CASE REPORT

Primary pulmonary hypertension during pregnancy: A case report

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We describe a case of a 25-year-old pregnant woman who presented with severe primary pulmonary hypertension (PPH). Her echocardiogram showed severe right ventricular hypertrophy with dilatation and Moderate right ventricular systolic dysfunction. Right ventricle systolic pressure (RVSP) was estimated to be 125 mm Hg. She had an elective caesarean section under general anaesthesia at 32 weeks of gestation. Pulmonary artery pressures measured by a pulmonary artery catheter before anaesthesia were 102 mm Hg and pulmonary vascular resistance was 429. Intraoperative nitric oxide was used to reduce pulmonary artery systolic pressure (PASP). After the delivery of a healthy infant, PASP was controlled with nebulized iloprost and silandifil. Five days later she was transferred from intensive care unit after she was started on silandifil 50 mg three times daily and a small dose of warfarin.

Keywords: Primary pulmonary hypertension, Pregnancy, General anaesthesia, Pulmonary vasodilators

Introduction

The mortality rate of primary pulmonary hypertension (PPH) complicating pregnancy is very high [1,2]. The only long-term 'cure' is a heart–lung transplant. Increased pulmonary vascular resistance combined with the normal physiological changes of pregnancy and delivery is difficult to manage. A successful outcome has been described following general anaesthesia for caesarean section in a woman with PPH and coarctation of the aorta [4]. We describe in this report the management of a pregnant lady with severe PPH delivered successfully with elective caesarean section under general anaesthesia.

Case report

A 25-year-old primigravida presented at 32 weeks of gestation with one month history of progressive dyspnea associated with central localised chest pain, palpitation and mild intermittent cough. There was no significant past medical history.

On examination she was tachypneic and tachycardic. Arterial pressure was 100/60 mmHg. She had signs of right ventricular overload including elevated jugular venous pressure (14 cm above sternal angle), lower limb edema and right parasternal heave. On auscultation she had an accentuated pulmonary component of the second heart sound and a pan systolic murmur of tricuspid regurgitation.

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ECG showed right heart strain pattern, and chest radiograph showed a prominent right heart soluette with prominent pulmonary arteries.

An echocardiogram showed severe right ventricle (RV) dilatation and hypertrophy (RVH) with moderate RV systolic dysfunction in addition to a huge right atrium (RA), Figs. 1 and 2, and severe tricuspid regurgitation (TR). Right ventricle systolic pressure (RVSP) was 125 mmHg (Fig. 3) with no evidence of left to right shunt with negative contrast injection. Left side of the heart was normal.

A ventilation/perfusion scan and duplex ultrasound (US) of the lower limbs excluded pulmonary embolic disease. Blood work for vasculitis screen did not reveal any evidence of vasculitis or connective tissue disease.

A clinical diagnosis of PPH was made and she was transferred to the intensive care unit and assessed by cardiology and pulmonary teams. She was started on diuretics and her dyspnea and lower limbs edema improved. Additionally she was given dexamethazone, silandifil 50 mg PO three times daily and intravenous infusion of heparin for two days prior to delivery.

An elective caesarean section and tubal ligation under general anaesthetic was planned after two days of admission. She was prepared for the procedure and anaesthetised by cardiac anaesthesia team. In the operating room she received midazolam 1.5 mg i.v. after which an arterial cannula and pulmonary artery catheter were inserted. Systolic pulmonary artery pressure (PAP) was 102 mmHg. After pre-oxygenation, anaesthesia was induced with propofol 40 mg, fentanyl 1 mg. She was intubated and the ventilated with oxygen and isoflurane. Vecuronium was used for muscle relaxation.

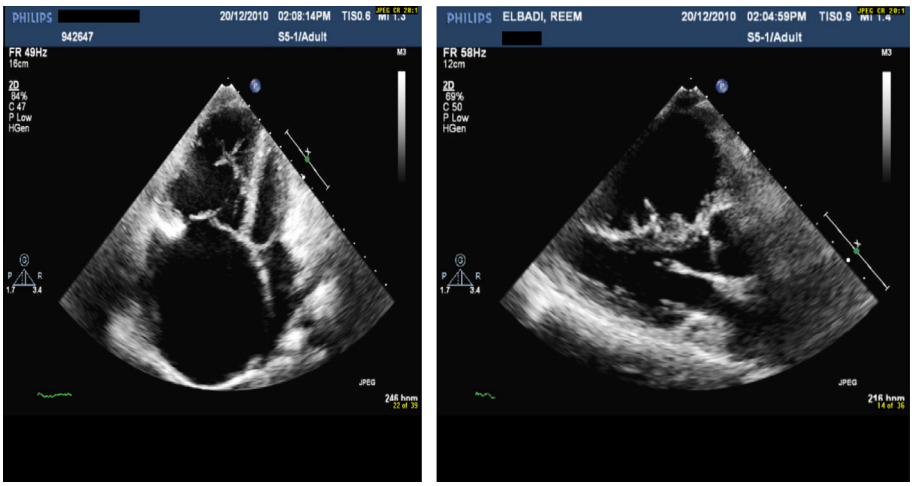

Figure 1. Severe right ventricle and right atrium dilatation.

