Emergency aortic valve replacement and Caesarian section in a primigravida with severe aortic stenosis: a case report

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ABSTRACT

Introduction Congenital bicuspid aortic valve with severe aortic stenosis (AS) is a rare condition (3–6% of patients with congenital heart disease). Pregnancy in these patients carries a high risk of maternal and fetal mortality. With advancing gestational age, these women may develop cardiac failure due to increased cardiorespiratory requirements. When medical therapy proves insufficient, cardiac surgery becomes mandatory to save the patient’s life. Balloon valvuloplasty is only palliative treatment, the duration of benefit being only 6 months. Valve replacement is thus recommended. Cardiopulmonary bypass (CPB) surgery with valve replacement has been reported to carry a lower risk of maternal mortality (1.5–13%) but a very high fetal risk (16–40%). This paper reports the case of a 30-year-old primigravida with severe AS with bicuspid aortic valve and pulmonary congestion clinically uncontrolled, in whom CPB surgery and aortic valve replacement was performed as an emergency procedure, along with a lower segment Caesarian section.

Conclusion The outcome of unrelieved severe symptomatic AS in pregnancy is poor. Multidisciplinary management is important to avoid deterioration in cardiac performance in parturients with severe AS. CPB during pregnancy carries a high risk to the fetus. Therefore, open heart surgery during pregnancy should be advised only in extreme emergencies (ie, heart failure refractory to conventional therapy).

A 30-year-old primigravida woman with severe aortic stenosis (AS) presented at 28 weeks of gestation with a 2-week history of palpitations, precordial pain, orthopnoea and dyspnoea on exertion. She had no history of syncopal attacks and had recovered fully from a right-sided hemiplegia following a cerebrovascular accident 3 years earlier. There was no family history of heart disease, hypertension, or dyslipidaemia.

On general physical examination, she was aca- notic and not dyspnœic at rest. Her height was 1.54 m, weight 40 kg (body mass index 22.3 kg/m²), her pulse rate was 90/min regular and blood pressure 98/60 mm Hg. She had an ejection systolic murmur (5+/6+) in the aortic region radiating to the carotid arteries. There was equal air entry in both lung fields with no adventitious sounds. On abdominal examination, the uterus was 28 weeks’ size, and the fetus was in cephalic presentation with a heart rate of 140 bpm.

Electrocardiography revealed sinus rhythm and hypertrophy of the left chambers. Chest radiography showed cardiomegaly. Echocardiogram showed a bicuspid aortic valve, severe AS, mild left ventricular systolic dysfunction with ejection fraction of 45% and highly calcified aortic valve (figures 1–4). Haemogram, serum electrolytes, renal and liver function tests were within the normal range. Antistreptolysin-O titre was not raised. Abdominal ultrasound showed a single normal live fetus of 28 weeks’ gestation. She was started on oral furosemide 40 mg once a day and cardiology opinion was taken. Intramuscular corticosteroids (betamethasone 12 mg two doses 24 h apart) were given to enhance fetal lung maturation.

At 31 weeks of gestation, the patient developed progressive pulmonary congestion, congestive cardiac failure and hypotension. She required infusions of dopamine and dobutamine to maintain blood pressure at 80/60 mm Hg. Oxygen therapy (fractional inspired oxygen 0.5 at 10–12 l/min) was given by mask. Given that her clinical condition continued to deteriorate, a multidisciplinary team decision was reached to deliver the baby and manage her condition more aggressively including a cardiopulmonary bypass (CPB) surgery with aortic valve replacement.

Endotracheal intubation was performed after establishing an adequate anaesthetic depth and muscle relaxation. Arterial carbon dioxide tension was maintained at less than 40 mm Hg to avoid a possible increase in intracranial pressure associated with hypercapnia. Mean arterial pressure was maintained between 65 and 75 mm Hg. After the induction of general anaesthesia, the fetal heart rate dropped to 80 bpm and a female baby weighing 1500 g was born by Caesarean section with an Apgar scores of 5, 7 and 7 at 1, 5, and 10 min, respectively. The baby was resuscitated and transferred to the neonatal intensive care nursery. The patient’s abdomen was kept open to deal with any postpartum haemorrhage during the CPB as heparin was being injected during the valve replacement surgery. Prophylactic B-Lynch uterine compression sutures were placed to prevent uterine atony.

