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Successfully treated respiratory failure and pulmonary hypertension caused by Pickwickian syndrome with nasal bilevel positive airway pressure ventilation

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Abstract A 36-year-old woman (height 157cm, weight 114kg, body mass index 46.2kg/m²) with intolerable sleepiness and cyanosis was diagnosed with respiratory failure caused by Pickwickian syndrome. Echocardiography showed normal left ventricular systolic function and moderate pulmonary hypertension. Pulmonary artery systolic pressure was estimated at 50mmHg with doppler echocardiography. Oxygen inhalation and diuretics were started. Although the patient was treated with nasal continuous positive airway pressure ventilation, her hypoxemia and hypercapnia did not improve. Bilevel positive airway pressure ventilation with inspiratory pressure 10cmH₂O and expiratory pressure 2cmH₂O was quite effective in improving hypoxemia and hypercapnia, and pulmonary hypertension had disappeared at the discharge.

Introduction

Pickwickian syndrome was first described by Burwell, which was characterized as chronic hypercapnia and daytime hypersomnolence in extremely obese patient without underlying lung disease.¹⁾ Rapoport et al. demonstrated the effectiveness of nocturnal nasal-airway pressure ventilation for Pickwickian syndrome.²⁾ But when a patient has repeated apnea, nasal continuous positive airway pressure (n-CPAP) ventilation is not so effective. In this report, we present the effectiveness of nasal bilevel positive airway pressure ventilation (bilevel PAP) for repeated apnea caused by Pickwickian

syndrome.

Case Report

A 36-year-old woman was admitted to our ward because of cyanosis, anasarca and intolerable sleepiness. For two months before the admission, she had felt general malaise and her weight had gone from 110kg to 114 kg. She had not pointed out having any diseases. She had smoked 10 pieces of cigarette per day for 15 years. Relevant findings included the following: height 157cm; weight 114kg; body mass index (BMI) 46.2 kg/m²; blood pressure 114/58mmHg; pulse rate 86/min; respiratory rate 14/min. The respiration was irregular and repeated apnea

was observed.

On auscultation, the pulmonic closure sound was accentuated and no wheezing was heard. The neurological examination was normal. Arterial blood gas values with breathing room air were as follows: pH 7.428, PaCO₂ 73.4 mmHg, PaO₂ 38.5 mmHg, and HCO₃⁻ 47.6 mmol/L. A chest X-ray showed cardiac enlargement (cardio-thoracic ratio: 60%) and pulmonary congestion. Echocardiography showed as follows: the left ventricular end-diastolic dimension was 48mm and the left ventricular ejection fraction was 60%. The pressure difference between the right ventricle and the right atrium obtained by doppler echocardiography was approximately 50mmHg.

The patient was diagnosed as Pickwickian syndrome. We administered supplemental O₂ (0.5L/min) via a nasal cannula, but her PaCO₂ increased to 90mmHg. Apnea lasting more than 60 seconds was observed frequently. We began treatment with doxapram hydrochloride at a rate of 1mg/kg/min and furosemide 40mg/day. On the second day, we began n-CPAP ventilation with oxygen inhalation (5L/min). n-CPAP at a level of 10cmH₂O continued for 30 minutes, but was discontinued because of frequent apnea. Alternatively, bilevel PAP (Quantum PSV, Respironics Healthdyne technologies, Georgia, U.S.A.) with inspiratory pressure (IPAP) 6cmH₂O and with expiratory pressure (EPAP) 2cmH₂O with oxygen inhalation (5L/min) was started. Ventilation was set in the synchronous-timed mode with respiratory rates 15/min. IPAP had been increased gradually to 10cmH₂O as we had monitored PaCO₂ and her symptoms, such as dyspnea during inspiration and easiness in breathing.

On the tenth day, her somnolence and anasarca disappeared entirely. Polysomnography was performed on the first day (December 14th, 1997) and seven months after discharge (August 6th, 1998) (Fig 1). On the first day, frequent apnea with loud snoring was observed. The apnea-hypopnea index (AHI) was 39.8 (apnea index: 32.8, central apnea; 1.8, obstructive apnea; 29.4, mixed apnea; 1.6, hypopnea index; 7.0). SpO₂ decreased to lower than 80% frequent-

ly. Pulmonary function data before discharge were as follows: VC 3.06L, %VC 109%, FVC 3.05L, FEV_{1.0} 2.58L, and FEV_{1.0}% 84.3%.

She admitted on August 3rd again to take polysomnography. At the time of the admission, her weight was 86kg. The examination was taken on August 6th. She didn't use bilevel PAP at night during the admission, but less time was spent on snoring. SpO₂ sometimes decreased to lower than 85%. Her AHI was 8.4 (apnea index; 5.6, central apnea; 2.0, obstructive apnea; 2.9, mixed apnea; 0.8, hypopnea index; 2.8).

