ORIGINAL ARTICLE

Pregnancy Outcome in Eisenmenger Syndrome at an Indian Tertiary Center

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Abstract

Aim: To evaluate the materno-fetal outcome of pregnancy with Eisenmenger syndrome (ES).

Materials and methods: This retrospective observational study was carried out at a tertiary teaching institute over a 1-year period. The occurrence of ES in pregnant women with the cardiac disease along with their materno-fetal outcomes was analyzed.

Results: Among 27,259 antenatal women who delivered over a period of 1 year, 104 had cardiac disease with 31 congenital heart disease (CHD). Only three cases of pregnancies with ES were found among 104. Thus, ES was seen in 1:9086 pregnant women, 1:35 pregnant women with cardiac disorders, and 1:10 pregnant women with CHD. The mean age was 25.3 ± 0.58 years while the mean gestational age (GA) at delivery was 35.23 ± 0.67 weeks. The mean pulmonary arterial pressure was 68.7 ± 13.3 mm Hg. All had preterm deliveries. Vaginal delivery was conducted in 66.7% while 33.3% had a cesarean delivery. No maternal mortality was seen. Fetal demise was seen in one of three cases.

Conclusion: Multidisciplinary care with the availability of a critical care unit can help achieve a positive materno-fetal outcome in ES during pregnancy which till now has been known to have a very high mortality rate.

Clinical significance: Of all CHD in pregnancy, ES is associated with a very high maternal mortality rate. However, with multidisciplinary effort and approach, optimum outcomes are achievable.

Keywords: Cyanotic heart disease in pregnancy, Pulmonary hypertension in pregnancy.

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Introduction

Eisenmenger syndrome (ES) refers to pulmonary hypertension due to a reversed (right to left) or bidirectional shunt between pulmonary and systemic circulation.[1,2]

Eisenmenger syndrome (ES) has an incidence of 3% in pregnant females with CHD.[3] The mortality rates of ES in pregnancy (ESP) are 30–50% and even up to 65% in those undergoing cesarean section (CS).[4] Pulmonary vascular remodeling seen in these patients limits the ability to cope with the pregnancy-induced physiological changes in the cardiovascular system (CVS) including a decrease in systemic vascular resistance (SVR) and an increase in cardiac output (CO) and blood volume, subsequently leading to right heart dysfunction (RHD). This RHD leads to a rise in pulmonary vascular resistance (PVR), decreased CO resulting in hypotension, and finally death.

Owing to high maternal mortality rates in this condition, women are advised against conception or to opt for medical termination of pregnancy (MTP). However, if GA for MTP has surpassed, or they begin or continue a pregnancy against advice, well-coordinated multidisciplinary care becomes mandatory.

Most literature on ESP outcomes is from developed countries. There is only limited data on the effect of this condition on pregnancy in the Indian population. This paper describes the clinical course and materno-fetal outcome in ESP along with a literature review of its management.

Materials and Methods

This retrospective observational study was done at a tertiary teaching institute over a 1-year duration. Retrospective data collection of pregnancies with cardiac disease, delivered over 1 year, was done from the medical records. The number of antenatal cases (ANC) with the cardiac disease was assessed. Of these, ANC with ESP was evaluated. The occurrence of ES in the pregnant population and in the pregnant population with the cardiac disease was thus calculated. The presentation, symptomatology, management, mode of delivery, and materno-fetal outcome were studied in detail in all these women.

Results

The total number of ANC who delivered at our institute over 1 year was 27,259. Of these, 104 ANC had cardiac disease. Of these 104 ANC, 31 had CHD, of which, three had pulmonary arterial hypertension (PAH) due to ES.

Eisenmenger syndrome (ES) was seen in 1:9086 pregnant women, 1:35 pregnant women with cardiac diseases, and 1:10 pregnant women with CHD. The mean age and BMI of these women were 25.3 ± 0.58 years and 20.5 ± 1.1 kg/m^2, respectively.

