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A Case of Emergency Caesarean Section in Patient with Large Atrial Septal Defect with Severe Pulmonary Arterial Hypertension

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ABSTRACT

Pregnancy with pulmonary hypertension carries a very high risk of morbidity and mortality and therefore women with severe pulmonary hypertension are advised not to become pregnant. However, many women with severe pulmonary hypertension become pregnant and present with complications. Here we present successful management of a patient with severe pulmonary hypertension presented for emergency caesarean section under epidural anaesthesia.

Key words: pulmonary arterial hypertension (PAH), atrial septal defect (ASD), pregnancy, epidural anaesthesia.

INTRODUCTION

During pregnancy many physiological changes occur to meet the nutritional demand of developing foetus. These changes are well tolerated in a healthy pregnancy. However, in pregnant women with cardiac diseases, these changes increase the complications and may lead to increased maternal mortality and morbidity. One such cardiac condition is pulmonary arterial hypertension.

Uncorrected congenital heart disease is the most common cause of pulmonary hypertension in developing country like India. [1] Advancement in surgical intervention of congenital heart diseases has increased the survival of patients up to child bearing age group and has resulted in increased incidence of pregnant women presenting with cardiac conditions. [2]

Here we present anaesthetic management of pregnant women with severe pulmonary hypertension with Pulmonary Artery Pressures (PAP) of 120 mm hg due to large ASD for emergency low segment caesarean section (LSCS).

CASE REPORT

A 30-year-old female G4 P1 L1 A2 at 30 weeks of gestation presented with epigastric pain and breathlessness. She was a known case of atrial septal defect with situs inversus diagnosed at seven years of age. She was diagnosed with severe pulmonary hypertension in her previous pregnancy in 2009 when she presented with intrauterine growth retardation (IUGR). The PAP then was 90mmhg. She had undergone an uneventful LSCS under general anaesthesia. She was advised during that admission to avoid future pregnancies. She had been discharged on oral Sildenafil which had been discontinued in current pregnancy and had been started on Aspirin. She was on Thyroxin for hypothyroidism.

On presentation, uterine ultrasound revealed severe IUGR with reversal of diastolic flow with oligohydramnios. Hence, she was scheduled for an emergency LSCS.

On preoperative evaluation, her heart rate was 76/min, BP was 126/70 mm Hg, SPO2 was 86% at room air and was 92-93% with oxygen on nasal prongs at flow of 4l/ min. We determined her functional capacity to be less than 4 METS. Auscultation was significant for loud P2 with narrow S2 split. Her lab work revealed haemoglobin of 12.8 g/dl with normal liver function tests, renal function tests and coagulation profile. ECG showed sinus rhythm with T wave inversions in V1 to V6. 2D ECHO showed large SV type ASD, 30mm in diameter with bidirectional shunt. The right atrium, right ventricle and pulmonary artery were dilated. The PASP was 120mm Hg. The left ventricle function was normal with no RWMA. EF was 60%.

A multidisciplinary informed consent was taken from the patient and patient's family highlighting the perioperative risks involved.

Intra operative course:

When patient arrived at OT, ASA standard monitors - ECG, pulse oximeter and NIBP were connected. Her initial vital signs were: HR was 97/min, NIBP was 189/84 and SPO2 was 93% with 4L/min supplemental oxygen. Then a 20 gauge right radial arterial line and a 7Fr triple lumen right internal jugular central venous catheter were placed for continuous IBP and CVP monitoring respectively.

With patient in sitting position, a central neural epidural block was performed with 18G tuohy needle at L1- L2 intervertebral disc space with loss of resistance technique to saline and an epidural catheter was threaded and fixed at 10cm. A test dose of 3 ml of 2% Lignocaine with 1:100,000 Adrenaline was injected via epidural catheter. Intravascular or intrathecal placement of catheter was ruled out. 100 mcg (2ml) of Fentanyl was given through the epidural. Epidural anaesthesia was then established in a graded manner with 0.5 % Bupivacaine (3 ml every 5mins). A total 15 ml (12 ml 0.5% Bupivacaine + 3 ml of the test dose) was given. Block achieved up to T6 level. A low dose Phenylephrine infusion (20mcg/ml) was started to support blood pressure. Four minutes into surgery, a live male baby delivered. The Apgar score was 7 at 1 minute, 7 at 5 minute and 8 at 10 minutes. Birth weight was 850g. Baby was shifted to NICU for severe IUGR. Following delivery, 10 units of Oxytocin was given as slow IV infusion. 100 mcg of Carbetocin was given IM. The patient received a total 1100 ml of crystalloids. Her urine output was 150ml/hr and blood loss was 250 ml. Blood pressure was maintained above 130 mmHg systolic with Phenylephrine infusion throughout the surgery.

