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Evaluation of Outcomes of Pregnancy in Women with Eisenmenger Syndrome: Is There Any Prognostic Criterion?

Eisenmenger Sendromu Olan Gebe Kadın Hastalarda Gebelik Sonuçlarının Değerlendirilmesi: Prognostik Bir Kriter Var Mı?

ABSTRACT

Objective: Pregnancy in women with Eisenmenger syndrome (ES) presents very high morbidity and mortality rates for both the mother and fetus. In this study, we aimed to evaluate the outcomes of pregnancies affected by ES.

Methods: This retrospective cohort study reviewed pregnancies in women with ES at two university hospitals over the past 10 years. The primary outcomes examined were maternal, perinatal, and pregnancy-related outcomes in women with ES.

Results: The study enrolled eight pregnant women diagnosed with ES. The average maternal age was 26.7 years (range 21-36 years). The causes of ES included ventricular septal defect (five patients), patent ductus arteriosus (two patients), and atrial septal defect (one patient). Four patients had been diagnosed with ES before pregnancy. Two patients died in this group; one developed right heart failure in the third trimester and unfortunately died immediately after delivery due to cardiogenic shock, and another died on the 10th postpartum day from sudden cardiac arrest despite having had a successful delivery. Four patients were diagnosed with ES during the postpartum period; none in this group died, and there were no cases of fetal mortality. We could not identify any clinical or echocardiographic predictors for postpartum mortality. The two deceased patients showed tendencies of severe right heart failure and lower oxygen saturation during the third trimester and before delivery.

Conclusion: Despite advanced obstetric care and targeted treatments for pulmonary arterial hypertension, maternal mortality is still unacceptably high in patients with ES. No significant predictors of maternal mortality were identified.

Keywords: Congenital heart disease, Eisenmenger syndrome, pregnancy, pulmonary arterial hypertension

ÖZET

Amaç: Eisenmenger sendromu (ES) olan hastalarda gebelik, hem anne hem de bebek için yüksek mortalite ve morbidite riski taşır. Bu çalışmada, ES olan gebe hastalarda gebelik sonuçlarını değerlendirmeyi amaçladık.

Yöntem: Bu retrospektif kohort çalışmaya, iki üniversite hastanesinde son 10 yılda görülen ES'li gebe hastalar alınmıştır. Çalışmanın primer sonlanımı, ES olan gebe hastalarda maternal, perinatal ve gebelik sonuçlarının değerlendirilmesidir.

Bulgular: Bu çalışmaya, 8 ES'li gebe kadın hasta dahil edildi. Ortalama anne yaşı 26,7 (21-36 yıl) idi. Eisenmenger sendromu nedenleri, 5 hastada ventriküler septal defekt, 2 hastada patent duktus arteriyozus ve 1 hastada atriyal septal defekt olarak bulundu. Eisenmenger sendromu tanısı dört hastaya gebelik öncesinde konulmuştu. Bu grupta iki hasta hayatını kaybetti. Bu hastalardan birinde üçüncü trimesterde sağ kalp yetersizliği gelişti, ne yazık ki kardiyojenik şok nedeniyle doğumdan hemen sonra öldü. Diğer hasta sorunsuz bir doğum eylemi geçirmesine rağmen doğum sonrası 10. gününde ani kardiyak arrest nedeniyle öldü. Dört hasta doğum sonrası dönemde ilk kez ES tanısı aldı. Bu grupta hayatını kaybeden hasta olmadı. Fetal mortalite tespit edilmedi. Doğum sonrası mortalite için, herhangi bir klinik ve ekokardiyografik öngördürücü belirleyemedik. Ölümle sonuçlanan iki hastada, üçüncü trimesterde ve doğum öncesi ciddi sağ kalp yetersizliği ve daha düşük oksijen saturasyonu mevcuttu.