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Table 1: Table showing materno-fetal parameters evaluated in the antenatal women with ES

Sl. no.	*Parameters evaluated*	
1	Occurrence of ES among antenatal women	1:9086
2	Occurrence of ES among antenatal women with cardiac disease	1:35
3	Occurrence of ES among antenatal women with congenital cardiac disease	1:10
4	Mean age (years)	25.3 ± 0.58
5	Mean BMI (kg/m^2)	20.5 ± 1.1
6	Mean gestational age at presentation (weeks)	26.2 ± 14.08
7	Mean gestational age at delivery (weeks)	35.23 ± 0.67
8	Mean pulmonary arterial pressure (mm Hg)	68.7 ± 13.3
9	Maternal mortality (%) (0/3)	0
10	Preterm delivery (%) (3/3)	100
11	Vaginal delivery (%) (2/3)	66.7
12	Cesarean delivery (%) (1/3)	33.3
13	Fetal growth retardation (%) (3/3)	100
14	Fetal survival (%) (2/3)	66.7

BMI, body mass index; ES, Eisenmenger syndrome

One woman had an atrial septal defect (ASD) while two others had both ventricular septal defect (VSD) with ASD. The mean pulmonary arterial pressure was 68.7 ± 13.3 mm Hg. The mean GA at delivery was 35.23 ± 0.67 weeks.

While 66.7% (2/3) had vaginal delivery, 33.3% (1/3) underwent CS for fetal indication [severe fetal growth restriction (FGR) with absent end-diastolic flow (AEDF)]. All three (100%) had preterm delivery (PTD) and FGR. Fetal demise was seen in 33.3% (1/3). No maternal mortality was observed (Table 1).

Summary of Cases (Table 2)

Case 1

A 26-years primigravida at 34 + 3 weeks presented with acute breathlessness and generalized edema. Eisenmenger syndrome (ES) was diagnosed after a cyanotic episode and dyspnea, 5 years back. While on irregular torsemide, metoprolol, digoxin, and sildenafil, she conceived. She came as an unbooked and uninvestigated case. On admission, vitals were: PR was 120/min, BP – 130/88 mm Hg, RR – 28/minute, SpO_2 – 88% on oxygen by face mask, and bilateral chest crepitation along with peripheral cyanosis and clubbing (Fig. 1). Abdominal examination revealed 28 weeks uterus with a single live fetus with vertex presentation. The patient was stabilized, cardiology consultation was sought, and started on cardiac drugs (metoprolol, digoxin, and sildenafil). Chest X-ray showed cardiomegaly with bilateral chest infiltrates. A two-dimensional (2D) Echo showed VSD with ASD with PAH (84 mm Hg) with ES. Dexamethasone was given for fetal lung maturity. Hb and hematocrit were 13 gm% and 62%, respectively. She was kept on oxygen by face mask with regular oxygen saturation (SpO_2) monitoring. She underwent spontaneous preterm labor (PTL) with forceps delivery of a 1.2 kg baby with an Apgar score of 5,7 at 0 and 5 minutes. The baby was shifted to NICU. She was given intrapartum endocarditis (IE) prophylaxis. The patient had atonic postpartum hemorrhage (PPH) which was managed by oxytocin, crystalloid, and blood transfusion with an aim to maintain SpO_2 without letting hypotension ensue. Two units of packed-cell transfused, and sildenafil and metoprolol were continued in the