Following closure of the uterus, CPB was started with a pulsatile flow at a rate of approximately 2 l/min. The stenotic aortic valve was replaced with a mechanical valve (25 mm St Jude Medical aortic heart valve St. Jude Medical, St. Paul, MN, USA). CPB time was 145 min and aortic cross clamp time was 90 min. The patient was transfused 6 units of whole blood, 4 units of fresh frozen plasma and 4 units of platelet concentrate perioperatively.

Postoperatively, she was maintained on vaso-pressors for 48 h, and extubated on the third postoperative day. Warfarin was used for...
anticoagulation, maintaining an international normalised ratio of between 2.5 and 3.5. Abdominal sutures were removed on the tenth day postoperatively. Intravenous antibiotics were continued for 14 days after which she was discharged from the cardiac coronary care unit on warfarin, ecosprin and diuretics. She was transferred to the obstetric ward as the baby was still in the nursery. On the 17th day after surgery, she had an episode of generalised convulsions. Physician referral was taken and tab. phenytoin 500 mg HS was started. Contrast-enhanced CT (head) showed gliosis in the third ventricle. There was no fresh infarct or haemorrhage. Echocardiography showed no vegetation or clot. All other investigations were within normal limits.

The newborn infant had bronchopneumonia and patent ductus arteriosus but both the mother and baby were discharged home on the 25th postoperative day.

DISCUSSION
Heart disease is the most important cause of non-obstetric mortality in pregnancy. Among women of childbearing age, 2–4% are found to have concomitant heart disease. While congenital heart disease (CHD) is more common in the western world, acquired heart disease (rheumatic valvular heart disease) is more often found to affect women in Asia. Mitral stenosis is the most common lesion seen. Significant AS (acquired or congenital) is uncommon in this age group.

Severe AS with congenital bicuspid aortic valve is found in 3–6% of patients with CHD. The turbulent blood flow results in thickening, fibrosis, calcification and stiffness of the valve leading to clinical manifestations from the third decade of life. Earlier diagnosis of AS has been facilitated with the routine use of echocardiography.

Haemodynamic changes in pregnancy with AS
Pregnancy induces a 20–100% expansion of the blood volume, a 30–40% increase in cardiac output and an increase in heart rate of 10–20 bpm. Increase in cardiac output is limited in patients with severe AS because of obstruction of the left ventricular outflow. This may lead to haemodynamic decompensation (particularly if the valve area is <1 cm²), which frequently manifests as symptoms and signs of pulmonary congestion, syncope and sudden death. Therefore, these patients...
are extremely sensitive to small alterations in ventricular volumes, arterial pressure, cardiac frequency and rhythm. The outcome of unrelieved severe symptomatic AS in pregnancy is poor. In late pregnancy, compression of the inferior vena cava by the gravid uterus in the supine position decreases venous return and further reduces cardiac output in these patients. Finally, the hypercoagulable state associated with pregnancy can increase the risk of thrombosis in diseased or prosthetic valves.

Reported outcome
In a recent literature review, significant cardiac complications were seen in 11% of 2491 pregnancies among women with structural CHD. Hypertensive disorders and thromboembolic events were the only obstetric complications found to be more prevalent. Premature delivery rates ranged from 22% to 65%, and the neonates were small for gestational age.

However, pregnancy in patients with severe AS has been reported very infrequently, thus there is no consensus about the ideal management. In 1978, Arias and Pineda, analysing 38 gestations with 23 severe aortic stenoses, reported a maternal mortality rate of 17.4% for untreated patients and a fetal mortality rate of 34%. Attempted therapeutic abortion was associated with 40% maternal mortality. Other reported adverse outcomes include spontaneous abortion, small for dates, prematurity, low Apgar scores, neonatal hypoglycaemia, bradycardia, impaired response to anoxic stress and an increased frequency of CHD in the infant.

An invasive cardiac intervention may be necessary if the clinical condition deteriorates during pregnancy. The development of dyspnoea, syncope, angina and arrhythmias indicate critical left ventricular outflow obstruction. Surgical intervention (balloon valvuloplasty or valve replacement) must be considered in these cases.

Figure 3 Continuous wave Doppler of aortic valve suggestive of severe aortic stenosis.

