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The achievement of pregnancy with eisenmenger syndrome

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Objective

Eisenmenger syndrome(ES) is a rare congenital heart defect in pregnant women and is seen in around 3% of pregnancies. Pregnancies with pulmonary hypertension (PHT) due to ES are associated with significant maternal and fetal mortality. ES is associated with eventful pregnancies due to circulatory disorders caused by the disease and the increased cardiac afterload due to pregnancy itself. Effective contraception methods are applied in those cases and if the patient is pregnant the option of termination is recommended before 10 weeks. A multidisciplinary consultation is required if the family wishes to continue pregnancy. Maternal mortality in pregnant women with ES is reported to be 30-50% and perinatal risks have been increased considerably. We present a rare case of a pregnancy complicated by Eisenmenger syndrome due to severe PHT that successfully led to a term delivery.

Methods

Patient 27 years old, G5P1, at 35 + 1 weeks of gestational age was referred to our department due to maternal ES. The mother had been operated for atrial and ventricular septal defect at the age of 8 years. The patient medical history is also remarkable for chronic hypertension and hypothyroidism. During pregnancy, she was on enoxaparin, levothyroxine and multivitamins. First and second trimester scans were normal, the risk for chromosomal abnormalities was low and the glucose tolerance test was normal. She was referred to us after being diagnosed with severe PHT. The obstetric measurements according to the ultrasound were consistent with 35w and no fetal anomaly was detected. Fetal echocardiography was normal. In the pelvic examınatıon, there was no cervical dilatation and no effacement. After a multidisciplinary meeting and consultation it was considered appropriate to admit and follow up the patient in the ICU. The patient was treated with 12 mg intramuscular betamethasone, a cesarean section under epidural anesthesia was performed and a female baby with APGAR score 8/9 and 2200 grams was delivered. On postoperative day 2, treatment with bosentan, sildenafilsitrat and oxygen was initiated. She was followed up for 15 days in the ICU and she was discharged with an oxygen device for home use. The patient was followed up one month after the delivery with her general condition and her vital signs being stable and within the normal ranges. The patient was informed about the available methods of contraception and was referred to the cardiology outpatient clinic for further assessment and management.

Results

Pregnancy is associated with high maternal mortality and morbidity in patients with Eisenmenger syndrome. There are literature sources indicating that the risk of perinatal maternal mortality increases to 50%. Due to the decreased systemic vascular resistance during pregnancy, the right to left shunt is getting increased and the pulmonary perfusion decreases, fact that causes deterioration of hypoxia and even sudden death. ES cases diagnosed at advanced gestational age should be evaluated by a multidisciplinary team. Intrauterine growth restriction is expected in 30% of the fetuses. Epidural anesthesia should be preferred in cesarean delivery because it reduces the possibility of rapid hemodynamic changes. An early onset of induction and an atraumatic birth under the epidural block is preferred generally. The high incidence of maternal death is related to hypovolemia, preeclampsia and thromboembolic events. For the above reasons, contraception should be explained to patients with ES and tubal sterilization should be recommended. If the pregnancy is less than 10 weeks, the curettage is recommended at unwanted pregnancies, whereas, if more than 10 weeks, a termination option should be provided.

Conclusion

Medical termination of pregnancy is a treatment method for women with pulmonary hypertension due to high mortality. Information about the prognosis should be given to patients who wish to continue the pregnancy; the patient should be hospitalized and followed up along with the cardiology and pulmonary department. The birth should be considered when fetal lung maturation is achieved.