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A case of Eisenmenger's syndrome treated with extracorporeal lung and heart assist in the postpartum period

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Introduction

Extracorporeal Lung Assist (ECLA) and Extracorporeal Lung and Heart Assist (ECLHA) have been successfully applied to infant and adult patients with respiratory failure and/or heart failure [1,2]. However, only a few cases have been reported in Japan. We present a patient who was saved by ECLHA from moribund cardiopulmonary failure which developed in the postpartum period.

Case report

A 32-year-old woman developed dyspnea before the term of pregnancy. She underwent a Cesarean section at term. After the Cesarean section, dyspnea improved for a while under medical treatments. About 1 month after the delivery, however, she was admitted to a local hospital for severe dyspnea. The echocardiogram revealed a markedly dilated right ventricle and tricuspid valve insufficiency. An electrocardiogram demonstrated right ventricular hypertrophy, incomplete right bundle branch block, and ST segment depression in the V3 to V5 leads. Arterial blood gas analysis showed severe hypoxemia (Pao_2 < 40 torr). She was endotracheally intubated and mechanically ventilated, but her Spo_2 remained between 58% and 80%. Pao_2 further decreased to 29 torr on the 3rd day after admission. Her general condition gradually deteriorated and urine output was almost zero on the 4th day. She was transferred to the Intensive Care Unit of the Kumamoto University Hospital.

Though she and her family stated that she had no history of persistent cardiopulmonary disease, except for pneumonia 3 years before, the chest X-ray taken in the previous hospitals showed increased pulmonary vascular markings, enlargement of the bilateral pulmonary arteries and a dilatation of the right ventricle, indicating hidden cardiac abnormalities and prolonged pulmonary hypertension.

Her cardiopulmonary failure was refractory to all conventional treatments. Therefore, we decided to apply ECLHA as a life-support procedure and obtained informed consent from her family. The clinical course of this patient is summarized in Fig. 1.

The ECLHA circuit consisted of an artificial membrane lung with a surface area of 0.8 m^2 (MENOX EL 4000, Kuraray, Kurashiki, Japan), a pump-tube with a blood reservoir of 70 ml, a latex rubber tube for blood sampling, and interconnecting silicone tubes. The circuit was primed with 350 ml of lactated Ringer's solution containing 10 Unit/ml of heparin.

The left femoral vein and the right femoral artery were cannulated with a 21 Fr and a 17 Fr cannula (DLP, Grand Rapids, MI), respectively, by the Seldinger method under local anesthesia. We initiated extracorporeal bypass circulation at a flow rate of 3 l/min. The artificial lung was insufflated with 5 l/min of warmed oxygen. The mode of mechanical ventilation was changed from intermittent positive pressure ventilation (IPPV) to pressure support control. Pao_2 did not improve at the beginning even under an FIo_2 of 1.0, but it began to rise 14 h later.

Pulmonary arterial pressure transiently dropped after the initiation of ECLHA, but gradually rose again 12 h later. To decrease the pulmonary arterial pressure, we started a drip infusion of prostaglandin E_1 (PGE_1). When the dose of PGE_1 was increased to 21 ng/kg per min, a massive foamy pink secretion began to come out

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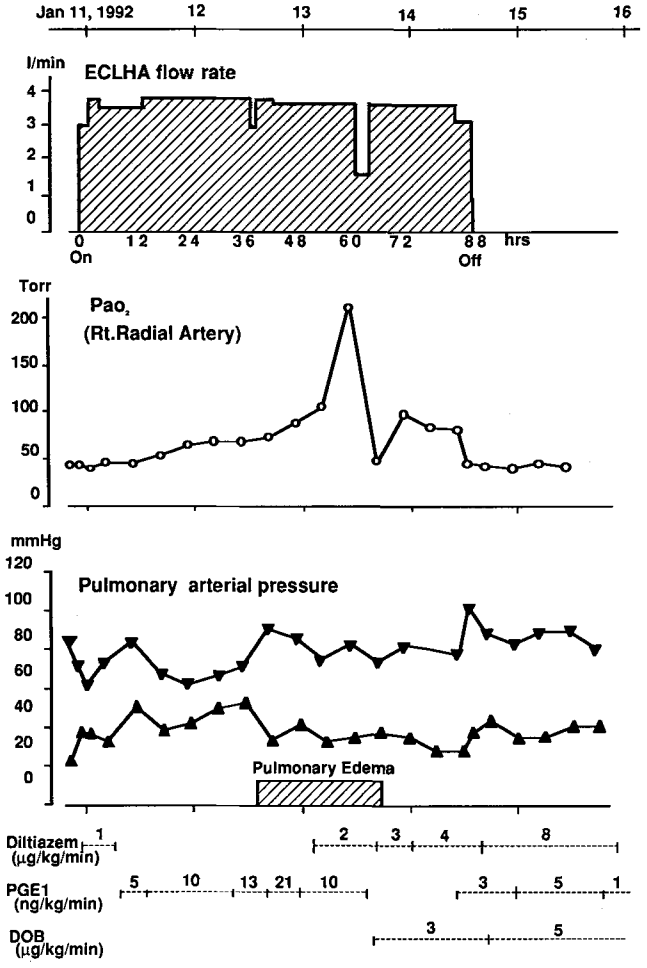

