



A RARE CASE REPORT ON EISENMENGER'S SYNDROME WITH PREGNANCY : A GLIMPSE OF LIFE IN OCEAN OF DEATH

Obstetrics & Gynaecology

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ABSTRACT

Pregnancy complicated with Eisenmenger syndrome is associated with high risk to the fetus as well as the mother. There is approximately 50% risk of sudden maternal death, frequently occurring a few days postpartum and the overall fetal wastage is reported to be up to 75%. Patients with Eisenmenger syndrome are advised to refrain from pregnancy or to terminate pregnancy by the end of first trimester itself. Management of these patients requires a co-ordinated multi-specialist care when such pregnancies reach a stage where safe termination is not advisable. However, in spite of all the risks, a few patients deliver successfully with a good maternal and neonatal outcome. We present 2 cases reported till third trimester and delivered a healthy baby and were subsequently discharged on the 10th postpartum day without any serious complications.

KEYWORDS

Pregnancy, Eisenmenger's syndrome, Mortality

CASE REPORT 1

- 31 year old primi gravida patient referred to our emergency department from PDU medical college RAJKOT as a 37 weeks 3 days gestational age with chief complaint of abdominal pain & breathlessness since 5 days with known case of congenital heart disease (large inlet ventricular septal defect), Eisenmenger syndrome & severe PAH.
- Our patient was k/c/o congenital heart disease diagnosed since 2 years with large peri membranous ventricular septal defect with bidirectional shunt with eisenmenger physiology.
- Patient was on Tab. ENDOBLOC T (AMBRISANTAN + TADALAFIL) 0-1-0 before pregnancy. Patient had taken routine antenatal care from private hospital. But fetal 2D ECHO was not done.
- On examination she was normal stature and fairly built, weight around 54kg. Examination revealed no edema or clubbing. Vitals were Spo2 = 90% on room air, with pulse rate 88 beats/min and blood pressure 110/76mmhg in right upper limb, her respiratory rate was 18/min. On CVS examination she had thrills over apex. Pan systolic murmur were present. Apex beat was shifted to midclavicular line & left paratracheal heave present. Respiratory examination revealed bilateral vesicular breath sound in all lung fields.
- Abdominal examination shows uterus 36 weeks size with cephalic presentation with floating head, fetal heart rate 146 beats/minute taking contractions of 5 secs /2/10 minutes. On per vaginal examination cervical os was 2 finger loose, 20% -30% effaced, vertex presentation, membrane present with high station.
- On investigations Hb = 15.2 gm/dl, hematocrit = 46.4 %, Platelets were 1,34,000/cmm.
- Cardiology consultation done, advised immediate termination of pregnancy. Patient was prepared for emergency LSCS in view of early delivery in case of primi patient in early stage of labour under general anesthesia after high risk consent. A full term male child with 2.5 kg baby weight delivered in cephalic position, liquor was clear & adequate & baby cried soon after birth. APGAR 7/8/9.
- Intrapartum period was uneventful, 4 unit Platelets were transfused, but patient did not reversed, not extubated electively. Shifted to ICU for elective ventilation on CMV mode of ventilation post operatively. Post operatively spo2 96% on CMV mode of ventilation, P = 120/min, BP= 142/84mmhg. No crepitations on respiratory examination. Uterus was well contracted with No active bleeding per vaginum.
- Postpartum cardiology consultation done advised NO active intervention.
- Patient extubated on POD2 gradually to CPAP & then on NRBM &

kept on NRBM for 1 day & gradually weaned off to simple o2 mask on POD3, then shifted to ward on room air. 1 episode of syncope with spo2 dropped to 70% was there on starting phase of mobilization; which was immediately recovered by itself. 1 unit PCV was transfused postnatally to prevent adverse outcome with low oxygen saturation.

- Both mother & healthy baby were discharged after cardiologists opinion with vitals of spo2 = 90% on room air, p= 78/min, BP= 110/60mmhg. Different Contraceptive methods were advised.

