapy, but adapting it to meet Sarah’s ability and medical condition. 7. Establishing communication strategies through leak speech. 8. Increasing calories when exercise tolerance increased. 9. Establishing following alternative therapies: 1 Pet therapy once per week 1 Music 1 Art 1 Guided imagery 1 Biofeedback 1 Reflexology 10. Monitoring reduced to a minimum.

Specific exercise program: 1. To sit out twice daily, progressing to all day 2. Standing transfers, bed to chair 3. Hourly breathing exercises 4. Resisted upper limb exercises using weights 5. Resisted trunk exercises 6. Treadmill work in the gym, while on portable ventilator. 1 Progression in time or speed as per tolerance-defined as; SpO 2 < 85% or patient fatigue using a dyspnea visual analogue scale. 1 Use of Heliox during treadmill exercise entrained through the ventilator 7. Step ups.

Within three weeks of starting rehabilitation Sarah was weaned and decannulated. Table A1 shows Sarah’s exercise tolerance over the three-week period.

<table>
<thead>
<tr>
<th>Day 1, week 1</th>
<th>4 m (on pressure support ventilation)</th>
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</thead>
<tbody>
<tr>
<td>Day 5, week 1</td>
<td>120 m (on pressure support ventilation)</td>
</tr>
<tr>
<td>Week 2</td>
<td>280 m (on pressure support ventilation)</td>
</tr>
<tr>
<td>Day 18, week 3</td>
<td>108 m (using entrained heliox via ventilator on reduced pressure support)</td>
</tr>
</tbody>
</table>
Day 20, week 3: 67 m on a tracheostomy mask using heliox

Day 21, week 3: 80 m following decannulation

Day 25: 120 m prior to discharge on oxygen only
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10 Chapter 10. Peripheral and Respiratory Muscle Training


46. Williams PE. Use of intermittent stretch in the prevention of serial sarcomere loss in immobilised muscle.


58. Chang AT, Boots RJ, Brown MG, et al. Reduced inspiratory muscle endurance following successful weaning


11 Chapter 11. Transcutaneous Electrical Muscle Stimulation


44. Plankeel JF, McMullen B, MacIntyre NR. Exercise outcomes after pulmonary rehabilitation depend on the initial mechanism of exercise limitation among non-oxygen-dependent COPD patients. Chest 2005;


76. Troosters T, Gosselink R, Decramer M. Chronic obstructive pulmonary disease and chronic heart failure:


86. Brown MD, Cotter MA, Hudlicka O, et al. The effects of different patterns of muscle activity on capillary density, mechanical properties and structure of slow and fast rabbit


97. Gregory CM, Bickel CS. Recruitment patterns in human skeletal muscle during electrical stimulation. Phys Ther


Chapter 12. Psychological Aspects in Patients with Chronic Respiratory Failure


25. Wong HLC, Lopez-Nahas V, Molassiotis A. Effects of


Chapter 13. Definition and Indications for Prolonged Mechanical Ventilation (PMV)

1. Mulligan S. AARC and Gallup estimate numbers and costs for chronic ventilator patients. Am Assoc Respir Care Times 1991; 150:30-36.


11. Lin CC. Comparison between nocturnal nasal positive pressure ventilation combined with oxygen therapy and


33. American College of Chest Physician. Mechanical
ventilation beyond the intensive care unit. Chest 1998; 113(5 suppl):289s-344s.


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pulse oximetry/cutaneous carbon dioxide tension monitoring during colonoscopies: pilot study with a smart ear clip. Digestion 2004; 70:152-158.


67. Lofaso F, Quera-Salva MA. Polysomnography for the management of progressive neuromuscular diseases. Eur


78. Gomez-Merino E, Bach JR. Duchenne muscular dystrophy:


122. McKim DA, LeBlanc C. Maintaining an “Oral Tradition”: specific equipment requirements for mouthpiece ventilation
instead of tracheostomy for Neuromuscular Disease. Respir Care 2006; 51:297-298.