Sonuç: Eisenmenger sendromu olan gebe hastalarda, gelişmiş obstetrik bakım ve pulmoner arteriyel hipertansiyona yönelik spesifik tedaviye rağmen, maternal mortalite hala kabul edilemez derecede yüksektir. Çalışmamızda maternal mortalite için belirgin bir öngördürücü tespit edemedik.

Anahtar Kelimeler: Konjenital kalp hastalığı, Eisenmenger sendromu, gebelik, pulmoner arteriyel hipertansiyon

ORIGINAL ARTICLE
KLİNİK ÇALIŞMA

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Congenital heart disease is the most common cardiovascular disease detected during pregnancy after preeclampsia in developed countries. Pulmonary arterial hypertension (PAH) is present in 5% of patients with congenital heart disease.[1] Pregnancy in women with PAH is associated with very high morbidity and mortality rates for both the mother and fetus.[2] According to the modified World Health Organization (WHO) classification of maternal cardiovascular risk, PAH is associated with very high maternal mortality and morbidity risks.[3]

Eisenmenger syndrome (ES) represents the most severe phenotype of PAH associated with congenital heart defects, which can occur in patients with large, unrepaired intra- or extracardiac shunts.[3,4] Maternal mortality and morbidity in patients with ES range between 26% and 66%, primarily caused by right heart failure, cardiogenic shock, thrombotic complications, and sudden cardiac arrest.[5-7] Additionally, fetal outcomes are often unfavorable, with increased risks of miscarriage, intrauterine growth retardation, preterm birth, low birth weight, and congenital heart defects.[7] Due to the high maternal and fetal mortality and morbidity, the 2022 European Society of Cardiology (ESC) guidelines regard pregnancy in patients with PAH as a contraindication.[8]

Even though physicians do not recommend pregnancy, we more commonly encounter patients with ES in the clinic either by chance or intentionally. Some patients may also be diagnosed with ES for the first time during pregnancy or in the postpartum period. In this retrospective study, we aimed to evaluate the maternal and perinatal characteristics and pregnancy outcomes in women with ES.

Materials and Methods

This retrospective cohort study investigated pregnancies in women with ES at Mersin University and Gazi University Hospitals in Türkiye over the past 10 years. The study population included pregnant women with ES who were managed in these two hospitals over the past 10 years. The inclusion criteria consisted of all pregnant or postpartum women diagnosed with ES. There were no exclusion criteria. Ethical approval for this study was received from the Non-Interventional Research Ethics Committee of Gazi University (Approval Number: 132, Date: 17.02.2021). Informed consent was obtained from all accessible patients. The study was conducted in accordance with the Declaration of Helsinki.

The primary outcomes of this study were maternal, perinatal, and pregnancy outcomes in women with ES. We evaluated patients' characteristics, such as age, WHO functional class, N-terminal pro brain natriuretic peptide levels, oxygen saturation, and PAH treatment pre-pregnancy, at the first trimester, third trimester of pregnancy, and again in the postpartum period. Time and mode of delivery, and anesthetic management were reviewed. Pregnancy outcomes including maternal mortality, obstetric complications, heart failure, mode of delivery, and baby birth weight were evaluated. Medication during pregnancy and the peripartum period were described. Lengths of hospital stay and time in the intensive care unit were also recorded.

Transthoracic echocardiography (TTE) reports of all patients were reviewed using an Echo Vivid E9 XDClear machine and EchoPAC system (General Electric Healthcare, US). The probability of pulmonary hypertension related to congenital heart defects was estimated as systolic pulmonary arterial pressure (sPAP) by adding the tricuspid regurgitation maximal pressure gradient and estimated right atrial pressure.[9] Tricuspid annular plane systolic excursion (TAPSE) and the presence of pericardial effusion were also documented. The TAPSE/sPAP ratio, included in the latest ESC guideline as a diagnostic and prognostic criterion, were also calculated.[8] Baseline TTE reports before pregnancy were evaluated as basal TTE. Eisenmenger syndrome was first diagnosed during the postpartum period in four patients. In these cases, echocardiography was performed during the initial evaluation and repeated 14 days after delivery, with the findings documented.

