1. Introduction

The simultaneous occurrence of acute type A aortic dissection and pregnancy is rare and commonly results in catastrophic outcomes unless emergent surgical intervention is undertaken [1]. Marfan syndrome is a distinct predisposing factor for aortic dissection, especially if the aortic root is already dilated. We report the case of successful aortic root and hemiarch replacement and cesarean delivery in a Marfan patient with twin pregnancy.

2. Case report

A 29-year-old female with known Marfan syndrome and pregnant at 34 weeks presented to the emergency room with severe chest pain, hypertension and increased painful uterine contractions. She had the classic stigmata of Marfan Syndrome. She was followed at the high risk obstetric clinic due to her aortic root dilatation (4 cm), however, with poor compliance. A CT-scan with IV contrast showed the thoracic aorta dissected with a diameter of 6 cm and extension of the dissection into the arch vessels and coronary sinuses.

Fetal viability and presence of twins was confirmed by ultrasound. She underwent fentanyl-based general anesthesia with invasive arterial and pulmonary artery monitoring. The intraoperative TEE showed severe AI and the intimal flap extending into the non-coronary sinus and right coronary ostium (Fig. 1). A cesarean section and twin males were delivered with low Apgar scores (1–7 and 2–8 at 1 and 5 min, respectively). This was most likely related to the use of fentanyl and maternal stress and pain. Babies’ weights were 2.18 kg and 1.79 kg. After the abdominal closure, a median sternotomy incision was performed and a large hematoma at the root of the aorta was found with impending rupture. Cardiopulmonary bypass (CPB) was instituted through the right femoral artery and right atrial cannulation using circulatory arrest (31 min) to complete the hemiarch anastomosis with velvety #28 mm Dacron graft (Vascutek, Terumo Cardiovascular Systems Corp., Ann Arbor, USA). Biologic glue (Bioglue, Cryolife, Inc., Kennesaw, USA) was used to reconstruct the layers of aorta before suturing. Circulatory arrest was started with a nasopharyngeal temperature of 17 °C with a flat EEG. Retrograde cerebral perfusion was implemented through an SVC cannula and monitored with transcranial Doppler and cerebral oximetry to confirm symmetric delivery of cold blood. The technique of cerebral protection is described elsewhere [2].

While rewarming, the coronary ostia patches were developed and reconstructed with glue, and a composite mechanical valved conduit (St Jude Medical Inc., Minneapolis, USA) #27 mm was implanted with the coronary ostia were attached individually. At a temperature of 36.5 °C the patient was weaned from CPB using a low dose of epinephrine (0.05 µg/kg/min). Myocardial arrest times were 208 and 170 min, respectively.

3. Discussion

Type A aortic dissection complicating pregnancy is a rare occurrence and life threatening for both the mother and...
the fetus, and requires immediate surgical intervention. The association of Marfan syndrome, pregnancy and aortic dissection is extremely rare. An English literature review [1] of dissection in pregnancies (n=16) showed the majority of pregnancies (1983–2002) in the third trimester patients with enlarged aortic roots (>4 cm) or increasing aortic size during pregnancy were at higher risk for dissection. Weiss et al. [3] analyzed 32 patients reported worldwide with aortic-arterial dissection (type not specified) and pregnancy. Maternal morbidity and mortality were 34% and 22%, respectively. It was this point on the higher risk for maternal death when surgery was performed late in pregnancy, most likely related to the hemodynamics burden that peak in the antepartum period [4]. The aortic repair in the mother has a reported mortality higher to the one in the non-pregnant patient. Overall reported fetal outcomes are relatively poor [5, 6], likely secondary to factors associated with delivery, fetal maturity and condition of the fetus in situ. The mechanisms underlying fetal distress include hypothermia, placental vasoconstriction and increased uterine contraction resulting in hypoxia and potentially fetal demise. Fetal outcome is superior in later pregnancy. At 28 weeks of gestation surgical strategy mandates cesarean delivery immediately followed by aortic repair. If the dissection occurs before the 28th week, fetal outcome is likely poor, but newer reports such as Sakaguchi et al. and others [7, 8] have successfully addressed the use of deep hypothermic circulatory arrest in early pregnancy with fetal viability in term. Matsuda et al. [9] reported a hemiarch aortic replacement at 19 weeks of pregnancy using normothermic selective cerebral perfusion and high flow pulsatile systemic perfusion resulting in good materno-fetal outcome.

Immer et al. [1] pointed out the importance of counseling in pregnant Marfan patients as well as in those with bicuspid aortic valves or enlarged aortas. In patients who are diagnosed with an enlarged aorta during pregnancy, a close echocardiographic follow-up (biweekly) is recommended. Development of the fetus should be closely monitored and fetal lung maturation at 26 weeks and/or early hospitalization in the third trimester should be considered, selectively.

To our knowledge, this is the first report in the English literature describing the occurrence of acute type A aortic dissection, Marfan syndrome and twin pregnancy. This report emphasizes the importance of counseling of all Marfan patients undergoing pregnancy.

References