Chronic Respiratory Failure due to Bilateral Vocal Cord Paralysis Managed with Nocturnal Nasal Positive Pressure Ventilation*

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A patient with bilateral vocal cord paralysis developed chronic respiratory failure. Treatment with nocturnal inspiratory positive airway pressure via nasal mask improved symptoms and reduced hypercapnia.

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Bilateral vocal cord paralysis causes variable extrathoracic airway obstruction. Such obstruction can cause dyspnea and may lead to acute or chronic respiratory failure. Frequently these patients are managed with a tracheostomy. Nocturnal nasil ventilation has been used to treat chronic hypoventilation of various etiologies and thus offers hope of management without a tracheostomy. We report a patient with bilateral vocal cord paralysis causing chronic respiratory failure who was managed successfully with nocturnal positive pressure ventilation via a nasal mask.

CASE REPORT

A 40-year-old white man presented with a right upper lobe mass in 1975. Resection led to the diagnosis of Hodgkin's disease. During the surgery, the right phrenic nerve was damaged. He was treated with 5,400 rad of radiation therapy and nine cycles of chemotherapy. In November 1990, he complained of hoarseness. At that time, he had bilateral true vocal cord paresis and arytenoid joint stiffness. The vocal cord paresis may have been idiopathic due to previous surgery or possibly due to radiation damage. In February 1991, he complained of morning headaches, daytime drowsiness, and exertional dyspnea. He was referred to the pulmonary section for further evaluation. He denied smoking, alcohol, and drug use. He had no fever, chills, cough, or sputum production. His voice was hoarse and he was unable to speak fluently. He was 165 cm tall, weighed 55 kg and appeared lethargic. There were no palpable lymph nodes. Breath sounds were normal, and there was no wheezing or stridor. A radiograph of the chest showed an elevated right hemidiaphragm and atelectatic changes in the right upper lobe area. The lung fields were free of infiltrates. These findings were unchanged since 1986. Spirometry showed: FEV₁, 1.2 L (32 percent of predicted); forced vital capacity (FVC), 1.55 L (30 percent of predicted); FEV₁/FVC ratio, 84 percent; FEV₁/FEV₁total, 1.5. Lung volumes as a percentage of predicted showed vital capacity, 37 percent; total lung capacity, 61 percent; functional residual capacity, 89 percent; expiratory reserve volume, 37 percent; and residual volume, 134 percent. The specific airway conductance was 0.13 cm H₂O⁻¹·L⁻¹·s⁻¹ (normal: 0.2 to 0.35 cm H₂O⁻¹·L⁻¹·s⁻¹). The diffusing capacity was 43 percent of predicted. The diffusion per unit of alveolar volume was 6.39 (126 percent of predicted). The flow-volume curve showed reduced flow rates and a ratio of mid-expiratory flow to mid-inspiratory flow of 1.25 (normal, <1). Arterial blood gas analysis with the patient breathing room air showed a pH level of 7.35, a PaCO₂ value of 61 mm Hg, and a PaO₂ value of 52 mm Hg. The hemoglobin value was 16.4 g and the hematocrit value, 50.6 percent. The electrolyte values were normal except for a carbon dioxide level of 35 mmol/L. Thyroid function was normal.

Standard nocturnal polysomnography showed no discrete instances of apnea or hypopnea. On falling asleep the patient's arterial oxygen saturation (SaO₂) fell quickly to 82 to 84 percent. It remained at that level until the onset of rapid eye movement (REM) sleep (latency, 9 min) when the SaO₂ dropped to 56 percent. With supplemental oxygen at 1 L/min, the SaO₂ remained above 92 percent for the remainder of the night. Although the short REM latency suggested that REM deprivation was present, the percentage of REM sleep was reduced (3.3 percent). However, despite improved SaO₂, the patient awoke with a headache and was still sleepy during the day.

Bronchoscopy was performed to evaluate the upper and lower airways. Examination of the larynx showed the vocal cords to be midline with poor movement on phonation. The airway was slit-like with the cords bowed slightly. No endobronchial lesions were visualized. A sniff test, performed with fluoroscopy, showed a paralyzed right hemidiaphragm. This was confirmed by phrenic nerve conduction studies which showed no response on the right and a normal conduction time of 5.0 ms on the left. A maximal negative inspiratory force of 35 cm H₂O and a maximal expiratory force of 120 cm H₂O were recorded. An exercise test showed a maximal oxygen consumption of 0.92 L/min (40 percent of predicted).