Post operatively epidural infusion with Ropivacaine 0.2% at 2ml/hr was started for postoperative pain relief. Postoperative vitals were stable. Patient was shifted to ICU for observation. Anti coagulation with Injection Clexane was started after 12 hours. Epidural catheter was removed the next day. She was shifted to ward with supplemental oxygen on day two of surgery and was discharged home after 1 week in a stable condition.

DISCUSSION

Pulmonary hypertension (PH) is defined as mean pulmonary artery pressure (PAPm) ≥25 mm Hg at rest, measured during right heart catheterization. Pulmonary arterial hypertension (PAH) is a subset of pulmonary hypertension, defined as PAPm ≥ 25 mmHg and pulmonary artery wedge pressure (PAWP) ≤15 mm Hg with a pulmonary vascular resistance >3wood units.[1,3,4,].

WHO (World Health Organization) divides pulmonary hypertension into five categories. [3, 5, 6, 7]

1) Primary pulmonary hypertension (familial and sporadic) and PAH related to collagen vascular disease, congenital systemic to pulmonary shunts, HIV

infection, portopulmonary hypertension, drug induced.
2) PAH related to disorders of the respiratory system or hypoxemia
3) Pulmonary venous hypertension due to mitral valve disease, chronic left ventricular dysfunction and pulmonary veno occlusive disease.
4) PAH due chronic thrombotic and/or embolic disease.
5) PAH related to disorders directly affecting the pulmonary vasculature (inflammatory pulmonary capillary hemangiomatosis)

Our case is PAH due congenital heart disease 'atrial septal defect' (category 1). ASD is one of the most common congenital heart diseases causing pulmonary hypertension in pregnancy. It is an acyanotic condition with left to right shunt. There are three types of ASD - Ostium primum type, ostium secondum type and sinus venosus (SV) type. In the third type the defect is located in the upper segment of the atrial septum close to the superior vena cava and can lead to multiple complications like pulmonary arterial hypertension, cardiomegaly, arrhythmias and finally myocardial ischemia.[8,9] Our patient had SV type of ASD with severe pulmonary arterial hypertension.

Chronic uncorrected left to right shunt produces right ventricle volume overload, pulmonary congestion, pulmonary vascular remodelling and increased pulmonary arterial pressures which in turn leads to right ventricular hypertrophy and right ventricular dysfunction. When pulmonary arterial pressures exceeds systemic pressure there will be reversal of shunt i.e. the shunt direction changes from "left to right" to "right to left". This results in hypoxemia. The condition is termed as Eisenmengers' syndrome. Once this occurs correction of ASD will not help reverse the PAH [2, 3, 4, 9] and specific treatment for pulmonary hypertension needs to be started. There are a number of pulmonary vasodilators now available like endothelin receptor antagonist (Bosentan, Ambrisentan), phosphodiesterase 5 inhibitors (Sildenafil, Tedalafil), prostacycline analogues (Iloprost, Epoprostenol, Treprostinil) and inhaled nitric oxide[1,4]. Intravenous vasodilators like Milrinone, Dobutamine, Nitroglycerine, Sodium Nitroprusside and Prostacycline can also be used to reduce right ventricle after load. But they are not specific to pulmonary vasculature and can produce significant systemic vascular dilatation and hypotension and hence are used with caution. [6]

In pulmonary hypertension, if systemic arterial pressure falls below pulmonary arterial pressure, coronary blood flow to right ventricle also will decrease. Normally right ventricle receives blood flow both during systole and diastole. If the right ventricular systolic pressure becomes equal to or more than systemic pressure or if the right end diastolic pressure becomes higher, coronary blood flow decreases. This leads to myocardial ischemia and right ventricular failure. [10] Therefore it is very important to always maintain systemic pressure above pulmonary artery pressure. Various vasoconstrictors are used to increase systemic vascular resistance and therefore systemic arterial pressure.[4] Norepinephrine, phenylephrine and vasopressin have all been used successfully.[3,4,6,10,11,12,13] Various studies done on use of vasoconstrictors in pulmonary hypertension give conflicting results as which one is better than other.