Right heart catheterization (RHC) reports of patients diagnosed with ES before pregnancy were reviewed. For those diagnosed with ES in the postpartum period, RHC was performed 14 days after delivery. Pulmonary artery pressure (systolic, diastolic, and mean) and pulmonary vascular resistance (PVR) values were recorded from these reports. Artificial intelligence-assisted technologies were not used in the production of the submitted work.

Statistical Analysis

All analyses were conducted using Statistica version 13. Continuous data were presented as means ± standard deviations. Categorical data were presented as frequencies and percentages, with comparisons made using chi-square tests. *P*-values of less than 0.05 were considered statistically significant.

Results

This study reviewed and enrolled eight women. The mean maternal age was 26.7 years (range 21-36 years). Two women had second pregnancies, and their outcomes during these subsequent pregnancies were evaluated. The underlying causes of ES were as follows: five patients with ventricular septal defect, two patients with patent ductus arteriosus, and one patient with atrial septal defect.

The diagnosis of ES and PAH had been established before pregnancy in four patients (Table 1, cases 1, 4, 5, 7). These patients were treated with PAH-specific drugs before pregnancy: one patient with monotherapy (bosentan) and three patients with combination therapy (bosentan and tadalafil/sildenafil). These patients were diagnosed as pregnant during the first trimester. Termination of pregnancy was advised for all patients. However, all the women chose to continue their

ABBREVIATIONS

C/S	Cesarean section
ES	Eisenmenger syndrome
ESC	European Society of Cardiology
IV	Intravenous
PAH	Pulmonary arterial hypertension
PVR	Pulmonary vascular resistance
RHC	Right heart catheterization
sPAP	Systolic pulmonary arterial pressure
TAPSE	Tricuspid annular plane systolic excursion
TTE	Transthoracic echocardiography
WHO	World Health Organization

Table 1. Clinical Characteristics of Patients During Pregnancy

Case	Cause of ES	Defect Size (cm)	Age	Diagnosis of ES	FC Before Pregnancy/1st Trimester/3rd Trimester/ Postpartum	SpO_2 (%) Before Pregnancy/ Delivery/ Postpartum	NT-proBNP (pg/mL) Basal/3rd Trimester	PAH Treatment Before Pregnancy	PAH Treatment During Pregnancy
#1	VSD	3	21	Pre-pregnancy	2/2/3/Ex	82/75/40	-	Bosentan	Sildenafil
#2	VSD	3.3	25	Postpartum	2/2/3/4	-/70/85	-	-	-
#3	ASD	4.2	36	Postpartum	2/2/3/4	-/75/85	-	-	-
#4	VSD	1.7	29	Pre-pregnancy	2/2/3/2	85/80/85	199/863	Bosentan, Tadalafil	Tadalafil, Inhaled Iloprost, IV Epoprostenol
#5	VSD	1.4	27	Pre-pregnancy	2/2/2/3	77/70/70	349/550	Bosentan, Tadalafil	Tadalafil, Inhaled Iloprost
#6	PDA	1.1	31	Postpartum	2/2/3/3		-	-	-
#7	VSD	1.8	22	Pre-pregnancy	2/2/3/3	89/-/69	90/350	Bosentan, Sildenafil	Sildenafil, Inhaled Iloprost
#8	PDA	0.9	23	Postpartum	2/2/3/3	-/75/85	-	-	-

ASD, Atrial Septal Defect; ES, Eisenmenger Syndrome; FC, Functional Capacity; PAH, Pulmonary Arterial Hypertension; PDA, Patent Ductus Arteriosus; VSD, Ventricular Septal Defect.

pregnancies. At their initial visits, all patients were New York Heart Association (NYHA) functional class II. During pregnancy, two patients received inhaled iloprost and tadalafil, one patient received sildenafil, and one patient received inhaled iloprost and sildenafil. Case 4 experienced an increase in dyspnea during the last trimester, so intravenous (IV) epoprostenol treatment was initiated, and the patient underwent delivery under IV epoprostenol treatment. After delivery, the patient refused the use of parenteral prostaglandin, so the treatment was gradually stopped during the postpartum period. All these patients also received anticoagulation therapy during pregnancy with enoxaparin 1 mg/kg subcutaneously once daily or twice daily according to the patient's risk of bleeding.

