Case report - Congenital
Emergency redo mitral valve replacement and caesarean section in a patient with previous atrioventricular septal defect repair in childhood

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1. Introduction
Cardiac disease is seen in 1–2% of all pregnancies. Cardiac surgery has been reported to have a limited but defined role in the management of valvular heart diseases associated with pregnancy. Reported maternal and fetal mortality associated with cardiac surgery varies from 1.5% to 4.2% and 9.5–33%, respectively [1].

2. Case history
A 19-year-old woman was booked at 12 weeks’ gestation to the obstetricians at a district hospital. Her previous history included correction of atrioventricular septal defect at the age of two and mitral valve replacement at the age of six with a size 27 mm Medtronic Hall mechanical prosthesis. Comorbidities included systemic lupus erythematosus, previous left-sided hemiparesis and recent transient ischaemic attacks. Her anticoagulation was changed from warfarin to Enoxaprin at confirmation of pregnancy. She was managed by a multidisciplinary team of obstetrician, cardiologist, haematologist, paediatrician and rheumatologist.

Whilst under regular antenatal surveillance she became increasingly dyspnoeic from 24 weeks’ gestation with production of pink frothy sputum. She was admitted to the local hospital and subsequently transferred to our centre two weeks later. On admission she was in pulmonary oedema, which was treated with diuretics, antibiotics and steroids. Her condition seemed to improve. After two days her condition deteriorated as she developed cardiogenic shock. Her echocardiogram (Fig. 1) showed features of mitral prosthesis dysfunction with left ventricular inflow obstruction, pulmonary hypertension (pulmonary artery systolic pressure of 130 mmHg) and poor right ventricular function raising the suspicion of thrombus or pannus. She was then referred for emergency cardiac surgery. At this point the patient was clinically shocked; the blood pressure was 80 mmHg systolic with cold peripheries and metabolic acidosis of –13.

After a joint discussion with the concerned specialists a combined emergency caesarean section and redo mitral valve replacement (MVR) was planned.

She was taken to theatre where the left femoral vessels were cannulated under local anaesthesia following intravenous administration of 5000 U of heparin. General anaesthesia was induced followed by rapid caesarean section delivery of the baby by the obstetric team. After haemostasis was achieved and the caesarean section wound was closed satisfactorily, the patient was fully heparinized with a dose of 4 mg/kg. Cardiopulmonary bypass was established through femorofemoral route and the chest was opened. The heart was densely adherent and required careful dissection, which was followed by cannulation of the superior vena cava. Following aortic cross-clamping cardioplegia was delivered to achieve diastolic arrest and the patient was cooled to 32 °C. The left atrium was opened through a trans-septal approach. The minor orifice of the Medtronic Hall prosthesis was found to be completely obstructed by the pannus preventing the disc from opening. The mitral valve was replaced with a St Jude mechanical prosthesis.
After atrial closure and deairing of the heart, multiple attempts at weaning from the cardiopulmonary bypass (CPB) failed secondary to refractory right ventricular failure, despite incremental support with enoximone, epinephrine and nor-epinephrine. Following insertion of a right ventricular assist device (RVAD) contractility of the right heart improved and the patient was successfully weaned from bypass. One-hour post bypass right ventricular recovery was adequate to allow discontinuation of the RVAD and closure of the chest. Total bypass time was 300 min and cross-clamp time was 80 min.

In the immediate postoperative period vaginal bleeding was managed by balloon tamponade and administration of blood products. She developed severe inflammatory response syndrome (SIRS) in the early postoperative period requiring vasoconstrictor, inotropic and ventilatory supports which were weaned successfully over the next 72 h. The patient was discharged from the critical care unit to the ward after one week. Once in the ward she made steady progress and was discharged to her district hospital after two weeks. The baby required prolonged neonatal intensive care support but made a good recovery and was discharged home. Both mother and baby continue to do well at her recent clinic visit three months after surgery.

3. Discussion

With recent advances in screening, increasing numbers of patients with congenital cardiac disease are being detected during pregnancy. Although patients with good functional class tolerate pregnancy well, worsening of symptoms occur in 25% of the patients while premature delivery occurs in 12% of patients. Other complications such as bleeding, arrhythmia, endocarditis, cardiac failure, and death are known to occur in a significant proportion (11.3%) of patients [1].

The most common valvular problem in pregnancy is rheumatic valve disease accounting for 60–70% of cases [2]. Mitral valve is the most commonly affected valve. Percutaneous balloon valvuloplasty and closed mitral valvulotomies (CMV) are established procedures with low risk in pregnancy [3]. Where CMV is not feasible open mitral valvulotomies are reported with acceptable risks in this clinical setting. There are a number of reports of combined first time mitral valve operations with caesarean sections [3]. Mahli and colleagues have described the indications, risk factors, operative interventions and feto-maternal outcomes in this context [4].

Though symptomatic primary mitral valve disease in pregnancy is a relatively common presentation, malfunction of a mechanical mitral valve requiring surgical intervention in pregnancy is rare. Massad et al. have reported three cases of thrombosed Bjork-Shiley mitral valves requiring surgical de-clotting [5]. We could not find any report of a mechanical mitral valve dysfunction due to pannus in pregnancy requiring emergency redo mitral valve replacement and caesarean section.

Malfunction of mechanical valve is a surgical emergency. The risk of adverse fetal outcome diminishes progressively as the baby becomes more mature. Neonatal mortality is reported to be as high as 90% at 25 weeks of gestational age but decreases to <15% by 30 weeks [6]. Intervention before 30 weeks has to be balanced with increased maternal risk as pregnancy continues. A multidisciplinary team approach to management is essential. When surgery is indicated a short cardiopulmonary bypass time with high flow, high perfusion pressure is recommended to ensure fetal well-being [7]. Femoral access for the cardiopulmonary bypass under local anaesthesia was established before the start of general anaesthesia (GA) because of the possibility of cardiovascular collapse on induction of GA due to fixed output state. The patient was anesthetized only after the cannulae were in place in readiness to commence CPB. It was felt this strategy would allow CPB to be established immediately in the event of catastrophic cardiopulmonary collapse at induction of anaesthesia. The patient tolerated the anaesthesia for caesarian section well. Thus, full heparinisation for CPB was avoided until the caesarian section was completed; blood loss was thus minimized. CPB was established once LSCS was completed.

4. Conclusion

Emergency caesarean section with redo mitral valve replacement is a feasible option when indicated clinically.

References