Noninvasive Positive Airway Pressure in Hypercapnic Respiratory Failure in Noncardiac Medical Disorders

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- Noninvasive positive pressure ventilation
- Chronic obstructive pulmonary disease • Cystic fibrosis
- Kyphoscoliosis • Neuromuscular disease

Noninvasive positive pressure ventilation (NIPPV) is commonly used to improve gas exchange in patients with various noncardiac medical conditions. This article discusses NIPPV in chronic obstructive pulmonary disease (COPD), cystic fibrosis (CF), kyphoscoliosis, and neuromuscular disorders, including amyotrophic lateral sclerosis (ALS), Duchenne muscular dystrophy (DMD), myasthenia gravis (MG), and postpolio syndrome (PPS). NIPPV is typically delivered by bilevel positive airway pressure (BPAP) or, in some cases, ventilatory support other than BPAP.

POSITIVE AIRWAY PRESSURE AS A TREATMENT MODALITY

Positive airway pressure (PAP) is delivered by medical devices that generate either a continuous PAP (CPAP) or a pressure gradient with different inspiratory and expiratory pressures, such as BPAP. BPAP is preferred in acute and chronic hypercapnic respiratory failure because it increases alveolar ventilation.

CPAP is effective in patients with obstructive sleep apnea (OSA) by increasing the extrathoracic airway pressure to a level above the atmospheric pressure, thereby preventing collapse of the upper airway. This process has been called a ''pneumatic splint''.[1]

CPAP may also increase the lung volume by increasing the functional residual capacity (FRC). This volume increase may improve oxygenation but may also increase hypercapnia by increasing respiratory dead space, which generally makes CPAP less effective for hypercapnic respiratory failure.[2]

BPAP may be effective for treating patients with hypercapnic respiratory failure. The inspiratory PAP to expiratory positive airway pressure (EPAP) differential provides a gradient to inflate and deflate the lungs. The EPAP is, in effect, a form of positive end-expiratory pressure that provides some of the benefits of CPAP (increase in FRC to improve oxygenation) combined with ventilatory support to reduce hypercapnia.[2]

NIPPV is delivered to patients through an interface that may include a nasal mask, nasal pillows,

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a full-face mask, or an oral mouthpiece.[3] The complications of using a noninvasive interface, such as pressure sores, are beyond the scope of this article. Detailed methods of titration are also not covered in this article. However, the reader should be aware that inspiratory pressures greater than 20 cm may be required and ventilator support other than BPAP may be needed. Documentation of successful NIPPV is generally obtained through attended polysomnography (PSG), nocturnal monitoring of arterial oxygen hemoglobin saturation (saturation), transcutaneous partial pressure of carbon dioxide ($Paco_2$), or end-tidal CO_2.

PAP IN STABLE COPD

COPD is a treatable and preventable disease that results in airflow obstruction that is not fully (ie, partially) reversible.[4–6] The obstructive component of COPD is documented by spirometry with a post-bronchodilator forced expiratory volume in 1 second/forced vital capacity (FEV_1/FVC) ratio (in liters) of less than 0.70 (**Table 1**). The severity is determined in part by the FEV_1, with severe COPD being an FEV_1 of less than 50% predicted. COPD is primarily the result of cigarette smoking, a common cause of morbidity, and the fourth leading cause of mortality in the United States and the world.[4]

The clinical-pathologic conditions most associated with COPD are one or a combination of chronic bronchitis (chronic cough and sputum production) and emphysema. Symptoms may include dyspnea, COPD exacerbations, and fatigue.

Etiology and Pathophysiology

The pathologic results of exposure to cigarette smoke and the resulting neutrophil airway inflammation may include squamous metaplasia of the respiratory epithelium, respiratory ciliary loss and dysfunction, inflammation and fibrosis of airways, mucous gland hyperplasia and hypersecretion, increased airway smooth muscle, loss of alveolar attachments, and bronchoconstriction from vagally mediated release of acetylcholine.[4]

The differential diagnoses include asthma, bronchiectasis, bronchiolitis, upper airway obstruction (eg, from a tumor), postviral airway inflammation, eosinophilic bronchitis, and congestive heart failure.

Arterial blood gases are generally obtained only in borderline cases of hypoxemia (saturation $\leq$92% by pulse oximetry) and to determine the presence of hypercapnia. Patients with very low FEV_1 levels (<30% predicted) are particularly at risk of having hypercapnia[4–6] and may be candidates for NIPPV.

Treatment of COPD

Pharmacologic treatment is primarily directed at relaxing the airway smooth muscle with bronchodilators. Attempts to reduce inflammation have not been obviously successful, but inhaled corticosteroids and phosphodiesterase inhibition (eg, theophylline) may have a limited anti-inflammatory effect. In addition, nonpharmacologic therapy includes pulmonary rehabilitation, smoking cessation, and long-term oxygen therapy (LTOT). The use

Table 1
Some measures of pulmonary function in obstructive and restrictive lung disease

Pulmonary Function	No Lung Disease	Obstruction	Restriction
FEV_1	80%–120% predicted	Normal or reduced	Normal or reduced
FVC	80%–120% predicted	Normal or reduced	Reduced
FEV_1/FVC	$\leq$0.7[a]	Reduced	Normal or increased
TLC	80%–120% predicted	Normal or increased	Reduced
DLCOsb	80%–120% predicted	Normal in chronic bronchitis, reduced in emphysema, may be increased in asthma	Normal or near normal in chest wall and neuromuscular disease, reduced in interstitial lung disease

Predicted ranges are approximate and should be modified according to specific patient populations. The abnormalities of pulmonary function are generalizations, individual patients may vary.

Abbreviations: DLCOsb, single breath diffusing capacity for carbon monoxide; TLC, total lung capacity.

[a] A rough rule of thumb, which may require adjustment based on the age of the patient.

Data from Global Initiative for Obstructive Lung Disease. Available at: www.goldcopd.org. Accessed April 2, 2010.