In pregnancy small ASD with moderate left to right shunt is well tolerated. However in the presence of large ASD with severe PAH, the normal physiological changes of pregnancy cannot be tolerated. The physiological changes of pregnancy which worsens pulmonary hypertension are as follows:[2, 14, 15]

1) About 50% increase in intravascular volume that peaks by mid trimester, increases right ventricular preload.
2) Progressive decrease in systemic vascular resistance due to vasodilating effect of estrogen and progesterone, reduces systemic pressure less than PA

pressure leading to decreased coronary perfusion. It also causes reversal of shunt.

3) About 15% increase in heart rate, increases myocardial oxygen demand in already compromised coronary perfusion.
4) Reduced functional vital capacity further worsens hypoxia.
5) Increase in total blood volume by around 500 ml during each uterine contraction causes volume overload and increases right ventricular pre load
6) Hypercoaguability of pregnancy increases risk of thrombo embolic event.

Anaesthetic goals in pregnant patients with pulmonary arterial hypertension includes: [3, 10, 14, 16]

1) Avoid increase in PAP by avoiding hypoxia, hypercapnia, acidosis, pain and hypothermia.
2) Maintain mean systemic arterial pressure within 15% above or below the baseline. Systemic pressure should always be higher than pulmonary arterial pressure.
3) Avoid tachycardia and arrhythmias as this may increase myocardial oxygen demand.
4) Avoid acute hypovolemia which decreases systemic vascular resistance and lead to reversal of shunt.
5) Avoid sudden hypervolemia
6) Avoid even small amount of accidental intravenous injection of air as it can lead to systemic embolization.

All the above goals are better met with regional anaesthesia compared to general anaesthesia. In a study done by Elisabeth Bedard et al, higher mortality was observed in pregnant women with PAH receiving general anaesthesia. [17]

Disadvantages of general anaesthesia in pulmonary hypertension are: [4, 16]

1) All general anaesthetic agents have negative ionotropic effect and therefore reduce RV contractility in PAH.
2) Intubation response like tachycardia and hypertension increases pulmonary artery pressure.
3) Positive pressure ventilation increase increases RV after load.
4) Chances of aspiration is more in pregnancy due to delayed gastric emptying.
5) Pregnant women also have difficult airway both for mask ventilation and intubation

Disadvantages of spinal anaesthesia: Spinal anaesthesia causes acute massive sympatholysis resulting in profound vasodilatation and systemic hypotension. Fall in systemic pressure below PA pressure increases the risk of reversal of shunt via the ASD and also causes coronary hypoperfusion. [2, 4]

Unlike spinal anaesthesia, epidural anaesthesia causes gradual sympatholysis, very slow changes in preload and after load and no sudden fall in blood pressure. This gives time for the heart to compensate and allows relatively better haemodynamic stability. [18]

Dr Saya Raghavendra Prasad *et al* have also done lscs in pulmonary hypertension with right ventricular pressure of 90 mmHg, under sole epidural anaesthesia. [12] In a case report by Dr Raja Avinash *et al*, the authors performed a LSCS in severe pulmonary hypertension with PA pressures of 128 mmHg under combined spinal epidural anaesthesia where Fentanyl was given in subarachnoid space followed by 0.5% Bupivacaine + Fentanyl in epidural space. [19] Similarly Dr Bhavana Gupta *et al* have done LSCS in PH under combined spinal anaesthesia. [16]

We preferred graded epidural anaesthesia over Combined Spinal Epidural as the onset and level of blockade can be titrated. Hence sudden fall in blood pressure can be prevented and good hemodynamic stability can be achieved. [18]. Epidural anaesthesia avoids airway manipulation and risk of aspiration. It also provides good postoperative analgesia.

CONCLUSION

Pregnancy complicated by pulmonary hypertension is a life-threatening condition.

A thorough knowledge of physiological changes of pregnancy and pathophysiology of pulmonary hypertension is necessary to administer a safe anaesthesia. Regional techniques like graded epidural or combined low dose spinal epidural can be safely administered in patients with severe PAH.

Declaration by Authors

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