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Effects of maternal hyperoxygenation in a case of severe congenital diaphragmatic hernia accompanied by hydrops fetalis☆

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ABSTRACT

We report a case of severe congenital diaphragmatic hernia accompanied by hydrops fetalis in a fetus conceived by a 33-year-old woman. Fetal magnetic resonance imaging performed at 27 weeks of gestation revealed herniation of the stomach, bowel, and left liver into the thoracic cavity, confirming severe left-sided CDH. Presence of hydrops fetalis at 32 weeks of gestation and its progression over the next 2 weeks was confirmed by fetal echography. Subsequently, fetal echocardiography revealed severe tricuspid regurgitation and reverse flow in the inferior vena cava at 33 weeks, with abnormal left and right ventricular Tei indices, extended left ventricular isovolumetric relaxation time, and increased preload index. Maternal hyperoxygenation (MHO) therapy was initiated at 35 weeks of gestation to increase pulmonary blood flow and promote venous return to the left ventricle. The hydrops was attenuated by MHO for 2 weeks, and fetal echocardiography demonstrated improved cardiac performance. A female infant was born at 37 weeks of gestation and underwent diaphragmatic repair shortly after birth. She survived surgery and was discharged at 4 months of age with no adverse sequelae. This case report suggests that MHO may be a potential therapy for severe congenital diaphragmatic hernia associated with hydrops fetalis.

Perinatal mortality associated with congenital diaphragmatic hernia (CDH) is high because of the presence of lung hypoplasia [1]. The association of hydrops fetalis further portends poor prognosis [2]. Cases of lethal nonimmune hydrops fetalis have been reported [3]. Here we report the successful treatment of severe CDH and hydrops fetalis by maternal hyperoxygenation (MHO) therapy administered before birth and surgical CDH repair performed after birth.

1. Case report

A 33-year-old woman was referred to our hospital with left-sided CDH in the fetus and polyhydramnios at 27 weeks of gestation. Her past medical history was unremarkable, and she was not treated for hydrops fetalis during her ongoing pregnancy. At 31 weeks of gestation, fetal echocardiography demonstrated that the fetus' lung to head ratio (LHR) was 1.57, cardio-thoracic area ratio (CTAR) was 20%, Tei index of the left ventricle (LV) was 0.70, and Tei index of the right ventricle (RV) was 0.59. Fetal magnetic resonance imaging revealed herniation of the stomach, bowel, and left liver into the thoracic cavity, confirming severe left-sided CDH.

At 32 weeks of gestation, fetal echography revealed mild pleural effusion and subcutaneous edema. By 34 weeks of gestation, pleural effusion was increased to 9 mm, with evidence of mild ascites, and subcutaneous edema of 10 mm (Fig. 1). In addition, LHR decreased to 0.8 and CTAR decreased to 16%. Fetal echocardiography detected severe tricuspid regurgitation. Reverse flow was identified in the inferior vena cava, and the preload index (PLI) and RV Tei index were 0.73 (normal 50th percentile, 0.244) [4] and 0.57, respectively. The LV isovolumetric relaxation time (IRT) was remarkably prolonged to 75 ms, while the LV Tei index worsened to 0.87 (Fig. 2).

In order to increase pulmonary blood flow and promote venous return to LV, maternal hyperoxygenation (MHO) therapy was initiated

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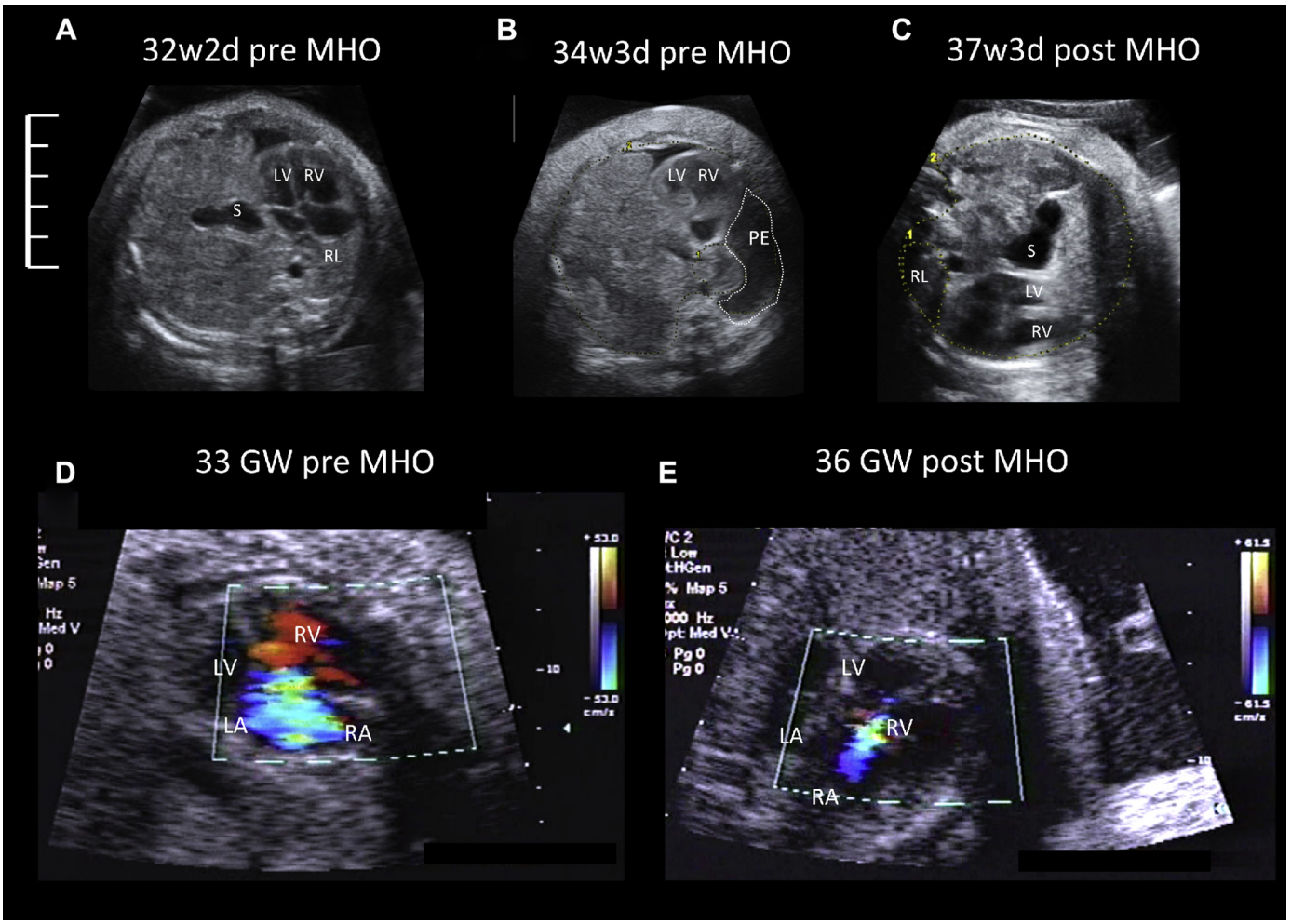

