# Biphasic Intermittent Positive Airway Pressure (BIPAP) Ventilation Support In The Postoperative Period For Patients With Myotonic Dystrophy

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#### Abstract

Patients with myotonic dystrophy (MD), a neuromuscular disorder, are significantly affected by general anesthesia. The most common post-operative complications seen in MD patients are respiratory complications; thus they are often in need of temporary post-operative ventilatory support. We present a case of a 65-year-old female who developed CO2 narcosis due to hypoventilation, after undergoing breast lumpectomy under general anesthesia. The ventilatory complication was treated with BIPAP using a full face mask. The patient recovered within 24 hours. This case proposes the option of the non-invasive method of BIPAP for MD patients with hypoventilation after general anesthesia.

## INTRODUCTION

Myotonic dystrophy (MD) is a multisystem disorder, manifested mainly by myotonia and accompanying muscle weakness. As the respiratory muscles are affected, ventilatory compromise is common, especially in the setting of surgery with general anesthesia. Aside from the preoperative investigation and intraoperative management, the correct plan for postoperative care is of prime importance for patients with MD. The most common and serious problem these patients encounter is postoperative hypoventilation resulting from the influence of hypnotic drugs, opioids and myorelaxants, thus exacerbating the underlying myotonic disease<sub>3</sub>. Occasionally, even after minor surgical intervention under light general anesthesia MD patients require a prolonged recovery period with mechanical ventilation<sub>4</sub>. While the issue of ventilation for MD patients post-operatively has been discussed in the literature, the case presented below is a first report describing the use of bi-level positive airway pressure (BIPAP) as temporary ventilatory support for a hypoventilating patient with myotonic dystrophy in the postoperative period.

## **CASE PRESENTATION**

A 65 year-old female patient was admitted for breast lumpectomy and axillary lymph node dissection due to breast carcinoma. She was known to suffer from MD, with clinical symptoms for the preceding 10 years. She presented with kyphoscoliosis, gastro-esophageal reflux and mild asthma as well. Regular medications included famotidine, aluminium hydroxide and tamoxifen. She had no known drug allergies. According to pre-operative anamnesis, she did not require assistance in daily activities of living, and preoperative examination did not show pathological findings aside from mild symmetric weakness in arms and legs. Her chest X-ray and blood tests were normal, and there were signs of left ventricular hypertrophy in her 12-lead electrocardiogram.

Premedication with diazepam 10 mg was administrated prior to surgery. General anesthesia was induced with Propofol 100 mg and Rocuronium 30 mg. Intubation was performed for ventilation and aspiration prevention. Morphine 10 mg was given intravenous for perioperative analgesia. Anesthesia was maintained with Isoflurane 0.2% and N2O 70%. The operation was completed after one hour and 20 minutes. Atropine 1 mg and Neostigmine 2.5 mg were given for reversal of muscle relaxation, and extubation was executed after verifying that the patient was able to breathe adequately without assistance. The patient was transferred to the PACU in full consciousness; she responded to her name and raised her limbs as commanded. The first measurement of patient's oxygen saturation in the PACU was 85% in room

air; after applying a non-rebreathing face mask, it increased to 93-95%. During the next 5 hours her pulse and blood pressure were stable, respiratory rate remained at 14-16/minute and only a worsening of her state of consciousness, responding to pain only, prevented her discharge to the surgical department. This neurological deterioration was unexplained by the drugs she received, and arterial blood gases were measured as a part of diagnostic work up. Severe acute respiratory acidosis with pH 7.1, PaCO2 of 88 mmHg and HCO<sub>3</sub> of 28 mEq was discovered. Naloxone 0.2 mg and Flumazenyl 0.2 mg were administered as an attempt to minimize any possible effect of the Morphine and Diazepam received earlier; the response to these antagonists was minimal. There were no pathological findings on a chest X-ray performed subsequently, and a CO2 narcosis due to hypoventilation was diagnosed.

BIPAP "Synchrony" (Respironics inc. USA) with full face mask was connected to the patient and ventilation with 18 respirations/minute; inspiratory pressure 16 cmH<sub>2</sub>O and expiratory pressure 5 cmH<sub>2</sub>O was initiated. The patient was detained in the PACU for close observation. During the next 24 hours, a gradual decrease in respiratory rate and ventilatory pressure was executed under monitoring of blood gases. Use of BIPAP was discontinued approximately 24 hours after initiation of BIPAP, with recovery of the patient to her preoperative state. The patient was transferred to the surgical department on the second post-operative day, after a 36 hour stay in the PACU, and was discharged from the hospital on the fifth post-operative day without sequel.

# DISCUSSION

First described by Steinert, in 1909, MD is the most common of the myotonic syndromes with a prevalence of  $0.003-0.005\%_2$ . It is an autosomal dominant disorder, and aside from a rare congenital form of the disease, its onset is most commonly between the second and fourth decades of life. Weakness is most common complaint in MD, characterized by myotonia (muscle stiffness). The disorder may be accompanied by symptoms including, marked wasting of the muscles of mastication, neck, pharynx and distal limbs; ptosis; frontal baldness; mental impairment and multisystem involvement. Extramuscular features include cataracts; cardiomyopathy; cardiac conduction abnormalities; restrictive lung disease; central and obstructive sleep apnea syndrome; dysphagia; delayed gastric emptying; and endocrine abnormalities such as hypothyroidism, primary hypogonadism, infertility and

diabetes mellitus<sub>2</sub>.

The most common perioperative complications in MD patients are pulmonary complications<sub>5</sub>. Acute ventilatory failure, leading to CO2 retention, is a significant type of respiratory complication in MD patients<sub>6</sub>. Most anesthetic drugs, including volatile anesthetics, hypnotics, opioids, and nondepolarizing and depolarizing muscle relaxants, can induce postoperative hypoventilation and respiratory insufficiency in MD patients. Therefore, the issue of ventilatory support is of prime importance in the post-operative care of these patients.

In cases of such a complication, mechanical ventilation with intubation has been used as ventilatory support. It must be acknowledged that endotracheal intubation is not without complications of its own. Mechanical ventilation with intubation, even for a short period of time, can increase the risk of nosocomial infection<sub>7</sub>. Moreover, intubated patients require sedation to prevent discomfort from the endotracheal tube. The use of sedative medications does not shorten time of recovery, and can yet increase it beyond the time required for recovery from the respiratory problem per se.

BIPAP is effective in improving oxygenation and decreasing hypercarbia<sub>9</sub>, and has been found to be beneficial to patients after lung resection surgery<sub>9</sub> and after coronary artery bypass graft (CABG) surgery<sub>10</sub>. Postoperative hypoventilation is a common occurrence in obese patients; the use of BIPAP has been recommended as part of postoperative care after gastric bypass surgery<sub>11</sub>.

Patients with MD may be a suitable group for similar management in the postoperative period after general anesthesia and deep sedation.

The use of (BIPAP) in the post-operative setting has been described in conjunction with use of a laryngeal airway mask (LMA)  $_{12}$ . The LMA itself is accompanied with potential complications, such as hypopharyngeal trauma and aspiration $_{13,14}$  Thus, in the case presented here, we emphasize that the patient required a full face-mask only, which is less uncomfortable and entails fewer potential complications than the LMA, and demonstrated a satisfactory ventilatory response to BIPAP.

## SUMMARY

MD patients are at risk for developing respiratory failure after general anesthesia. The use of BIPAP for MD patients

with ventilatory insufficiency in the postoperative period should be considered, as it can avoid hypoventilation, requires no invasive airway management, and may be an equally effective and safer alternative to invasive methods of ventilation.

# CORRESPONDENCE TO

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