134. Wicks AB, Menter RR. Long-term outlook in quadriplegic


144. Gay PC, Hubmayr RD, Stroetz RW. Efficacy of nocturnal nasal ventilation in stable, severe chronic obstructive


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22 Chapter 22. Legal and Ethical Issues

1. The Ethox Centre, Department of Public Health and Primary Health Care, University of Oxford, www.ethox.org.uk


13. Hardart MK, Burns JP, Truong RD. Respiratory support in spinal muscular atrophy-type I: a survey of physician


Chapter 23. Pharmacological Treatment for Patients with Chronic Respiratory Failure


Chapter 24. Patient-Ventilator Interfaces for Invasive and Noninvasive Ventilation


44. Hill NS. Complications of noninvasive ventilation. Respir Care 2000; 45:480-481.

Chapter 25. Tracheostomy Weaning from Longer Term Ventilation


11. Sinha R, Bergofsky EH. Prolonged alteration of lung mechanics in kyphoscoliosis by positive pressure


15. MacIntyre NR, Cook DJ, Ely EW Jr., et al., American College of Chest Physicians, American Association for Respiratory Care, American College of Critical Care Medicine. Evidence-based guidelines for weaning and discontinuing ventilatory support: a collective task force facilitated by the American College of Chest Physicians; the American Association for Respiratory Care; and the American College of Critical Care Medicine. Chest 2001; 120(Suppl):375S-379S.


33. Vallverdu I, Calaf N, Subirana M. Clinical characteristics, respiratory functional parameters, and outcome of a two-hour T-piece trial in patients weaning from mechanical ventilation. Am J Respir Crit Care Med


42. Hess DR. Noninvasive positive-pressure ventilation and ventilator-associated pneumonia. Respir Care 2005; 50(7):924-929, discussion 929-931.


62. Rossi A. Noninvasive ventilation has not been shown to be ineffective in stable COPD. Am J Respir Crit Care Med 2000; 161(3 pt 1):688-689.


27 Chapter 27. Electrophrenic Respiration


28 Chapter 28. Secretion Management


12. Servera E, Sancho J, Zafra MJ. Cough and neuromuscular


82. Bach JR. Cough in SCI patients. Arch Phys Med Rehabil


29 Chapter 29. The Importance of Overnight Monitoring in the Management of Chronic Respiratory Failure


31 Chapter 31. Nutrition in ICU


24. Sullivan DH, Bopp MM, Roberson PK. Protein-energy undernutrition and life-threatening complications among the


47. ASPEN Board of Directors and the Clinical Guidelines Task Force. Guidelines for the use of parenteral and


Chapter 32. Skin Integrity, Bowel and Bladder Care


Chapter 33. Palliative Care for the Ventilator Patient: End-of-Life Issues and Approaches


11. Desbiens Nam Wu AW. Pain and suffering in seriously ill


36. Guo YF, Sforza E, Janssens JP. Respiratory patterns
during sleep in OHS patients support. Chest 2007; 131:1090-1099.


35 Chapter 35. Progressive Neuromuscular and Degenerative Diseases


21. Sivasothy P, Smith IE, Shneerson JM. Mask intermittent positive pressure ventilation in chronic hypercapnic


31. Windisch W, Kostic S, Dreher M, et al. Outcome of


37 Chapter 37. Ventilation Among the Pediatric Population


38 Chapter 38. The Perspective of Patients


32. Schmid H, Hasenfeld Y. Organizational dilemmas in the provision of home-care services. Soc Serv Rev 1993; 67:40-54.


Chapter 39. The Perspective of Family and Caregivers


8. Writing Angel. Solutions for Living, PO Box 36116, Canton, Ohio, U.S.A., 44735 (888-8845483) www.solutionsforliving.us.


40 Chapter 40. The Perspective of Physicians: The Intensive Care Specialist and the Pulmonary Specialist


7. MacIntyre NR, Cook DJ, Ely EW Jr., et al. Evidence-based guidelines for weaning and discontinuing ventilatory support: a collective task force facilitated by the American College of Chest Physicians; the American Association for Respiratory Care; and the American College of Critical Care Medicine. Chest 2001; 120(6 suppl):375S–395S.


41 Chapter 41. The Perspective of the Allied Health Professionals


Chapter 43. Long-Term Ventilation: The European Perspective


Chapter 44. Long-Term Ventilation: The South American Perspective


46 Chapter 46. Long-Term Ventilation: The Taiwanese Perspective


3. The Ventilator Dependents Managed Care Demonstration, Bureau of National Health Insurance. Available at:


