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Review Article

Surgical repair of atrial septal defect with severe pulmonary hypertension during pregnancy: a case report with literature review

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Abstract We are reporting a case of a 37-year-old pregnant woman with a large secundum atrial septal defect with left-to-right shunt and severe pulmonary hypertension. Her atrial septal defect was undiagnosed before this pregnancy. After carefully considering all the options, we repaired her atrial septal defect with an open heart surgical closure at 20 weeks of gestation. A substantial and consistent reduction in pulmonary arterial pressure after the surgery and subsequent uneventful delivery indicate that surgical repair of atrial septal defects is a viable option that should be considered for such patients.

Keywords: Pulmonary hypertension; pregnancy; atrial septal defect; atrial flutter

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PULMONARY HYPERTENSION IS THE MOST OMINOUS cardiac condition during pregnancy. Despite recent achievements in management of pulmonary hypertension, it still is associated with 36% of maternal mortality.[1] In the most severe cases of pulmonary hypertension with Eisenmenger physiology, maternal mortality can reach 50%.[2]

A significant proportion of pulmonary hypertension in young women occurs because of congenital heart defects, most commonly atrial septal defects. Although small atrial septal defects are relatively common in adults, large unrepaired defects are rare. There is a higher possibility of uncorrected defects in women who have immigrated from an undeveloped country.

We are presenting the case of a woman who was diagnosed with severe pulmonary hypertension due to large unrepaired secundum atrial septal defect during her second pregnancy.

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Case

The patient is a 37-year-old Hispanic woman originally from Mexico who first presented for obstetrical care at 18 weeks of gestation. She was referred to the cardiology clinic after she said that she had pulmonary hypertension.

She was diagnosed with some congenital heart defect at the age of 7 years in Mexico, but at that time her mother declined suggested surgery and the girl did not have further follow-up. She was usually more fatigued than her peers but otherwise did not have particular complaints. At the age of 26 years, she had her first pregnancy, which was uneventful. She had a forceps-assisted vaginal delivery secondary to arrest in descent. Her son does not have any known defects and is healthy.

At the age of 34 years, she developed an episode of palpitations and was admitted to an outside

hospital where she had cardiac catheterisation and an echocardiogram. Her atrial septal defect was missed and she was diagnosed with primary pulmonary hypertension, put on sildenafil, and told she needed a heart/lung transplant. She was taking sildenafil for 2 years but then stopped because she felt no difference. She did not have any follow-up until her second pregnancy. This time, when we saw her in our clinic, an echocardiogram was performed. There was a large secundum atrial septal defect, with severe dilation of the right chambers, moderate tricuspid regurgitation, and estimated pulmonary artery systolic pressure of 80 millimetres of mercury. We admitted her to the hospital for further evaluation and management.

On admission, her heart rate was 86 beats per minute, and her blood pressure was 110/70 millimetres of mercury. There was mild elevation in jugular venous pressure and 2/6 systolic murmur heard best at the right sternal border. There was no palpable liver, oedema, or cyanosis, and her abdomen was distended because of 18-week gestation.

A transoesophageal echo demonstrated normal left ventricular ejection fraction of 60–65%, normal valves, markedly dilated right chambers, and a large secundum atrial septal defect measuring 2.9 centimetres, with left-to-right shunt. There was moderate tricuspid regurgitation with a peak gradient of 57 millimetres of mercury. All pulmonary veins were draining into the left atrium. Adding at least 10 millimetres of mercury for right atrial pressure, we estimated her pulmonary artery systolic pressure as being no less than 67 millimetres of mercury.

Cardiac catheterisation demonstrated a pulmonary artery pressure of 65/35 millimetres of mercury, pulmonary capillary wedge pressure of 16 millimetres of mercury, and pulmonary vascular resistance of 4.5 Wood units. Qp/Qs was calculated as greater than 10:1. Because table values used for oxygen consumption for cardiac output calculation may not be accurate in pregnancy, the shunt by echocardiography based on right and left ventricular outflow tract dimensions and time velocity integrals of pulmonary and aortic flow, respectively, gave a very close value of 9. The atrial septal defect was too large, stretched diameter 3.4 centimetres, for a percutaneous closure with any type of occluder device.

A foetal echo was also performed and was consistent with normal foetal anatomy and normal biventricular systolic function.