Table 2: Table showing details of individual cases with Eisenmenger syndrome

Sl. No.	*Age*	*Obstetric profile*	*Booked status*	*Gestational age at presentation (weeks)*	*Symptoms and signs at presentation*	*Abdominal examination*	*Treatment*	*Outcome*		*Underlying cardiac defect*
								Mode of delivery	*Maternal*	*Fetal*	
1	26	Primigravida	U[a]	34 + 3	Acute dyspnea, cyanosis, clubbing	28 weeks	• Pulmonary vasodilator[c] • Postpartum anticoagulation • Endocarditis prophylaxis[d] • Steroid for fetal lung maturity	Preterm (34 + 5) delivery	Discharged on 10th postnatal day	1.2 kg baby (shifted to NICU)	ASD + VSD[e]
2	27	Primigravida	U[a]	34 + 3	Dyspnea, generalized edema	30 weeks	• Pulmonary vasodilator[c] • Postpartum anticoagulation • Endocarditis prophylaxis[d]	Preterm (35 IUD) delivery	Discharged on 10th postnatal day	2 kg anencephalic stillborn fetus	ASD+VSD[e]
3	25	G2P1L1 with previous normal delivery	B[b]	10 weeks (booking) Readmission at 34 weeks	Exertional dyspnea	30 weeks	• Pulmonary vasodilator[c] • Postpartum anticoagulation • Endocarditis prophylaxis[d] • Steroid for fetal lung maturity	Elective CS at 36 weeks (severe FGR with AEDF)	Discharged on 10th postnatal day	1.9 kg, male baby (shifted to NICU)	VSD[e]

[a]U, unbooked; [b]B, booked; [c]Oxygen by face mask, sildenafil (5-phosphodiesterase inhibitor); [d]Ampicillin 2 gm + Gentamycin 80 mg intravenous; [e]ASD, atrial septal defect; VSD, ventricular septal defect

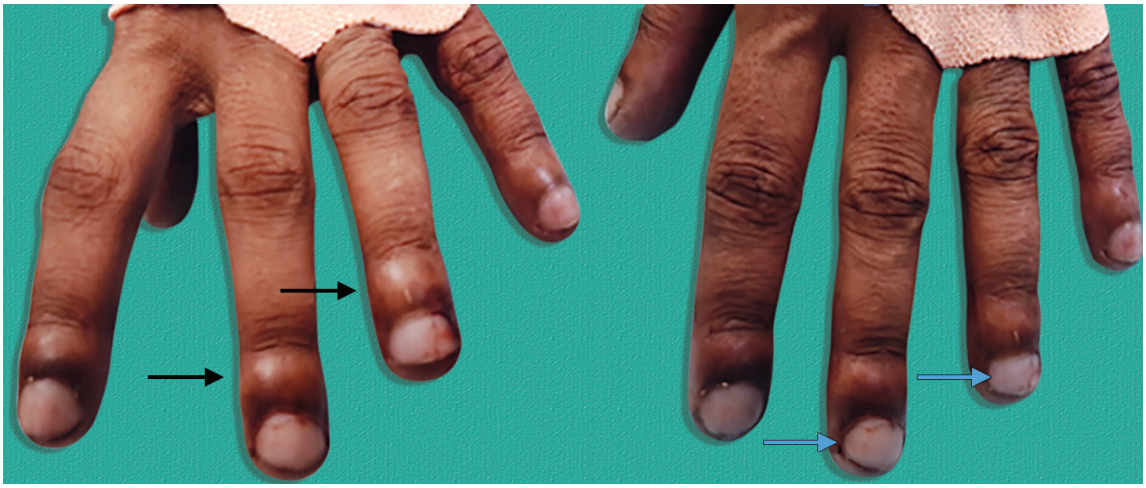

Fig. 1: Figure showing clubbing (black arrow) and peripheral cyanosis (blue arrow) in an antenatal patient with Eisenmenger syndrome (case 1)

postpartum period by the cardiologist. After controlling PPH, she was given postpartum thromboprophylaxis after 24 hours. The patient was discharged in satisfactory condition on the 10th postoperative day (POD) along with the baby.