Fig. 1. Clinical course of extracorporeal lung and heart assist (ECLHA). PGE_1, prostaglandin E_1; DOB, dobutamine; Pao_2, arterial oxygen tension

through the endotracheal tube. A chest X-ray showed marked infiltrating shadows in the bilateral lung fields. We suspected that PGE_1 caused pulmonary edema, and discontinued it. The pulmonary arterial pressure increased again, but tracheal secretion gradually decreased. Pao_2 increased under ECLHA, despite pulmonary edema.

Echocardiography, performed 38 h after the initiation of ECLHA, revealed that the tip of the venous cannula had moved into the left atrium through a large atrial septal defect. We pulled back the venous cannula to the right atrium. The echocardiography also disclosed a right-to-left blood shunt through the septum. The patient was diagnosed as having Eisenmenger's syndrome.

Eighty hours after the initiation of ECLHA, her physical condition became stable, though the pulmonary arterial pressure remained high. Considering the advantages and disadvantages of the prolongation of ECLHA on this patient with Eisenmenger's syndrome, we terminated ECLHA after obtaining the consent of her family. The duration of ECLHA was 88 h 15 min.

Though her pulmonary arterial pressure further increased and Pao_2 decreased after weaning from ECLHA, her general condition remained relatively stable. There were no symptoms of cerebral damage or other undesirable side effects of ECLHA. The endotracheal tube was removed 20 days after weaning from ECLHA. The patient was discharged from the Intensive Care Unit to an ordinary ward.

Though she recovered sufficiently to walk around in the hospital, and enjoy the care of her baby, she died of sudden cardiac arrest 57 days after weaning from ECLHA. The cause of sudden cardiac arrest was not clarified because the family did not grant permission to perform an autopsy.

Discussion

The clinical application of ECLHA on patients with cardiopulmonary failure is gradually increasing in many countries, though its indications are still controversial [1,2]. ECLHA supports oxygen delivery and gives rest to the heart and lungs to provide time for healing. However, it is usually not indicated in patients with cardiopulmonary failure with irreversible pulmonary hypertension because ECLHA cannot directly treat any primary disease.

Patients with Eisenmenger's syndrome have sustained pulmonary hypertension. The cause of sustained pulmonary hypertension, however, is not always irreversible. An active pulmonary vasoconstriction contributes in part to the increase of the pulmonary vascular resistance [3–6]. In this patient, the enlarged uterus compressed the diaphragm, hindered ventilation, and might have triggered hypoxic pulmonary vasoconstriction. Increased pulmonary vascular resistance, along with increased blood volume during pregnancy, could have led to an increase in the right atrial pressure, resulting in the right-to-left shunt of the venous blood through the atrial septal defect and in severe hypoxemia. This vicious cycle might have led to the development of right heart failure.

In severe right heart failure with pulmonary hypertension, pulmonary vasodilators have been used [7,8]. Pulmonary vasodilators, however, aggravate pulmonary oxygenation in some patients by increasing intrapulmonary veno-arterial shunt. Veno-arterial ECLHA, in combination with pulmonary vasodilators, can reduce right heart work by eliminating pulmonary vasoconstriction while supporting oxygen delivery. In this patient, Pao_2 improved in spite of pulmonary edema during PGE_1 administration, probably as a result of a decrease in right-to-left blood shunt through the atrial

septal defect with the veno-arterial bypass. Development of pulmonary edema might indicate pulmonary vasodilation and an increase in pulmonary blood flow. Though we could not find the cause of her sudden death 2 months after ECLHA, her activities of daily living showed the improvement of the right heart function during life support by ECLHA. This case suggests that even a patient with acute exacerbation of Eisenmenger's syndrome can achieve remission to a certain degree with appropriate cardiopulmonary support.

We had 25 survivors out of 49 patients who underwent ECLHA in whom cardiopulmonary failure was so severe that they had little chance of survival with conventional therapy [1,2]. These cases also showed the favorable effects of life support by ECLHA on the recovery from acute exacerbation of cardiopulmonary diseases.

In summary, a patient with Eisenmenger's syndrome, which was aggravated during pregnancy and after the Cesarean section, was treated with ECLHA. Although ECLHA is not a radical treatment of Eisenmenger's syndrome, it was effective in relieving the acute exacerbation of the syndrome by reducing the right heart work, improving oxygenation, and removing pulmonary vasoconstriction.

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