Owing to concern of haemodynamic changes and demand involved in pregnancy with uncorrected pulmonary hypertension and atrial septal defect, both high-risk obstetrics and cardiothoracic surgery were consulted. The options presented included:

1. Termination of the pregnancy because of severe pulmonary hypertension with subsequent surgical repair of atrial septal defect.
2. Medical management of pulmonary hypertension with continuation of the pregnancy assuming the risk of maternal and foetal mortality.
3. Surgical repair of the atrial septal defect early in the pregnancy and medical management of the pulmonary hypertension after the repair.

Despite counselling regarding the extremely elevated maternal risks for pregnancy, the patient declined therapeutic termination.

After extensive discussion with regard to the health of the mother and the baby and the risks and benefits of procedures, a decision was made for surgical repair of the atrial septal defect. It was universally agreed that this risk would increase if surgery was postponed to a later date in the pregnancy. Successful closure of the atrial septal defect was performed with a large Cormatrix patch at 20 weeks of gestation.

The patient tolerated the surgery well. Her postoperative echo demonstrated improved right ventricular systolic function and pulmonary artery systolic pressure of 50 millimetres of mercury.

Post-operatively, she had an isolated episode of atrial flutter with a rate of 140 beats per minute, which was terminated with electrical cardioversion.

Her follow-up was complicated by several more episodes of symptomatic atrial flutter. She was subsequently treated with flecainide, amiodarone, repeat electrical cardioversions, but eventually managed with oral diltiazem for rate control.

Throughout the rest of the pregnancy, she had serial echocardiograms that demonstrated right ventricular pressure consistently under 45 millimetres of mercury. No pulmonary vasodilator medications were required.

Her pregnancy was closely followed by the High Risk Pregnancy Clinic. Owing to the known increased maternal and foetal morbidity with congenital cardiac disease, the foetus was monitored with foetal well-being testing biophysical profiles and serial ultrasound evaluation for growth. At 37 weeks of gestation, the ultrasound foetal estimated weight of 2384 grams was at the 7th percentile – decreased from the 24th percentile 4 weeks earlier, suggestive of intrauterine growth restriction and foetal compromise. The decision was made for induction of labour with a planned passive second stage of labour with assisted vaginal delivery. The induction of labour was successfully accomplished with a low forceps-assisted vaginal delivery of a female infant weighing 2470 grams, Apgar score 9 at 1 minute and 9 at 5 minutes.

Before the delivery, Swan–Ganz catheter was placed for close haemodynamic monitoring. Pulmonary arterial pressure was in the range 35–40/20–25 millimetres of mercury. After delivery, the last reading of the pulmonary arterial pressure was 33/17 millimetres of mercury before the removal of the Swan–Ganz catheter.

On the second day after delivery, the radiofrequency ablation of atrial flutter was performed, with successful restoration of sinus rhythm. She is being followed up in our clinic, feeling well and taking care of the baby without difficulties.

Discussion

In this report, we are presenting a case of a young woman in the second trimester of pregnancy with previously undiagnosed large secundum atrial septal defect complicated by severe pulmonary hypertension with left-to-right shunt. Although management was challenging, we opted to perform an early open heart closure of the atrial septal defect without termination of pregnancy. Managing pulmonary hypertension in a pregnant patient is always challenging. We are unaware of other cases when such condition was successfully treated with open heart surgery.

Atrial septal defects, if untreated by surgical repair at an early age, can lead to pulmonary hypertension in about 35% of cases,[3] with subsequent right heart failure. The number of adult patients with congenital heart defects either repaired or unrepaired is increasing. Although timely surgical interventions increase the lifespan, some patients still present with unrepaired defects because of poor health care. In the recent review, 24% of women with congenital heart disease and pulmonary hypertension were diagnosed only during pregnancy.[4] In addition, as congenital heart disease survival rates have improved, the number of women surviving to childbearing age with complex congenital heart disease has increased.