There were two maternal deaths (case 1 and case 5) in these patients. Case 1 developed right heart failure during the third trimester, was hospitalized, and received positive inotropic therapy, IV diuretics, oxygen, and parenteral iloprost therapy. This patient was the first in our cohort, whom we followed 10 years ago. At that time, the only parenteral prostaglandin available in the hospital was iloprost. Case 1 underwent a cesarean section (C/S) under general anesthesia; unfortunately, after the end of the delivery, the patient could not recover from anesthesia and presented with cardiogenic shock and cardiac arrest. The other patient who died, Case 5, had oxygen saturation remaining below 80% throughout the pregnancy. Case 5 underwent C/S under epidural anesthesia and, although she experienced a successful delivery and had a healthy baby, she experienced sudden cardiac arrest on the postpartum tenth day.

Four patients were initially diagnosed with ES during the postpartum period (Table 1, cases 2, 3, 6, 8). All these patients were referred to our clinic during the postpartum period due to respiratory distress and hypoxia and were diagnosed with ES after delivery. Therefore, they could not take any PAH specific treatment before and during pregnancy. They were taken into the intensive care unit, and positive inotropic therapy, anticoagulation, diuretic, and parenteral iloprost therapy were started. No patients died in this group. However, after delivery, case 6 began experiencing frequent hospitalizations due to the worsening of PAH and started subcutaneous treprostinil therapy post-delivery. Unfortunately, she died in the fourth year after delivery.

Clinical characteristics of all patients during pregnancy are summarized in Table 1. Case 3 and 8 delivered via normal vaginal delivery. Four patients underwent C/S under general anesthesia, while the others had epidural or spinal anesthesia. The patients stayed in the hospital for an average of 11 days and received treatment in the intensive care unit for an average of 4 days. After delivery, PAH therapy continued as combination therapy with bosentan and tadalafil/sildenafil. Prophylactic low-dose anticoagulation therapy continued for 14 days during the postpartum period with enoxaparin 1 mg/kg subcutaneously. No significant bleeding complications were observed in patients using anticoagulants during pregnancy and the postpartum period.

As for pregnancy complications, one patient developed preeclampsia. Intrauterine growth retardation and low birth weight were the most common neonatal complications. The mean gestational period was about 35 weeks. There was no fetal mortality. The mean birth weight was 2087 grams (minimum 1900 - maximum 2300 grams). Clinical characteristics of patients during delivery and the postpartum period are summarized in Table 2.

Table 2. Clinical Characteristics of Patients During Delivery and Postpartum Period

Case	Week of Labor	Delivery Method	Anesthesia	Fetal Weight (g)	Cardiac Events During Pregnancy	Cardiac Events During Postpartum	Postpartum Hospitalization (Days)
#1	32	C/S	GA	1900	RHF in 3rd Trimester	Exitus after delivery	1 day
#2	32	C/S	GA	1950	RHF in 3rd Trimester	Hypoxia	17 days (3 Days ICU)
#3	35	VD	Epidural	-	Low O_2 saturation	-	10 days (2 Days ICU)
#4	34	C/S	GA	2300	Increased dyspnea	-	10 days (3 days ICU)
#5	34	C/S	Epidural	-	Increased dyspnea, low O_2 saturation	Exitus on 10th day postpartum	10 days ICU
#6	38	C/S	Spinal	-	Increased dyspnea	RHF	6 days (2 Days ICU)
#7	34	C/S	GA	-	Increased dyspnea, low O_2 saturation	RHF	20 days (5 days ICU)
#8	38	VD	None	2200	Preeclampsia	RHF, Hypoxia	15 days (3 days ICU)

C/S, Cesarean Section; GA, General Anesthesia; ICU, Intensive Care Unit; RHF, Right Heart Failure; VD, Vaginal Delivery.

