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Efficacy of Noninvasive Ventilation in a Patient with Hypercapnic Respiratory Failure Complicating Eisenmenger's Syndrome

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Abstract

Eisenmenger's syndrome is a severe type of congenital heart disease characterized by severe pulmonary arterial hypertension. In the cases that the pressure in pulmonary circulation exceeds the systemic pressure, there appears a right-to-left shunting of blood. Consequently, the syndrome exists hypoxemia and cyanosis. Hypercapnia is not common in these patients; however, it might coexist with hypoxemic failure if there are other restrictive pathologies associated. Meanwhile, it has been described high prevalence of sleep disorders in Down syndrome. There is no evidence about the role of noninvasive ventilation in the management of these patients. We present a 39-year-old man, suffering of Down and Eisenmenger's syndrome with multiple cardiac decompensations and obstructive sleep apnea (OSA), who was admitted to hospital due to severe somnolence, edema, and dyspnea. We observed a hypercapnic respiratory acidosis that ameliorated with noninvasive ventilation (NIV). The patient returned home with nocturnal NIV as a new treatment, and no further admission to hospital was seen in the following two years. To our knowledge, this is the first report about the utility of NIV in Eisenmenger's and Down syndrome patients.

Keywords: Congenital diseases, Eisenmenger's syndrome, hypercapnic respiratory, failure, hypoxemia, noninvasive ventilation

INTRODUCTION

Eisenmenger's syndrome (ES) is a severe type of congenital heart disease (CHD) characterized by severe pulmonary arterial hypertension (PAH). There is a connection between the systemic and pulmonary circulation; therefore, there is an existence of left-to-right shunt increasing pulmonary flow until it exists an irreversible pulmonary injury. Hypercapnia is not common in these patients; however, it might coexist with hypoxemic failure if there are other restrictive pathologies associated. Sleep disorders, mainly obstructive sleep apnea (OSA) due to anatomical alterations and hypoventilation, are very common in patients with Down syndrome (DS).

We present a case about an ES and DS patient with hypercapnic respiratory failure who was treated with noninvasive positive pressure ventilation (NIV). To our knowledge, this is the first report about the utility of NIV in ES patients.

CASE PRESENTATION

We have studied a 39-year-old man with personal history of DS, intraventricular communication, and PAH related to ES. Consequently, he had left chronic heart failure and a chronic hypercapnic respiratory failure treated with domiciliary oxygen, diuretics, bosentan, and sildenafil. He also had polyglobulia with hematocrit around 61% (which eventually needed phlebotomy to improve the level of hemoglobin) and symptomatic hyperuricemia treated with allopurinol. He had been admitted to hospital several times due to cardiac decompensation. He had a functional class IV NYHA score. The six-minute

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walking distance was 358 meters and his basal arterial oxygen saturation was about 80%.

The patient was admitted to hospital due to clinical worsening over a period of two months of worse central cyanosis and dyspnea, progressive somnolence, cough, and edemas in the legs and abdomen. At the emergency room, we noticed a saturation of 70% receiving oxygen at 2 L/min and the Glasgow scale was 13/15, blood pressure (BP) was 109/73 mmHg, respiratory rate (RR) was 30 bpm, and heart rate (HR) was 80 bpm. He was 150 cm high and weighed 65.5 kg (BMI 29). The patient showed excessive somnolence and important trembling. Cardiopulmonary auscultation showed crepitations in both sides. HR at 80 bpm with murmur.

On analyses, we noticed creatinine 1.28 mg/dL, BNP 996 pg/mL, troponin 0.2 ng/mL, uric acid 16 mg/dL, hematocrit 65%, hemoglobin 19.9 gr/dL, and platelets 77000, without other relevant alterations. An arterial gasometry showed pH 7.20, pCO_2 84 mmHg, and pO_2 65 mmHg. In the chest radiography (Figure 1) was observed, cardiomegaly, interstitial alveolar lesions that suggested pulmonary edema. Electrocardiogram was similar as previous. Echocardiography showed left ventricular ejection fraction (LVEF) of 80%, several pulmonary hypertension with a right-to-left shunt, interventricular communication and pulmonary arteries very dilated.

No clinical progress was made with only diuretics and oxygen, so NIV was initiated. A noninvasive ventilator was introduced (Vivo 40, Breas®, Mölnlycke Sweden) in ST mode setting at IPAP 19 cmH_20 and EPAP 8 cmH_20, I:E 1.2, slope of 2, and with 7 L/min of oxygen connected. In the following hours, the patient improved clinically, and an arterial gasometry after six hours showed pH 7.44, pCO_2 67 mmHg, pO_2 54 mmHg, HCO_3 45 mM/L, and SaO_2 88%. Two days later, analyses showed BNP 450 pg/mL, Hct 55%, and Hb 18.5 g/dL, and a satisfactory evolution with negative diuretic balance was appreciated.