Fig. 1. Fetal echocardiography. RV, right ventricle; RA, right atrium; LV, left ventricle; LA, left atrium; RL, right lung; PE, pleural effusion; S, stomach. (A) Mild fetal pleural effusion, and hydrops at 32 weeks of gestation. (B) Subcutaneous edema of 10 mm and pleural effusion of 9 mm. Small amount of ascitic fluid is visible at 34 weeks of gestation. (C) Fetal pleural effusion and ascites are undetectable at 37 weeks of gestation. (D) Severe tricuspid regurgitation at 33 weeks of gestation. The color signal extends to the posterior wall of RA. (E) Decrease in tricuspid regurgitation at 36 weeks of gestation. The width of the color signal has decreased and does not extend to the posterior wall of RA.

at 35 weeks of gestation. The mother received 60% humidified oxygen via a facemask for 3 h, 4 times a day [5]. After MHO for 2 weeks, the resistance index (RI), which was measured by the pulse Doppler method from the right pulmonary blood flow, decreased from 0.63 to 0.51. Tricuspid regurgitation markedly decreased, and the PLI improved to 0.51 at 37 weeks of gestation (normal 50th percentile, 0.223). In addition, the IRT improved from 75 ms to 50 ms, and the LV and RV Tei indices markedly improved from 0.87 to 0.26 and from 0.57 to 0.26, respectively (Fig. 2). Further, the fetal pleural effusion and ascites were also no longer detectable, with attenuation of the hydrops (Fig. 1).

A female infant was delivered by planned cesarean section after 2 weeks of MHO at 37 weeks of gestation. Echocardiography of the newborn revealed an LV diastolic diameter of 15 mm, a right pulmonary arterial diameter of 3.5 mm, and a left pulmonary arterial diameter of 2.2 mm. The infant did not exhibit hydrops fetalis, and her preductal oxygen saturation after birth was 60%. Immediately after birth, she was intubated and provided ventilator support with high frequency oscillatory ventilation and inhalation of nitric oxide (20 ppm). Catecholamine, prostaglandin E_1 (3 ng/kg/min) and prostaglandin I_2 (2 ng/kg/min) were administered. Her systolic blood pressure was maintained at 60 mm Hg, and her systemic circulation was stable. Prostaglandin I_2 administration was increased gradually every 30 min until preductal oxygen saturation reached 90%. Preductal oxygen saturation was increased to 97% by 90 min after birth.

She underwent definitive patch repair of CDH at 3 h after birth. There was a 6 cm defect in the left diaphragm. The lateral segmental branch of the left hepatic lobe, intestine, colon, spleen and stomach were herniated into the thorax, and the hernia sac could not be identified. She was weaned off prostaglandin E_1 and prostaglandin I_2 because of left to right shunt dominant through the ductus arteriosus on the third postoperative day; further, she was weaned off nitric oxide inhalation considering the improvement of pulmonary hypertension on the 14th postoperative day. In addition, she was weaned off catecholamines because she exhibited stable hemodynamics by the 20th postoperative day. Despite her hemodynamic stability, radiography and computed tomography revealed massive bilateral pleural effusion, and she began exhibiting signs of respiratory distress on the 29th postoperative day. Her respiratory distress was effectively relieved by thoracic cavity drainage. She was extubated 43 days after surgery, and discharged at 4 months of age without any further requirement of oxygen therapy or medications. No pulmonary hypertension was observed and she did not exhibit any neurological sequelae. No adverse effects of MHO were observed during or after her treatment.