During a normal pregnancy, several cardiovascular changes occur to maximise oxygen delivery to the foetus. Cardiac output increases up to 40% above baseline, with concurrent increases in both heart rate and plasma volume. As circulating blood volume increases, red cell mass increases correspondingly but at a lesser rate than the increase in plasma volume, producing a relative physiologic anaemia. In addition, there is a drop in both systemic and pulmonary vascular resistances, reducing the blood pressure during the first half of pregnancy.[5]

In patients with underlying pulmonary hypertension due to congenital heart defects with right-to-left shunts, and with increased pulmonary resistance, the above haemodynamic changes may not occur. The normal physiologic response to pregnancy of decreased systemic vascular resistance will promote right-to-left shunting of blood through the septal defects, thereby decreasing the amount of oxygenated blood circulating to patient and foetus. When this resistance is elevated during active labour, cardiac output demands are increased; however, with underlying cardiac disease, this cannot be adequately accomplished, causing increased cardiac strain and peripartum mortality due to cardiogenic shock.[6]

The data on evolution of pulmonary pressures during pregnancy are scarce. Despite decreased pulmonary vascular resistance, mean pulmonary artery pressure in healthy women remains grossly unchanged.[7] In primary pulmonary hypertension, pulmonary artery systolic pressure decreases somewhat during pregnancy but rebounds within hours of delivery.[8] In mitral stenosis, pulmonary artery systolic pressure increases by about 15 millimetres of mercury by mid-pregnancy.[9]

In atrial septal defects without right-to-left shunts, pulmonary arterial systolic pressure may not change from the baseline by mid-pregnancy, but the direction and the magnitude of change in right heart haemodynamics in late pregnancy when the cardiac output is high is difficult to predict. Increased cardiac output and blood flow during pregnancy may result in overestimation of pulmonary artery systolic pressure measured by velocity of tricuspid regurgitation. Nevertheless, pulmonary arterial systolic pressure was high in our patient by invasive haemodynamic studies as well. In addition, she was diagnosed with severe pulmonary hypertension before, with a recommendation to undergo a heart and lung transplantation, which indirectly speaks for severe pulmonary hypertension.

Although atrial septal defect as such is often well tolerated in pregnancy, severe pulmonary hypertension creates the highest risk during pregnancy, with high maternal mortality mostly in the first month after the delivery.[4]

Several options were considered for managing this patient.

Percutaneous repair of atrial septal defects has been successful in pregnant patients in the past,[10] but owing to the size of the atrial septal defect this approach was not feasible.

A second option was therapeutic pregnancy termination and then performing open heart surgery with atrial septal defect closure. The patient declined this option.

Another option was to try to decrease her pulmonary pressure medically till delivery, and to do an open atrial septal defect closure after delivery. The evidence on safety, tolerability, and efficacy of pulmonary hypertension drugs in pregnancy is

limited. A recent systematic review of cases of pulmonary arterial hypertension in pregnancy in the last two decades showed that prostacyclin analogues, phosphodiesterase inhibitors, and endothelin receptor blockers increase exercise capacity and functional class in patients with idiopathic pulmonary hypertension. Of the five patients who received sildenafil for pulmonary hypertension, four had congenital heart defects and all of them survived to 1 year.[4] There was a pattern of using inhaled nitric oxide and prostacyclin analogues as a last resort in patients who already were haemodynamically unstable, making it difficult to assess how well these medications might have worked if started at an earlier stage. A patient with atrial septal defect and Eisenmenger syndrome discovered in the third trimester of pregnancy was successfully managed with intravenous epoprostenol.[11] In another case, administration of inhaled nitric oxide during the labour of a woman with atrial septal defect and Eisenmenger syndrome resulted in improved oxygenation and pulmonary arterial pressure, but the patient died of worsening pulmonary hypertension and cardiac failure 21 days later.[12] None of the advanced pulmonary hypertension therapies were linked to increased maternal mortality risk.[4]

Finally, there was the option of doing the open heart surgery during pregnancy, either at the time of presentation or later during her pregnancy when the baby was viable and could be rescued in case of complications. Waiting until the third trimester has its own risks, including a greater cardiopulmonary demand from the pregnancy, as well as shifts in blood volume associated with delivery. Anaesthetic requirements for open heart surgery at this later stage also may cause an increase in pulmonary pressures with associated intubation and ventilation, possibly causing already delicate haemodynamics to have minimal room for error.

Cardiac surgery during pregnancy does not appear to increase the maternal mortality risk.[13,14] There is, however, a 10–15% risk of foetal mortality because of the non-pulsatile blood flow and hypotension associated with conventional cardiopulmonary bypass. In this case, although our patient had open heart surgery at 20 weeks of pregnancy, she later delivered a healthy girl.