Figure 4 Two-dimensional echo at the level of aortic valve.
However, balloon valvuloplasty has late results, with the duration of benefit lasting approximately 6 months, and is thus a palliative treatment only. The immediate appearance of aortic regurgitation or its progression and the later appearance of restenosis are the major complications of valvuloplasty. Other complications during the procedure include bleeding, arrhythmias, stroke, iliac-femoral arterial complications, injury to mitral valve and death. Exposure to x-rays during the procedure is another potential problem. Therefore, valve replacement is recommended.

CARDIOPULMONARY BYPASS

Effect on the mother
The effects of CPB are relatively well tolerated by the mother. Haemodilution, changes in coagulation, complement activation and hypotension during CPB may have a deleterious effect. However, in two studies on patients undergoing aortic valve replacement, no maternal death was reported, although the overall maternal surgical mortality rate with CPB has been reported to be 1.5%. With improvements in cardiac surgery and anaesthesia, the maternal risk of surgery is only marginally higher than the risk of CPB in a non-pregnant patient. The functional class of disease is the factor most commonly predicting a higher risk of maternal death. Other risk factors for maternal mortality during cardiac surgery include the use of vasoactive drugs, age, type of surgery and re-operation.

Effect on the fetus
During first two trimesters the incidence of teratogenesis is high. During the third trimester, with improvements in the outcome for premature infants with modern neonatal intensive care, delivery of the child immediately before commencing CPB is a safe option. If this is inappropriate, high-flow, high-pressure, normothermic bypass for as brief a period as possible should be used.

The initial response to CPB is fetal bradycardia, which can be corrected by increasing the perfusion rate. In most cases, the decrease in heart rate reverts to sinus rhythm on restoration of maternal circulation after cessation of CPB. This bradycardia is likely to be caused by reduced blood flow to the intervillous spaces resulting in fetal hypoxia and acidosis. The various causes postulated include maternal hypothermia, hypotension, haemodilution, reduced uterine perfusion pressure, fetoplacental dysfunction, uterine contractions and drugs that cross the placental barrier, such as β-adrenergic blockers. When CPB is prolonged, the fetus may develop sustained bradycardia.

Maternal core temperature also affects the fetal response during CPB. More severe hypothermia results in reduced perfusion and oxygen delivery to fetal organs. This induces a decline in fetal and placental function and increases the risk of fetal arrhythmias and cardiac arrest. Alterations in fetal heart rate may be observed even when maternal circulation, acid-base balance and perfusion pressure are stable. These may be related to the narcotic effect of anaesthetic drugs.

The overall fetal surgical mortality has been reported to be 16–33%, with no correlation to gestational age. Factors affecting fetal mortality are functional class, type of procedure, emergency surgery, re-operation, anoxic time and fetoplacental response to CPB.

Therefore, monitoring of the fetal heart rate is essential as it assists in the early recognition of the potentially deleterious effects of CPB, thus enabling timely intervention to minimise fetal mortality.

Effect on the uterus
Uterine contractions occur in response to bypass, possibly due to a dilutional effect from the stabilising influence of progesterone. Sustained uterine contractions reduce uterine blood flow and intervillus perfusion, resulting in fetoplacental insufficiency and fetal hypoxia. Uterine monitoring is essential to allow the early control of these contractions as they are associated with significant fetal loss.

Various techniques to modify this include the administration of progesterone, β 2-agonists and intravenous alcohol, all with some effect. The use of tocolytic therapy with ritodrine has been reported to prevent early labour, resulting in a favourable fetal outcome. However, more recent evidence suggests that there is no benefit. In fact, β-agonists increase myocardial work and oxygen demand in a circulation already compromised by the burden of pregnancy and the initiation of CPB.

Increased sustained uterine contractions are also associated with both the cooling and rewarming phases. This is supported by the fact that there is a notably higher fetal mortality rate with hypothermic than with normothermic CPB.

MANAGEMENT APPROACH

Women with severe AS require close monitoring by their obstetricians and cardiologists, especially during the third trimester, labour and delivery, and the early postpartum period. Good maternal and fetal outcome may be achieved through early preoperative detection of maternal cardiovascular decompensation, use of fetal monitoring, delivery of a viable fetus before CPB and scheduling surgery on an elective basis during the second trimester. Administration of maternal corticosteroids is recommended to initiate endothelial membrane stability and maturation of the fetal lungs, which can substantially improve fetal outcome. In addition, fetal morbidity may be reduced during CPB by optimising maternal oxygen-carrying capacity and uterine blood flow.