It was concluded that the patient's chronic respiratory failure was (due to respiratory muscle weakness or fatigue or both) induced by upper airway obstruction combined with right phrenic nerve paralysis. Because of the patient's reluctance to have a tracheostomy, nocturnal nasal ventilation was initiated with a ventilator. Sleep monitoring was repeated and adjustments were made to augment the tidal volume by increasing the inspiratory positive airway pressure (IPAP), eventually to 12 cm H₂O. With O₂ at 1 L/min, the SaO₂ remained above 94 percent during both REM and non-REM sleep. The distribution of sleep was normalized and when the patient awoke he felt refreshed and had no headaches. Blood gas analysis showed a pH level of 7.33, a PaCO₂ of 57 mm Hg, and a PaO₂ of 77 mm Hg. Spirometry 12 months after initiation of nasal ventilation showed a FVC of 1.8 L, FEV₁ of 1.56 L, and a PaCO₂ of 47 mm Hg. The maximal negative inspiratory force had improved to 46 cm H₂O. The patient reported marked improvement in his morning headaches and daytime drowsiness. He also was able to return to his usual daytime work.

DISCUSSION

The diagnosis of upper airway obstruction usually is aided by history, pulmonary function tests and radiographic imaging. In this case, the clinical history and reduction in inspiratory flows suggested upper airway obstruction. Chronic respiratory failure due to upper airway obstruction may develop if the diagnosis is delayed. In our patient, at least three months elapsed before the diagnosis was established. The cause of the bilateral vocal cord paralysis in our patient is not clear. There was no evidence of radiation fibrosis in the lung or recurrent disease. Etiology is likely idiopathic, but also could be related to the prior surgery or the radiotherapy. The management of bilateral vocal cord paralysis consists of tracheostomy or surgical procedures to improve the mobility of the vocal cords. Because our patient refused a tracheostomy, we looked for alternative ways to manage his symptoms and chronic hypoventilation. Nasal positive pressure ventilation has been used success-
fully to manage chronic respiratory failure in patients with neuromuscular disease, chest wall abnormalities, COPD, and sleep apnea. Randolph et al.10,11 used nasal inspiratory positive pressure ventilation in a patient with bilateral vocal cord paresis to manage hypoventilation. Our patient had right phrenic nerve damage and vocal cord paralysis. This combination might have induced respiratory muscle fatigue and weakness and subsequent chronic hypoventilation. Although the administration of oxygen improved the patient's nocturnal saturation, the Pco2 might have increased as suggested by the patient's report of a severe morning headache. With ventilatory assistance, a large increase in both slow wave and REM sleep was noted as well as improved symptoms. The level of IPAP was empirically adjusted to 12 cm H2O to facilitate patient acceptance and minimize leaks. In other patients, monitoring of tidal volume and end tidal Pco2 during sleep may allow more precise titration of the optimal IPAP level. In our patient, IPAP plus O2 resulted in both improved ventilation and oxygenation during sleep. We conclude that in those patients who are not candidates for surgical intervention nocturnal nasal inspiratory positive pressure ventilation might be an effective method for reducing chronic hypercapnia due to bilateral vocal cord paralysis.

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Resolution of Severe Intrapulmonary Shunting After Liver Transplantation*
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A major complication of hepatic cirrhosis is arterial hypoxemia, often the result of intrapulmonary arteriovenous shunting. While previously such hypoxemia was thought to preclude successful hepatic transplantation, more recent studies have suggested that hepatic transplantation should be considered if the hypoxemia is corrected by supplemental oxygen. We report the findings in a cirrhotic patient with severe hypoxemia associated with intrapulmonary arteriovenous shunting. The patient did not respond to supplemental oxygen (Pa02 < 40 mm Hg on O2 at 4 L/min). The patient underwent successful hepatic transplantation, with complete resolution of intrapulmonary shunting. We believe that patients with cirrhosis-associated intrapulmonary shunting, even with hypoxemia resistant to supplemental oxygen, are acceptable candidates for hepatic transplantation.

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Many factors contribute to the development of arterial hypoxemia in hepatic cirrhosis, but the most significant is the presence of shunting in the intrapulmonary vasculature. Right-to-left shunts result from pulmonary vascular dilatations, either at the precapillary level or by arteriovenous communications.1-3 Cirrhotic patients who develop such shunts often have severe hypoxemia, with a PaO2 less than 50 mm Hg, and associated dyspnea, orthodeoxia, and platypnea. Severe cyanosis, digital clubbing, and exercise intolerance may occur.1 This spectrum of abnormalities is termed "the hepatopulmonary syndrome." Recently, Krowka and Cortese4 suggested that cirrhotic patients with hepatopulmonary syndrome who have a PaO2 of less than 70 mm Hg on room air could be considered for hepatic transplantation if they respond to supplemental O2 with a rise in PaO2. Patients who do not respond to supplemental O2 were considered poor candidates for transplantation.4

In the following case report, we document the clinical course of a cirrhotic patient with severe hypoxemia unresponsive to supplemental O2 (PaO2 < 40 mm Hg on O2 at 4 L/min), which was associated with intrapulmonary arteriovenous shunting. This patient made a full recovery after hepatic transplantation, documenting the potential for patients with hypoxemia unresponsive to O2 to survive transplantation.

CASE REPORT
The patient is an 18-year-old white man who underwent orthotopic liver transplantation at the University of Minnesota in 1989. Liver disease associated with α1-antitrypsin deficiency (PIZZ) was

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CHEST / 103 / 4 / APRIL, 1993 1271