Pulmonary hypertension after atrial septal defect closure

The common objection to this course of action is the fear that pulmonary artery systolic pressure may increase further after the atrial septal defect closure. The literature review indicates that this fear is not well justified.

Experience with 54 adult patients with moderate and severe pulmonary hypertension who had percutaneous closure of atrial septal defect showed that in most of them pulmonary pressures decrease, although not to the normal levels.[15] In 2 months after the intervention, right ventricular systolic pressure decreased from a mean of 57 to 51 millimetres of mercury ($p = 0.003$), and further decreased to 44 millimetres of mercury on the 30-month follow-up. In this series, 44% of patients eventually normalised the pulmonary artery systolic pressure – less than 40 millimetres of mercury, but 15% had persistent severe pulmonary hypertension.[16] In another series of 29 adult patients with secundum atrial septal defect and mean pre-procedure pulmonary systolic pressure of 65 millimetres of mercury (three patients had bidirectional shunt due to suprasystemic pulmonary pressure), it decreased to 54 millimetres of mercury after 21 months of follow-up, with clinical improvement in all symptomatic patients.[17]

Several cases of late (several years after surgery) mortality were reported by Horer et al. They found that pre-operative systolic pulmonary artery pressure greater than 36 millimetres of mercury, and mean pulmonary artery pressure greater than 21 millimetres of mercury were predictive of late death from arrhythmia or cardiac failure.[18] In 11 patients with mean pulmonary artery pressure greater than or equal to 30 millimetres of mercury before transcatheter atrial septal defect closure, two patients developed progressive pulmonary vascular disease with one death.[19]

We were ready to add sildenafil to our patient's therapy, but on serial echocardiograms post-atrial septal defect closure her pulmonary artery systolic pressure did not exceed 45–50 millimetres of mercury with further decrease after delivery.

Anaesthesia and delivery

For better outcomes, it is preferable to have a delivery in a tertiary care centre.[20–22] With regard to anaesthesia during delivery, greater preference has been given to regional anaesthesia (59%), but there were patients managed with general anaesthesia (31%). Patients treated with general anaesthesia had quadrupled maternal mortality risk, likely because of both cardiodepressive effects of anaesthetics and increase in pulmonary vascular resistance during positive end expiratory pressure.[4] It is also difficult to rule out the possibility that sicker patients had general anaesthesia.

Although vaginal delivery may be associated with smaller shifts in blood volume, fewer clotting or bleeding complications, and less infections, caesarean section is more frequently used (72% versus 28%).[4,11]

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During delivery, forceps assistance or vacuum extraction may be utilised to decrease stress of pushing.[20]

Compared with healthy women, pregnant patients with atrial septal defect – not necessarily with pulmonary hypertension – had higher risk of small for gestational age births and foetal mortality.[23] Foetal and neonatal death occurred in 7% of 29 pregnancies in women with congenital heart defects and pulmonary hypertension, and another 24% had intrauterine growth restriction.[4] Our patient's baby was also small.

Atrial flutter

Atrial flutter post-operatively is most likely secondary to scar formation in the myocardial wall. We initially selected flecainide, a IC antiarrhythmic with an excellent safety record in pregnancy for both maternal and foetal arrhythmias. When it failed to prevent further episodes of atrial flutter, we substituted amiodarone. It is considered a second-line drug for foetal arrhythmia and its use in pregnancy is not uncommon.[24]

Electrical cardioversion in pregnancy is infrequent. Although there is a theoretical risk of inducing foetal arrhythmia, the risk is low as only a small amount of energy is actually distributed to the foetus.[25] Tromp et al[26] reported two cases and collected 44 other cases from the literature. All 13 cases with known pregnancy outcomes had normal term deliveries. In two cases, there was foetal distress directly after the cardioversion, necessitating an immediate caesarean section at 37 and 28 weeks of gestational age, respectively.[25,27] Our patient had several electrical cardioversions without immediate or remote complication, and underwent successful radiofrequency ablation after the delivery.

Conclusions

Management of severe pulmonary hypertension in a pregnant woman with a congenital heart defect is always challenging. Our patient had a large unrepaired atrial septal defect with severe pulmonary hypertension. After carefully exploring all the options, we successfully treated her with open heart surgery with the closure of the defect. A substantial and consistent reduction in pulmonary arterial pressure after the surgery and subsequent uneventful delivery indicate that surgical repair of atrial septal defects is a viable option that should be considered for such patients.

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