Case 2

A 27-years primigravida, unbooked, uninvestigated female, came to Obstetrics Emergency at 34 + 3 weeks with a complaint of dyspnea at rest and generalized edema. Her vitals were PR – 110/min, BP 100/60 mm Hg, RR – 32/min, and SpO_2 87% on oxygen by face mask. A pansystolic murmur (PSM) along with bilateral crepitations was auscultated. On abdominal examination, fundal height was 30 weeks along with fetal bradycardia. The patient was admitted to the critical care obstetrics unit. The poor materno-fetal prognosis was explained. She was nursed in a propped-up position with oxygen support and a cardiologist's opinion was taken. Her Hb and hematocrit were 12 gm% and 62%, respectively. A 2D Echo showed ASD+VSD+ES+PAH (60 mm Hg). She was started on sildenafil (25 mg TDS) with an injection of furosemide 40 mg once daily after cardiology consultation. She went in spontaneous PTL and delivered a 2 kg anencephalic stillborn fetus (35 weeks). The patient received postpartum thromboprophylaxis and IE prophylaxis. She received oxygen support and sildenafil during the postpartum period and was discharged on the 10th postnatal day (PND).

Case 3

A 25-year, G2P1L1 presented to ANC-OPD at 10 weeks with a history of exertional dyspnea and occasional hemoptysis over the last 1 year. Her last pregnancy was uneventful with a normal delivery at home 2-years back. On examination, PR was 106/min, BP 100/70 mm Hg, and no pallor/cyanosis/edema. On CVS examination, PSM in the left parasternal area was heard. Respiratory system and per-abdominal examination were normal, and per-vaginal examination revealed a soft, anteverted uterus of 10 weeks size. Ultrasound report confirmed a single intra-uterine 10 weeks fetus. A 2D Echo was suggestive of CHD with large peri-membranous non-restrictive VSD with balanced shunt with significant PAH. In view of the risks of ESP, she was offered MTP but she refused and continued pregnancy. She was treated in consultation with the cardiologist who advised conservative management (sildenafil) and further evaluation after delivery. Serial hemoglobin and hematocrit monitoring were done. Hb and hematocrit were 12 gm% and 35%, respectively. Rest antenatal investigations were normal. Iron supplementation was given. She followed in ANC and cardiology OPD. Level-2 ultrasound and fetal echo were normal. Her ultrasound at 28 weeks showed single 24-weeks live fetus of 990 gm, anterior-placenta, adequate liquor, and color Doppler indices of umbilical and MCA suggested signs of early placental insufficiency. She refused admission. She was admitted at 34 weeks in view of at-rest dyspnea and deranged Doppler. Repeated Echo showed PAH (60 mm Hg). She was nursed in a propped-up position with oxygen support, and started on sildenafil. Intrapartum endocarditis (IE) prophylaxis was administered, and elective CS was done at 36 weeks for FGR with AEDF under epidural anesthesia and delivered a 1.9 kg male baby with an Apgar score of 7,9 at 0 and 5 minutes. The patient was kept in the obstetric critical care unit during the postoperative period and was given postpartum thromboprophylaxis. She was discharged on 10th POD along with the baby with the advice of cardiology follow-up.

All these patients were seen after 6 and 12 weeks postpartum and their condition was satisfactory.

Discussion

Eisenmenger syndrome in pregnancy needs well-coordinated multidisciplinary care from a Cardiologist, high-risk Obstetrician, Critical-Obstetrics-Care-unit, and Anesthetist.

Development of ES occurs due to progressive pulmonary hypertension leading to a reversal of shunt or bidirectional shunting in presence of congenital left to right shunt. Pulmonary arterial hypertension, a least-tolerated condition in the antenatal period is defined as an increase in pulmonary arterial pressure ≥25 mm Hg at rest. Pulmonary arterial hypertension develops in long-standing left-to-right shunts leading to increased pulmonary blood flow resulting in increased PVR, leading to bidirectional or reversal of shunt.[5,6] These patients with cardiac right-to-left shunts have tendencies of:

- Syncope, paradoxical embolism, stroke, and sudden death
- Polycythemia, hemoptysis with pulmonary infarction
- Left heart failure
- Endocarditis

Thus, even an asymptomatic woman needs periodic evaluation. If MTP is declined, hospitalization is recommended in the second trimester.[5]

During pregnancy, CHD that ultimately evolves into ES is mainly VSD followed by ASD and PDA.[7] In our patients, one case had VSD while the other two had both ASD with VSD.