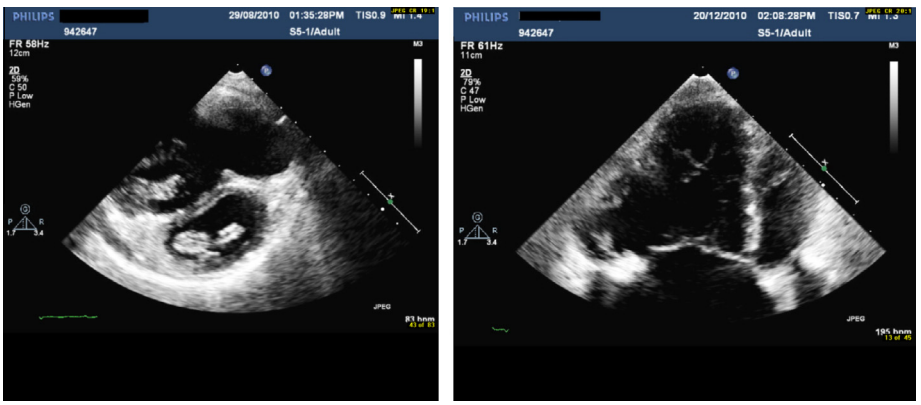

Figure 2. Severe right ventricular dilatation with right ventricular hypertrophy. Flattening of interventricular septum in systole indicating significant right sided pressure overload.

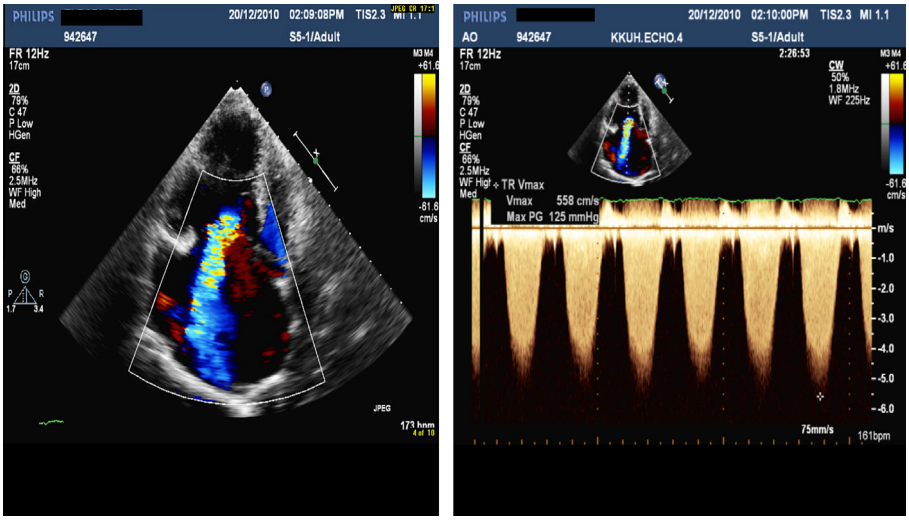

Figure 3. Severe Tricuspid regurgitation with peak TR velocity of (5.5 m/s) with high right ventricular systolic pressure (RVSP) of 125 mm Hg.

Nitric oxide 1.5 ppm was given after induction of anaesthesia and continued during the rest of the procedure. A live, healthy baby girl was delivered 10 min after induction of anaesthesia. The baby appeared heavily narcotized but promptly responded to naloxone. Augmentin 1.2 g i.v. was given after delivery and syntocinon 10 units i.v. as an infusion over 20 min to contract the uterus, which caused a slight increase in pulmonary artery pressures to 120 mmHg. Nitric oxide was continued after delivery and was well tolerated. The dose was gradually increased to 12 ppm. PAP decreased progressively to 90 mmHg by the end of the procedure. She was subsequently transferred to intensive care unit. The patient was extubated same day, 9 h after surgery with no complications.

Postoperative echocardiography was unchanged. Pulmonary artery systolic pressure (PASP) was equal to 102 mmHg. Intravenous heparin was recommenced four hours post-surgery to maintain an activated partial thromboplastin time of 2–2.5 the normal. Two hourly nebulized iloprost was introduced. This enabled the nitric oxide and pressure support ventilation to be weaned without any rebound increase in PAP. PAP remained at or slightly below systemic. Cardiac output remained fairly stable (8.1–9.2).