Table 3. Echocardiographic Findings of All Patients

Case	Pre-pregnancy sPAB (mmHg)	Pre-pregnancy TAPSE (mm)	TAPSE/sPAP Ratio (mm/mmHg)	Post-pregnancy sPAB (mmHg)	Post-pregnancy TAPSE (mm)	TAPSE/sPAP Ratio (mm/mmHg)
#1	85	15	0.18	-	-	-
#2	-	-	-	80	16	0.20
#3	-	-	-	110	21	0.19
#4	100	24	0.24	110	19	0.17
#5	120	19	0.16			-
#6	-	-	-			
#7	95	22	0.23			
#8	-	-	-	140	23	0.16

sPAB, Systolic Pulmonary Arterial Pressure (via Echocardiography); TAPSE, Tricuspid Annular Plane Systolic Excursion.

Table 4. Right Heart Catheterization Findings of All Patients

Case	Pre-pregnancy PAP (mmHg)	Pre-pregnancy PVR (WU)	Post-pregnancy PAP (mmHg)	Post-pregnancy PVR (WU)
#1	110/50/75	16	-	-
#2	-	-	125/30/60	15
#3	-	-	75/25/46	10
#4	115/56/80	20	-	-
#5	124/85/105	30	-	-
#6	-	-	95/34/40	8
#7	110/58/81	12	-	-
#8			140/43/80	13

PAP, Pulmonary Artery Pressure (Systolic/Diastolic/Mean); PVR, Pulmonary Vascular Resistance.

A notable characteristic of these patients was the presence of severe anemia during pregnancy or the postpartum period, despite having ES. The mean hemoglobin and hematocrit levels of the patients were 11.8 g/dL (range 9.2-15 g/dL) and 39% (range 26-53%), respectively. Upon checking the ferritin levels of the patients, it was found that they were significantly low, and their anemia was primarily due to iron deficiency, besides physiological anemia (mean ferritin level 12 ng/mL, range 4-23 ng/mL).

Four patients with a pre-pregnancy ES diagnosis had pre-pregnancy RHC data available. In these patients, an average PVR of 20 WU (minimum 12 - maximum 30 WU) was determined. Postpartum RHC was applied to four patients who were diagnosed with ES during the postpartum period. The average PVR values of these patients were lower, at 11.5 WU (minimum 8 - maximum 15 WU).

Echocardiographic data and RHC findings are presented in Tables 3 and 4. The TAPSE/sPAP ratio in the pre-pregnancy baseline TTE

of patients with mortality was in the high-risk zone, below 0.19. Only Case 6 exhibited mild pericardial effusion. There were no pre-pregnancy clinical or echocardiographic indicators predicting clinical deterioration and death in patients with mortality. Observing general trends, two patients with mortality showed a tendency toward high PVR levels during RHC, severe right heart failure, and low oxygen saturation, low TAPSE/sPAP ratio in the third trimester and before delivery. Given the small number of patients, reaching a statistical conclusion was not possible.

Discussion

Eisenmenger syndrome is an important subset of PAH that manifests with uncorrected large inoperable shunt lesions and severe cyanosis.[4] The European Registry of Pregnancy and Cardiac Disease (ROPAC) study was conducted as a prospective study screening pregnant women with heart disease between 2007 and 2018.[2] In this study, 5,739 pregnant women with heart disease were enrolled. According to this study, the most common heart disease was congenital heart disease (57%), and the highest mortality rate among pregnant women was detected in patients with PAH (9%). Women with ES faced the most adverse outcomes, with a 9.7% mortality rate and 48.4% experiencing heart failure in the ROPAC study.[2]