The pathophysiology is related to cardiovascular changes occurring during pregnancy, where reduced SVR leads to increased right-to-left shunting causing reduced pulmonary perfusion. In such patients, systemic hypotension leads to decreased right ventricular filling pressure. In presence of fixed pulmonary hypertension, such reduced right-ventricular pressure is insufficient to perfuse pulmonary arterial bed. This can be exaggerated in the event of fasting, dehydration, or any condition leading to hypovolemia (like hemorrhage or conduction anesthesia) during pregnancy. Thus both reduced right-ventricular filling and decreased pulmonary perfusion lead to hypoxemia. This hypoxemia causes secondary erythrocytosis or polycythemia or hypercoagulability and thromboembolism tendencies and hence adverse materno-fetal effects due to syncope and sudden death. Therefore, the avoidance of such hypotensive episodes during pregnancy is the principal clinical concern during intrapartum management.

In ES, the reduction in SVR of pregnancy increases right-to-left shunting. The main consequences of these hemodynamic changes are cyanosis and low oxygen saturation due to hypoxemia and secondary erythrocytosis (in response to hypoxemia), leading to polycythemia and hyper-viscosity with coagulation disturbances, heart failure, and serious arrhythmias.[8] The major risk factors for maternal mortality are congestive heart failure (CHF), bleeding/anemia, hematocrit >60%, oxygen saturation <80%, syncope or sudden PVR rise or SVR fall.[6] The maximum maternal death chances are in the peripartum period. The first postpartum weeks are the most critical time. But in all our three cases, the postpartum period remained uneventful.

The presentation may include cyanosis, dyspnea, easy fatigability, signs of right heart failure like raised jugular-venous distension, and extremities' edema. Physical examination may show clubbing and cyanosis (Fig. 1). Case 1 had both. There may be hemorrhagic tendencies like hemoptysis and epistaxis.[9] This was seen in case 3. There may be inspiratory crepts and loud P2 with a systolic murmur in the pulmonary area. Complications like low oxygen saturation (seen in cases 1 and 2), polycythemia (seen in cases 1 and 2), heart failure, endocarditis, and thromboembolic events are common. Chest X-ray may show cardiomegaly with bilateral pulmonary congestion (seen in cases 1 and 2). Electrocardiogram (ECG) may demonstrate right ventricular hypertrophy (RVH) or left ventricular hypertrophy (LVH) in few cases.

Amidst ESP, the fetal mortality rate of 25% and maternal mortality rate of 50% have been observed. Here, fetal mortality was seen in 33.3% of cases. Pregnancy must be avoided in this group of patients. These women are advised against pregnancy or to opt for early MTP within the first 10 weeks and tubal ligation is strongly recommended.[4] Therapeutic abortion is recommended in early pregnancy.[10]

Serial hemoglobin monitoring and hematocrit must accompany antepartum fetal surveillance. Iron supplementation must be given to avoid relative anemia. Hypovolemia should be avoided as hypotension exacerbates right-to-left shunt. Heavy exercise, dehydration, fasting, hemorrhage, high altitude, and air travel should be avoided in the antepartum period.