Post operative clinical course was uneventful in the intensive care unit and patient hemodynamics showed increased pulmonary vascular resistance (PVR) up to 800 and PAP up to 96. She was advised to continue on iloprost nebulizer 20mcg Q 6 h and silandifil 50 mg three times a day as well as home O2. She was also started on warfarin and was discharged from intensive care unit on day five post operatively and from the hospital in day 11 days postoperatively in a stable clinical condition.

She was given the appropriate outpatient clinic appointments to be followed and assessed for heart and lung transplantation.

Discussion

PPH is defined as a sustained elevation of PASP (mean greater than 25 mmHg at rest) in the absence of a demonstrable cause. Pulmonary vasoconstriction, medial hypertrophy, thrombosis in situ and dysfunctional pulmonary vascular endothelium are believed to be the underlying contributing mechanisms [4].

Pulmonary hypertension is poorly tolerated during pregnancy. Deterioration typically occurs in the second trimester with symptoms of fatigue, dyspnoea, syncope and chest pain. This corresponds to the physiological increase in cardiac output and blood volume of 40%. During labour, uterine contractions effectively add 500 ml of blood to the circulation. The labour pain increase right atrial pressure, blood pressure and cardiac output [5]. Women with PPH is advised against pregnancy. In early pregnancy a termination is considered. Where PPH is not diagnosed until late pregnancy an elective delivery with caesarean section is preferred. This facilitates cooperation between specialities, permits monitoring to be started in advance, the pain and haemodynamic consequences of labour to be minimized and an intensive care bed to be arranged. Premature spontaneous labour is common [2] therefore delivery is usually planned for 32–34 weeks gestation. In our patient the cardiovascular physiological

changes of pregnancy had already occurred by the time of presentation. Identification of these hemodynamic changes has led to the use of anticoagulants, oxygen, and vasodilators, which have been shown in several reports to lead to an improvement in hemodynamics and outcome in nonpregnant patients with PPH [6].

An opioid-based general anaesthetic was considered appropriate for a failing right ventricle. It facilitated the control of PAP and the use of nitric oxide. Nitric oxide can be administered by facemask but is poorly tolerated, difficult to monitor and cannot be scavenged. A narcotic-based technique minimizes increased pulmonary pressures during laryngoscopy and avoids the excessive negative intropic effect of inhalational agents. Care was taken to avoid reducing venous return during positive pressure ventilation. Narcotic-related neonatal depression is usually easily managed. O'Hare et al. [3] reported a successful outcome following an emergency caesarean section under general anaesthetic for a woman with PPH and coarctation of the aorta. They gave intravenous and aerosolized prostacyclin postoperatively.

Vasoconstriction is a prominent feature, leading to the rationale for using pulmonary vasodilators such as oxygen, nitric oxide, epoprostenol and iloprost in the short term and calcium antagonists in the long term. Nitric oxide has been used in the pre and post-delivery management of PPH in pregnancy, with clear reductions in PAP as evidenced by cardiac catheterization data, [8] but not during caesarean section.

Epoprostenol is a naturally occurring prostaglandin and potent vasodilator. It affects vascular remodelling and inhibits platelet aggregation. Iloprost is a synthetic analogue of epoprostenol with improved metabolic and chemical stability, which decreases PAP and PVR, increases cardiac output, and has minimal effect on systemic arterial pressure [9]. The pulmonary vasodilator effect lasts 60–120 min. In comparison, intravenous prostacyclin (epoprostenol) reduces PVR with similar efficacy but reduces systemic arterial pressure to a greater degree [10] with no clinically significant reduction in PAP. Neither iloprost nor prostacyclin is recommended in pregnancy because of concerns over the effect on uterine blood flow.

Patients with PPH are at risk of thrombosis and thromboembolism. The Mayo Clinic group has reported that anticoagulation may improve the outcome in severe pulmonary hypertension [11].

Pulmonary artery catheterization provides early warning of rising PAP, deteriorations in right ventricular function and the effects of therapeutic interventions. Its use has not been associated with improved survival [7], and there is an increased risk of pulmonary artery rupture and thrombosis in these conditions [12].

PPH complicating pregnancy remains a fatal condition with deaths reported to occur between two and nine days post-delivery [1], usually from right heart failure. Iloprost and nitric oxide therapies may have a role in controlling PAP in this condition but there is no evidence of improved survival.

An important component in the successful management of these patients involves a multidisciplinary team approach with an obstetrician, pulmonary or cardiology specialist, anesthesiologist, and experienced nursing staff [12].

Summary

PPH is likely to worsen during labour and delivery, resulting in a high maternal mortality rate. Early recognition and treatment with vasodilator and anticoagulation therapy may reduce the likelihood of complications. Elective caesarean section may be performed with intraoperative vasodilator administration. A multidisciplinary approach to the management of patients with PPH during pregnancy is of great importance for a successful maternal-foetal outcome.

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