Recent advancements in PAH treatment have also led to progress in the treatment of patients with ES. Despite these developments, according to the latest ESC guidelines, pregnancy is still contraindicated in patients with ES.[8] In some case series involving pregnant patients with ES, low mortality and morbidity outcomes have been reported.[10-14] However, despite these successful outcomes reported in these case reports, there are still case series with high mortality rates.[7,15-18]

Duan et al.[7] conducted a retrospective analysis of 11 pregnancies in women with ES and recorded four maternal deaths, resulting in a mortality rate of 36%. They found that arterial oxygen saturation and pre-delivery arterial oxygen tension during oxygen inhalation were significantly lower, and pre-delivery heart failure was higher in the maternal death group. Similarly, in our study, patients who died tended to have low oxygen saturation levels throughout their pregnancies and exhibited signs of right-sided heart failure before delivery.

In a study by Dachlan et al.,[16] out of 18 pregnant patients with ES, seven patients (38.8%) died. All deaths occurred within two weeks after delivery and were due to cardiogenic shock. Tokgöz et al.[17] conducted a single-center study in 35 pregnant patients with PAH, including eight patients with ES. Maternal mortality was also high in this study, with 13 PAH patients (37%, two of whom had ES) dying.

In our study, the incidence of maternal mortality was 25%. Consistent with the literature, maternal deaths occurred after delivery or within the postpartum two weeks. Half of the patients were diagnosed with ES during the postpartum period, and all these patients were from rural areas with low socioeconomic levels, including two refugees. Additionally, one patient deteriorated rapidly after childbirth and died within four years.

We could not identify any clinical or echocardiographic predictor for postpartum mortality. Observing general trends, two patients with mortality showed tendencies of having higher PVR and signs of right-sided heart failure, along with lower oxygen saturation during the third trimester and before delivery. The TAPSE/sPAP ratio is an indicator of right ventricle-pulmonary artery coupling and has been shown to be a prognostic criterion in PAH patients.[8,19] These patients also had low baseline TAPSE/sPAP ratios. However, there is no powerful clinical or echocardiographic criterion predicting clinical deterioration and death, making it very difficult to decide the prognosis before pregnancy. There was no mortality in patients diagnosed with ES during the postpartum period. Pulmonary vascular resistance levels in patients diagnosed with ES after pregnancy were lower than in those diagnosed before pregnancy. Although the sample size is too small to draw definitive conclusions, the postpartum diagnosis in these patients might be attributed to their relatively good clinical condition before pregnancy, which could result in lower symptom severity and consequently, a lower pregnancy risk.

Successful pregnancy outcomes published in the literature may give the impression of a good prognosis. However, even if patients experience a successful pregnancy, as seen in our cases, they may still face mortality in the postpartum period, and PAH can progress, necessitating parenteral PAH treatment.

Eisenmenger syndrome during pregnancy increases the risk of fetal complications such as miscarriage, small for gestational age, prematurity, intrauterine growth retardation, and neonatal death. In a case series of 11 pregnancies with ES, Duan et al.[7] reported exceptionally high fetal complication rates: preterm delivery (88%), small for gestational age (83%), fetal mortality (27%), and neonatal mortality (25%). In our study, intrauterine growth retardation and low birth weight were the most common neonatal complications. Fortunately, all infants, including those of the deceased patients, survived.

Most of the patients in the study had iron deficiency anemia besides the physiological anemia of pregnancy. The lack of polycythemia in pregnant patients with congenital heart disease might result in a missed diagnosis of ES. Moreover, iron deficiency anemia should be treated, as it further diminishes the patients' exercise capacity and adversely impacts their prognosis.[8]