She has continued auxiliary oxygen inhalation at home during sleep at night. Hormonal examinations, a head computed tomography, a head magnetic resonance imaging, an electroencephalogram and an electromyogram all exhibited normal findings. At the discharge, we couldn't detect pulmonary hypertension with echocardiography, because tricuspid regurgitation disappeared.

Discussion

The patient in the present case had typical characteristics of Pickwickian syndrome. Pickwickian syndrome is well known to be a lethal condition.³⁾ Before Abrahamsen et al.⁴⁾ revealed the effectiveness of artificial ventilation for extreme obesity with respiratory failure, sudden respiratory arrest had been common in this syndrome.

To treat respiratory failure associated with Pickwickian syndrome, a tracheostomy used to be performed. But an alternative to tracheostomy, non-invasive positive pressure ventilation has been established as an effective method for treating Pickwickian syndrome.⁵⁾

Bilevel positive airway pressure ventilatory assistance is pressure-based equipment that is capable of setting inspiratory and expiratory positive pressure independently. Compared with n-CPAP, bilevel PAP is useful when a patient fails to respond to initial n-CPAP or a patient's breathing is too weak to breathe out enough CO₂.⁶⁾

The patient sometimes refused to attach the nasal mask at the beginning of n-CPAP. She said that using n-CPAP made it hard to

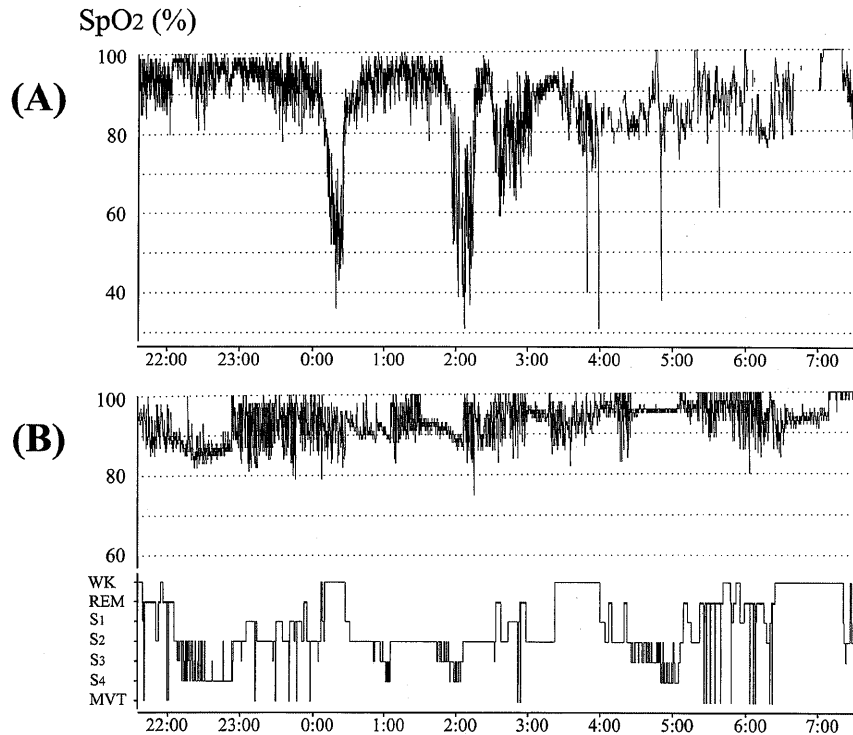


Fig. 1 Polysomnogram on the first day (A) and seven months after discharge (B). WK = awake, REM = rapid eye movement sleep, S1~ S4 refer to each sleep stages, MVT = Body movement.

(A) She was administered supplemental O₂ (5L / min) with a nasal cannula. We could not record an EEG, because she was not able to understand what she did at that time. (B) The examination was done seven months after discharge. She did not feel sleepiness during the daytime, but her SpO₂ at nighttime still decreased to lower than 85% several times.

breathe and, on the contrary, breathing was easier with bilevel PAP.

As the hypoxemia improved, the patient's urine output increased up to 3000ml/day in spite of administering the same dose of diuretics during using n-CPAP. In the present case, pulmonary hypertension improved with oxygen inhalation using bilevel PAP. The level of PaCO₂ was so high that we could not administer enough oxygen with a conventional way, i.e. via a nasal cannula. With bilevel PAP, we could administer oxygen aggressively. Although no studies have revealed the improvement in pulmonary hypertension with n-CPAP⁷⁾ or bilevel PAP⁸⁾, oxygenation relieved pulmonary hypoxic vasoconstriction and it improved the effect of diuretics. This is the reason why bilevel PAP was quite effective in improving pulmonary hypertension in the present case.

It is reported that approximately 500 thousands of Japanese suffer from sleep disorders such as sleep apnea syndrome.⁹⁾ These diseases have increased year by year, because the Japanese life style has gradually changed to the American or European one. We need to recognize that sleep disorders are more common than we have ever thought in Japan. Bilevel PAP should be considered as a first-line treatment for patients with respiratory failure caused by Pickwickian syndrome.

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