As most of these patients have a congenital cardiac defect, the risk of CHD in offspring of women with ES is 10% (depending on primary CHD). Thus fetal Echo between 18 and 22 weeks is recommended for these antenatal patients.[6,11]

In view of 30% risk of FGR, antenatal sonographic fetal surveillance becomes important. Maternal arterial oxygen saturation pressure should be aimed at a level ≥70 mm Hg as maternal hypoxemia leads to a high incidence of spontaneous abortions, preterm births (PTB), FGR, and low birth weight (LBW).[11] Moreover, Kansaria has reported that with hematocrit >65%, successful pregnancy outcomes are unlikely.[12]

Bèdard et al. reported that 86% of women with ESP had PTB while 24% had FGR.[13] The degree of maternal hypoxemia is an important predictor of fetal outcome. The fetal risks are due to chances of arterial oxygen desaturation, hypoxemia, and polycythemia. Successful pregnancy is unlikely with a hematocrit >65% and over 30% of fetuses FGR.[12] All three fetuses in our cases had FGR. Brach-Prever et al. showed that at least 54.9% of women had PTB.[14] A delayed diagnosis, delayed presentation, and severity of PAH are risk factors for poor outcome.[15] In our cases, although there was a delayed presentation in two cases yet multidisciplinary approach helped in salvaging the mother as well as fetuses. One fetus who succumbed probably to maternal hypoxemia episode had anencephaly.

Although there are no well-established controlled trials regarding continuous oxygen administration, prophylactic anticoagulation, and pulmonary vasodilator, a Brazilian series of 13 pregnancies showed improved maternal mortality with these.[16] Most of the vasodilators available are category B drugs. With the availability of pulmonary vasodilators, the pregnancy outcomes in ESP have improved and their initiation in early pregnancy has given promising results.[10]

The role of anticoagulants is controversial. The risk of thrombosis is high due to polycythemia. However, in a seven patients review with ES who received prophylactic anticoagulants, Pitts et al. implicated secondary hemorrhage as the cause of death in five patients.[17] We gave postpartum thromboprophylaxis to all three cases and had nil mortality. Pulmonary artery catheterization is also recommended during the intrapartum period. In addition to high inspired oxygen, all efforts should be done to avoid hypovolemia. Left uterine displacement should be done to ensure adequate venous return.

Mukhopadhyay reported a case managed in a tertiary center conservatively with oxygen, IE prophylaxis, and diuretics with successful materno-fetal outcome.[9] Ganguly reported three cases over 3 years where two of three mothers and all three babies survived.[18] Lacassie demonstrated successful use of sildenafil and L-arginine for the management of PAH in pregnancy.[19] Cartago et al. showed two cases of ES treated with sildenafil monotherapy leading to maternal stabilization and optimum clinical outcome.[20] Sildenafil (pulmonary vasodilator) along with oxygen was used successfully in all our patients with an optimal outcome.

The ideal mode of delivery is debatable. Cesarean section (CS) should be avoided as it significantly reduces circulating blood volume. Vaginal delivery appears to be safer.[4,10,21] Gleicher et al. reported 34% mortality as compared to 75% mortality with CS.[22]

Of all anesthetic techniques, epidural and incremental spinal anesthesia are the preferred ones.[4] Even for labor analgesia, epidural is considered a safe choice. General anesthesia can lead to a significant decrease in SVR thereby lowering the right-to-left

Table 3: Key points to be followed in the management of Eisenmenger syndrome in pregnancy

Sl. No.	*Key practice points*	
1	Preconception	• Advise against pregnancy • Tubal ligation; contraception choice • Comprehensive cardiology evaluation
2	Antepartum	• Offer therapeutic termination till 10 weeks • Multidisciplinary care at the tertiary center (If MTP declined/higher gestation) • Cardiologist opinion and treatment review • Role of pulmonary vasodilators • Role of prophylactic anticoagulants (controversial) • Avoid fasting/heavy exercise/high altitude/air travel/dehydration/hemorrhage/anemia • Serial Hb and hematocrit evaluation • Iron supplementation • Oxygen saturation monitoring at each visit
3	Intrapartum	• Delivery must be at a tertiary center with multidisciplinary care • Maintain fluid balance (avoid both volume depletion and volume overload) • Avoid hypotension • Continuous pulse oximetry • Oxygen by face mask • Use air filters in I/V lines to prevent paradoxical air embolism • Infective endocarditis prophylaxis • Vaginal delivery preferred. Cesarean section only for the obstetric indication • Assisted second stage
4	Postpartum	• Alert for postpartum hemorrhage • Prevention of DVT • Avoid dehydration • Early ambulation • Stockings • Postpartum thromboprophylaxis • Cardiac follow-up • Counsel for permanent sterilization