CASE REPORT 2

- Another 20 year old Primi patient who came to emergency department at 36 weeks gestational age with chief complaint of abdominal pain and breathlessness for 2 days.
- Our patient was known case of ventricular septal defect with eisenmenger syndrome with severe tricuspid regurgitation and pulmonary hypertension which was diagnosed at 6 month of amenorrhea. Patient was on regular medication (tablet toresemide 1 tablet once a day for three months).
- Patient did not take any ANC visit. On first visit at 6 month amenorrhea ANC doppler study was done is s/o compromised utero placental circulation with reversal of umbilical artery flow.
- Fetal cardiac condition s/o moderate cardiomegaly, left axis deviation, veno arterial and ventriculoatrial concordance noted, right atrium and right ventricle enlarged, right ventricular hypertrophy, pulmonary artery mildly dilated with pulmonary edema.
- On examination she was normal stature and poorly built, weight around 47 kg. Examination revealed edema, clubbing on all four limbs and vitals were stable with pulse rate 92 beats/min and blood pressure 130/90 mmhg in right upper limb. Her respiratory rate was 35/minute and JVP was raised. On CVS examination she had thrills over apex, pansystolic murmur with P2 loud. Apex beat was shifted to mid clavicular line and left paracentral heave was present. Respiratory examination revealed bilateral vesicular breath sounds in all lungs fields. Abdominal examination uterus 30-32 weeks size relaxed with cephalic presentation with fetal heart rate 140 beats/minute. On per vaginal examination cervical os was 2 finger loose, 30% effaced, membrane present with station -2
- On investigation hb 18.5 gm, hematocrit 61.50%
- Ecg showed irregular rhythm with ventricular hypertrophy. Cardiology consultation was obtained after she was put on sildenafil, dytor and spironolactone. Cardiologist opine to cut down second stage of labour. Patient was put on complete bed rest with 45 degree inclination with oxygen. Injection Ampicillin,

injection Betnasol for fetal lung maturity was given. Labour was augmented by 5 unit oxytocin and delivered normally after high risk consent . A pre term female baby weight 1 kg delivered with vertex presentation, liquor was clear and adequate and baby cried soon after birth. Apgar score 5/6/7.

- Intrapartum period was uneventful and fluid input and output was monitored strictly. On 1st post-partum day, her vitals were normal. Patient was admitted in Icu. Respiratory examination revealed bilateral crepitation present . In ICU she was given head up position, o2 inhalation via venti mask , higher anti biotics and patient kept for observation . Uterus was well contracted with no active bleeding per vaginum. Her condition was stable and she was transferred to cardiology hospital for further management where she was managed conservatively and stated on tab Toresemide / tab Spironolactone and tab Sildenafil. Patient was discharged on post partum day 5 from cardiology hospital. Patient was advised regarding contraceptive methods for future.

DISCUSSION

- The incidence of Eisenmenger syndrome is approximately 3% of patient with congenital heart defects . Due to high maternal & perinatal mortality rate approx 50% (Gleicher 1979) ; Eisenmenger syndrome is considered to be an absolute contraindication to pregnancy. (1)
- A Ventricular septal defect is the most common underlying shunt defect. Maternal mortality in association with VSD is higher (60%) than in association with ASD (44%) and PDA (41.7%). (1)
- A high incidence of maternal death was associated with hypovolemia, thromboembolic phenomena and pre eclampsia. (2)
- Therefore termination of pregnancy is a treatment of choice. (3)
- However , if a patient chooses to continue pregnancy ; even after risks have been explained; hospitalization for the duration of pregnancy is often appropriate.
- Fetal outcome correlates well with maternal hematocrit in cyanotic heart disease and successful pregnancy is unlikely with a hematocrit >65%.
- There is no evidence to support the choice in of either vaginal or cesarean delivery for cardiac reasons: vaginal delivery is associated not only with a lower average blood loss but also increased maternal effort. Gleicher et al. reported a 34% mortality associated with vaginal delivery and a 75% mortality associated with cesarean section. (6)
- The third trimester fetal surveillance with ultrasound and antepartum testing is important because at least 30% of the fetus will be growth retarded. Maternal arterial partial pressure of oxygen should be maintained at a level of 70mmHg or above . This is explained by high incidence of spontaneous abortions , a 30-50% risk of premature delivery and low birth weights as maternal hypoxia disturbs fetal growth. (5)
- The unique feature in our cases are that despite having an Eisenmenger syndrome with associated VSD our patient conceived , continued pregnancy without much major symptoms ; and delivered a healthy babies.

CONCLUSION

Although pregnancy should be discouraged in women with Eisenmenger's syndrome, it can be successful as seen in our cases wherein co-ordinated multispecialist care and proper monitoring helped in avoiding any adverse maternal or fetal outcome. These cases may help in giving glimpse of hope to all such pregnancies complicated with Eisenmenger syndrome (ocean of death) at an advanced gestational age where termination is not possible.

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