The patient was sent home with nocturnal NIV (Vivo 40, Breas®, Mölnlycke Sweden) and same treatment as always. It was made a polygraphy study after three months of clinical stabilization, and it was observed and apnea-hipoapnea index of 18 per hour and as expected a severe nocturnal desaturation. Whereby we maintained the NIV due to the OSA, the thoracic anatomy, and hypoventilation. In the following two years, there was no admission to hospital, no cardiac decompensation, always awakeness, and no more somnolence was seen. Dyspnea index ameliorated from NYHA class IV to II, and clubbing fingers were smaller. The patient had saturation at home for 90%. On consultation, the six-minute walking test (6MWT) showed a walking distance of 380 meters, starting with oxygen saturation at 90%, HR 86 bpm, and BP 89/56 mmHg and ending with saturation of 79%, HR at 114 bpm, and BP 112/75 mmHg. Spirometry showed FEV_1 1540 mL/59%, FVC 1440 mL/44%, and FEV_1/FVC 86%, with a poor collaboration of the patient. The arterial gasometry at this moment showed pH 7.44, pCO_2 44 mmHg, pO_2 55 mmHg, HCO_3 44 mM/L, and SaO_2 88%.

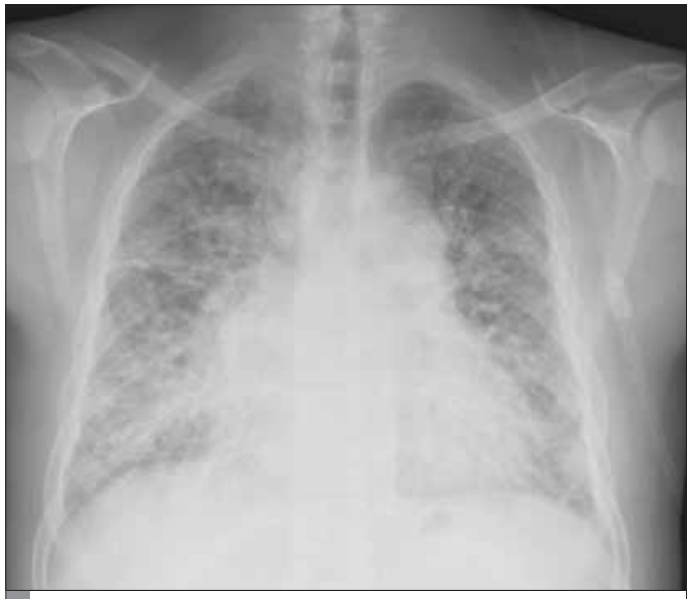

Figure 1. The chest X-ray of the patient showing a bilateral diffuse alveolar infiltrates

DISCUSSION

Eisenmenger's syndrome is a severe and irreversible CHD. Physiopathologically, it starts with a connection from systemic circulation of left ventricular to pulmonary circulation (left-to-right shunt). The pulmonary blood increases its flow, developing an irreversible injury with hypertrophic right ventricle and dilated pulmonary artery. Consequently, the blood flow becomes right-to-left, and hypoxia and erythrocytosis develop. Therefore, the severe form of PAH is the ES (1). Patients with ES have symptoms such as cyanosis, hypoxemia, dyspnea, syncope, paresthesia, and clubbing fingers. The following complications are observed: hemorrhage stroke, brain abscesses, erythrocytosis, gout, arrhythmias, heart failure, and sudden death. Dr. Victor Eisenmenger first described this condition in 1897. Among congenital cardiac disease patients with PAH, the estimated prevalence of ES is 4% (2).

OSA has more prevalence and is more severe in patients with DS than in the general population. Physiopathological explanations are mainly due to alterations on anatomy: Macroglossia, tonsil and palatal hypertrophy, narrowed throat, obesity, hypothyroidism, gastroesophageal reflux disease, and hypotonia in the mouth and upper airway (3). There are only three small studies that studied sleep-disordered breathing (SDB) in ES patients, and none of them showed differences between ES patients and the general population, the prevalence of SDB being the same (4). The prevalence of neonates with CHD in patients suffering from DS is 58%–100% (3, 5).

After exhaustive research, we have not found any literature that links together NIV and ES and DS. The role of NIV in the management of hypercapnic respiratory failure is well known (6). In the case of a cardiac right-to-left shunt, it seems like NIV would not be efficient due to the fact that an important amount of blood avoids the pulmonary alveolocapillary membrane. However, as we have seen in our patient, NIV can be useful, improving the clinical status and correcting the hypercapnia. We think hypercapnic respiratory failure in ES and DS patients could benefit from NIV, so this is an option to take into account in these situations.

Informed Consent: There was a verbal consent made between doctors and the patient, and the mother was the witness.

Peer-review: Externally peer-reviewed.

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