shunt and leading to difficult extubation. In case 3, CS was done under epidural anesthesia.

Intrapartum fluid balance, oxygen saturation maintenance (as it provides an estimate of the extent of right-to-left shunt), and strict BP control are mandatory. Optimizing BP is very critical to maintain a balance between systemic and pulmonary blood flow as decreased SVR or increased PVR may lead to increased right-to-left shunting leading to an increase in hypoxemia and increased feto-maternal death risk.

Thus, aim during labor is to maintain oxygen saturation, pain relief, and avoid hypotension (decreased vascular resistance). Intrapartum endocarditis (IE) prophylaxis should be given.[10] The second stage should be cut short and assisted delivery is recommended.[10] Cesarean section (CS) should be done only for an obstetric indication as already discussed. All measures to prevent deep venous thrombosis (DVT) should be taken as these patients have hyperviscosity although the role of anticoagulants is controversial as discussed already. Early ambulation, use of elastic stockings, and anticoagulant administration 12 hours postdelivery should be practiced.

Venovenous extracorporeal membrane oxygenation (ECMO) has been utilized in peripartum settings for stabilizing maternal hemodynamics and optimizing fetal oxygenation.[23]

Patients with ES should refrain from pregnancy. Tubal ligation is strongly recommended. Progesterone-only contraception, progesterone-only pills, depot preparations, and progesterone implants are safe as they don't have a tendency for thrombosis, unlike combined contraceptive methods. Intrauterine devices especially Copper devices are not recommended as they can cause menorrhagia and can increase the risk of endocarditis. Levonorgestrel intrauterine contraceptive device is considered safer. Partner vasectomy may be an option for couples in a monogamous relationship. But since the male partner outlives the female partner, the decision of vasectomy should be contemplated keeping in mind that later he may wish to have a family.[10]

The studies by Katsuragi et al. in their data on the Japanese females and Subbaiah et al. in their data on the Indian females found an increase in pulmonary arterial blood pressure (PABP) in later stages of pregnancy.[24,25] This increase in PABP may cause decompensation and deterioration of symptoms in advanced pregnancy.[25] These changes in PABP can be monitored by right heart catheterization. In the studies cited above, the women were monitored by Echocardiography. This was the limitation of our study. We could not monitor the changes in PABP in the first two cases as the time of presentation was in the third trimester and the duration of the antenatal period was small as compared to these studies. In the third case, however, Echo done in the third trimester picked up the raised PABP.

The key points of management of a pregnant female with ES are summarized in Table 3.

Thus, multidisciplinary care at a tertiary center equipped with an obstetrics critical care unit can help in achieving optimum pregnancy outcomes in ESP.

Conclusion

Eisenmenger syndrome in pregnancy is a very high-risk condition. Ideally, pregnancy is contraindicated. In case of unplanned pregnancy, MTP must be advised. But in cases where pregnancy goes beyond the GA limit for MTP, antenatal care and delivery should be at a tertiary center under multidisciplinary care involving a cardiologist, obstetrician, intensive care physician, anesthetist, and neonatologist for the optimum materno-fetal outcome.

Clinical Significance

Managing ESP, calls for dedicated multidisciplinary care at a tertiary center preferably with obstetrics critical care unit. The knowledge of key practice points while managing this rare condition in pregnancy can definitely help in reducing maternal morbidity and mortality.

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