Current maternal bypass recommendations include:

1. Maintaining a high pump flow rate greater than 2.5 L/min/m² and perfusion pressure greater than 70 mm Hg (to maintain uterine blood flow);
2. Maintaining the haematocrit at greater than 28% (to optimise oxygen-carrying capacity);
3. Using normothermic perfusion when feasible;
4. Using pulsatile flow (may better preserve uterine blood flow); and
5. Using α-stat pH management (for maintenance of carbon dioxide homeostasis as hypocapnia causes uteroplacental vasocostriction).

The principles for the management of pregnant patients undergoing cardiac surgery with CPB include maintenance of maternal wellbeing, avoidance of teratogenic drugs, avoidance of intrauterine hypoxia and prevention of premature labour. The chief concerns during CPB are the control of temperature, perfusion pressure and nature of the bypass flow. Current evidence favours maintaining normothermic CPB, avoiding the use of vasoconstrictors (which may have a profound effect on the placental unit), and maintaining both high haematocrit and high flow rates.

PERIOPERATIVE MONITORING

Fetal monitoring
If the fetus is viable, intraoperative fetal monitoring is indicated, with a preoperatively agreed plan of action with regard to how a non-reassuring tracing will be managed. Persistent fetal
bradycardia should be treated aggressively by optimising maternal oxygen saturation, correcting any acid-base abnormalities, increasing CPB blood flow, minimising anaesthetic depth, replenishing fetal glycogen stores (avoiding maternal hypoglycaemia) and delivery by Caesarean section.\textsuperscript{26}

Uterine monitoring

Monitoring of uterine contractions can also be performed. If uterine contractions are detected, maternal intravascular volume can be increased and tocolytic treatment may be given.

Invasive haemodynamic monitoring

Severe AS is a known indication for pulmonary artery catheterisation in the parturient undergoing cardiac surgery; (other indications are New York Heart Association class 3 or 4 heart disease, cardiac failure, refractory pulmonary oedema and preeclampsia with refractory oliguria or pulmonary oedema).\textsuperscript{26}

USE OF ANTICOAGULANT IN CPB IN PREGNANT WOMEN

Oral anticoagulation with warfarin has the lowest incidence of maternal mortality and thromboembolism. However, in the first trimester, warfarin can cause fetal growth retardation, spontaneous abortion and embryopathy. In addition, warfarin is associated with premature birth and fetal and placental haemorrhage in the third trimester.\textsuperscript{27} For the purposes of CPB, heparin may be safely used in the usual doses for anticoagulation without risk to the fetus.\textsuperscript{28} Post-delivery, warfarin has been found to be safe in breast-feeding women (despite minimal secretion in breast milk), with no reported adverse neonatal effects.

OBSTETRIC MANAGEMENT DURING CPB

When the fetus is not viable, the mother’s condition takes priority. If indicated, cardiac surgery should be performed after informing the parents that the fetus may tolerate CPB poorly. In contrast, when the baby is viable, it may be in the best interests of the mother and baby to perform a Caesarean delivery before the cardiac operation, with a neonatologist available for resuscitation, because of the high fetal mortality associated with cardiac surgery. In such situations, delivery by Caesarean section using general anaesthesia is preferred, making sure the mother is controlled,\textsuperscript{29} maintaining adequate ventricular depth, replenishing fetal glycogen stores (avoiding maternal hypoglycaemia) and delivery by Caesarian section.\textsuperscript{27}


CONCLUSION

The outcome of unrelied severe symptomatic AS in pregnancy is poor. Multidisciplinary assessment and caref ul anaesthetic planning is important to avoid deterioration in cardiac performance in parturients with severe AS. Although the valve lesion can be corrected surgically before delivery at a low risk to the mother, CPB during pregnancy carries a high risk to the fetus. Therefore, open heart surgery during pregnancy should be advised only in extreme emergencies (ie, heart failure refractory to conventional therapy).

Competing interests None to declare.

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REFERENCES