Pregnancy in patients with ES is a dangerous duo because cardiovascular changes that occur during pregnancy are poorly tolerated and contribute to high maternal morbidity. The mechanical compression caused by the growing uterus during pregnancy, increased sex hormones, increased intravascular volume, and cardiac output both during pregnancy and in the peripartum period exacerbate physiological stress.[20] During pregnancy, because of the physiological decrease in protein S and increase in prothrombotic factors, the risk of thrombosis increases, and pregnancy-induced hypercoagulability increases the risk of pulmonary thromboembolism. Additionally, the decrease in systemic vascular resistance leads to an increase in right-to-left shunting, ultimately resulting in pulmonary vasoconstriction and right heart failure. The highest risk occurs in the early postpartum period.[16,21] Therefore, patients should be counseled against pregnancy. If pregnancy occurs, termination is ideally recommended before 10 weeks of gestation. If the patient decides to continue the pregnancy, a close and multidisciplinary follow-up should be planned.

Patients should be hospitalized earlier during the third trimester of pregnancy. Maternal arterial oxygen saturation must be kept above 70 mmHg. Administering oxygen via a mask will improve the patient's oxygen saturation and reduce pulmonary artery pressure.[12] Diuretic therapy may be added to manage congestive symptoms and right heart failure. In patients with ES, a prothrombotic state may cause embolic complications, but risks of hemoptysis and postpartum hemorrhage are also high in these patients. Therefore, anticoagulation must be used with caution. Since endothelin-receptor antagonists (bosentan, ambrisentan, macitentan) and guanylate cyclase stimulators (riociguat) are teratogenic, they must be discontinued during pregnancy.[1,6,7] Phosphodiesterase inhibitors (sildenafil, tadalafil) and prostacyclin analogs (epoprostenol, treprostinil, iloprost) are recommended during pregnancy.[22,23] Vaso-reactive patients can continue to use calcium channels blockers.[8]

The ideal mode of delivery is controversial. Compared with C/S delivery, vaginal delivery is associated with less risk of hemorrhage, infection, and venous thromboembolism. However, vaginal delivery is also associated with increased basal cardiac output with every uterine contraction, which may worsen right heart failure. Cesarean section may be safer than vaginal delivery for patients with severe hemodynamic abnormalities due to the greater stability of hemodynamics. To prevent further pregnancies, tubal sterilization may be performed during the C/S operation if the patient wishes. Early delivery as planned C/S between 32 and 36 weeks of pregnancy is usually proposed.[20] According to the 2022 ESC guidelines for the management of cardiovascular diseases during pregnancy, C/S is advised in severe forms of PAH.[1]

Anesthetic management of patients with ES is controversial. General anesthesia may have a negative effect. Epidural anesthesia may prevent increases in PVR by preserving spontaneous ventilation to minimize mean airway pressures. Epidural anesthesia or a combined spinal-epidural approach is generally well-tolerated hemodynamically.[13] After delivery, all PAH-specific medications can be started without concern for teratogenicity. The first postpartum week is considered a period of maximum mortality because of the redistribution of fluid in postpartum periods and volume overload causing pulmonary edema or cardiogenic shock.[24] Thus, intensive postoperative monitoring is necessary as most hemodynamic changes of pregnancy usually resolve by two weeks postpartum.

Study Limitations

The number of subjects included in the study is quite limited. Additionally, since the study was retrospective, some data were missing.

Conclusion

Eisenmenger syndrome and pregnancy is an uncommon association characterized by a high risk of maternal and fetal mortality and morbidity. Despite advanced obstetric care and PAH-specific treatment, maternal mortality is still unacceptably high and unpredictable. The ESC recommends counseling against pregnancy and termination in the case of pregnancy. Unfortunately, most patients refuse termination. If the patient wishes to continue the pregnancy, close monitoring by a multidisciplinary team, including obstetricians, cardiologists, and anesthesiologists, is necessary. For a successful outcome, close monitoring, pulmonary vasodilator therapy, early hospitalization, longer postpartum follow-up, and individualization of treatment are very important.

Ethics Committee Approval: Ethical approval for this study was received from the Non-Interventional Research Ethics Committee of Gazi University (Approval Number: 132, Date: 17.02.2021).

Informed Consent: Informed consent was obtained from all accessible patients.

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