

Eisenmenger Syndrome in Pregnancy

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Eisenmenger's syndrome consists of pulmonary hypertension with a reversed or bidirectional shunt at the atrioventricular, or aortopulmonary level. Eisenmenger's syndrome in pregnancy is usually associated with high mortality rates (nearly 30-50%). Unfortunately, pulmonary hypertension is aggravated during pregnancy and often leads to an unfavorable outcome. Here, we report a successful pregnancy in a woman with Eisenmenger syndrome.

Introduction

Eisenmenger's syndrome consists of pulmonary hypertension with a reversed or bidirectional shunt at the atrioventricular, or aortopulmonary level. Eisenmenger's syndrome in pregnancy is usually associated with high mortality rates (30-50%). Unfortunately, pulmonary hypertension is aggravated during pregnancy and often leads to an unfavorable outcome. Here, we report a successful pregnancy in a woman with Eisenmenger syndrome.

Case Report

A 26 year-old woman with Eisenmenger syndrome and moderate persistent asthma presented at 8 weeks amenorrhea with sporadic history of dyspnea and cyanosis of the extremities. She was a gravida 3 with past history of one stillborn due to extreme prematurity and one pregnancy with intrauterine growth restriction. Home oxygen therapy associated with formoterol and budesonide were in use.

On physical examination, her heart rate was 80 bpm and blood pressure was 110x70 mm Hg. There was peripheral cyanosis, polycythemia (hematocrit of 71 %), digital clubbing, hypophonesis of the second heart sound in pulmonary focus and systolic blow +/-6 in the left sternal border. The two-dimensional echocardiography (2D echo) and Doppler report showed a 15 mm interventricular septal defect with bidirectional flow and pulmonary hypertension (pulmonary artery pressure of 115 mmHg) and slight increase of the right chambers. Medical approach included limited physical activity,

continuous oxygen therapy, and aspirin (100mg/d). Due to polycythemia, two phlebotomies were performed, the first at 8 weeks and the last at the 25th week. Fetal growth was evaluated by means of serial ultrasound. The pregnancy progressed uneventfully until the 26th week, when the patient complained about worsened dyspnea associated with edema of the lower limbs and weight gain; a diuretic was then introduced. After one week the patient was hospitalized to be followed until delivery. Subcutaneous heparin was introduced at the 29th week. At the 32nd week, the patient had a bronchial infection and was treated with cephalothin, which improved her condition. At the 33rd week, corticotherapy was administered for pulmonary maturity. An amniocentesis revealed a mature fetus and the presence of meconium. A cesarean section was performed with tubal ligation, and a girl was born weighing 2.250g, with Apgar of 7/9/10.

Six days after the cesarean, the patient had sudden dyspnea with a non-confirmed hypothesis of pulmonary embolism. Nevertheless, anticoagulation procedures were started. The patient's condition improved and she was discharged 17 days after the cesarean. She returned 40 days after the surgery for reevaluation, with exclusive breastfeeding, when she reported dyspnea at moderate efforts; only oxygen therapy was in use. Currently she has been followed up by cardiologists while waiting for heart-lung transplantation.

Discussion

Several of the hemodynamic changes that occur during a normal pregnancy contribute to the high maternal mortality in patients with Eisenmenger syndrome. The progressive increase in plasma volume, which peaks at about 50% above baseline early in the third trimester, adds to the burden of a compromised right ventricle and may precipitate right heart failure. The preexisting pulmonary vascular disease restricts this increased flow of blood to the lungs and increases right ventricular work. Systemic vasodilatation is a physiological adaptation of normal pregnancy and it is associated with an increase in cardiac output and renal blood flow. As peripheral vascular resistance falls, the patient with Eisenmenger syndrome may augment the right-to-left shunting that exacerbates the preexisting hypoxia, which, in turn, may cause more pulmonary vasoconstriction. At the time of labor and delivery, severe hemodynamic compromise may occur. Acidosis and hypercarbia may further increase pulmonary vascular resistance. Any hypovolemia resulting from blood loss or hypotension from a vasovagal response to pain may result in sudden death. In addition, death may also occur from pulmonary thromboembolism or in situ pulmonary infarction¹.

The degree of maternal hypoxemia is the most important

Key words

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predictor of fetal outcome; prepregnant levels of arterial oxygen saturation of 85% or less are associated with rates of live births as low as 12%, while saturation of 90% or more results in 92% of live births². This is explained by a high incidence of spontaneous abortions, a 30-50% risk of premature delivery and low birth weights as maternal hypoxemia disturbs fetal growth¹⁻³.

Maternal mortality in the presence of Eisenmenger's syndrome is reported as 30 to 50%^{1,4}. Gleicher et al¹ reported a 34% mortality associated with vaginal delivery and a 75% mortality associated with cesarean section.

Because of the high mortality associated with continuing pregnancy, abortion is the treatment of choice for women with Eisenmenger's syndrome. For the patient with a continuing gestation, hospitalization in the second trimester is highly recommended⁵.

Continuous administration of oxygen, anticoagulation and pulmonary vasodilator is controversial. Although there aren't any controlled trials, a Brazilian series of 13 pregnancies⁶ reported improved maternal mortality (23%) with a regimen of oxygen, heparin before delivery, and warfarin after 48 hours. Sixty per cent of infants were live births, mostly premature.

Anesthesia for patients with pulmonary hypertension is controversial. Theoretically, conduction anesthesia, with its accompanying risk of hypotension, should be avoided. The use of epidural or intrathecal morphine sulphate, a technique devoid of effect on systemic blood pressure, may be the best approach to anesthetic management of these

difficult patients.

There is no evidence to support the choice of either vaginal or cesarean delivery for cardiac reasons: vaginal delivery is associated with a lower average blood loss but also increased maternal effort⁷. After delivery, anticoagulation can be restarted once there is no evidence of bleeding and then coumadin can be given. In-hospital monitoring should be continued for at least 2 weeks after delivery.

The mortality of patients with Eisenmenger's syndrome who become pregnant remains prohibitively high. Appropriate advice regarding contraception should be given to all patients. If a patient becomes pregnant, therapeutic termination should be offered. If pregnancy continues against medical advice, treatment strategies as outlined above may be helpful, with prolonged hospital care both pre and post partum.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Study Association

This study is not associated with any graduation program.

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