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Pregnancy with Marfan's syndrome and outcome

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Objective

The Marfan syndrome is an autosomal dominant condition with an incidence of 1 in 3000 to 5000. Pregnancy and postpartum period is a high-risk time for aortic dissection and rupture due to increased arterial wall stress associated with the hypervolemic and hyperdynamic circulation and/or hormonal effects on aortic wall composition.

Methods

We had two women in one-year period presented in their first pregnancy. Case 1: Primigravida, Spontaneous conception. She had pectus excavatum, asymptomatic apart from palpitations not on any medicine. Being followed up in cardiology. Normal Ascending Aorta. History of atrial septum defect (ASD) and ventricle septum defect (VSD) repair at age of 5, Also ablation for supraventricular tachycardia (SVT) in Germany. The patient had emergency cesarian section (CS) for PPROM at 34+6 weeks and breech presentation. Birth weight 1. 7kg. The newborn has been admitted in neonatal intensive care unit (NICU) for 2 weeks with follow up appointment for ASD. Case 2: 35 years, primi, had Marfan syndrome complicated with type B aortic dissection in 2015. Aortic root replacement and re-implantation of coronaries with mitral valve ring and VSD repair in 2009 followed by infective endocarditis of mitral valve, underwent valve replacement in 2009. She was on Aldomet 500mg 3x/day and labetalol 200mg once daily for hypertension and was on warfarin. She had thoracolumbar scoliosis and was obese with BMI 44, after having abdominoplasty in 2004. Booked early in pregnancy and currently 28 weeks pregnant and asymptomatic. Elective CS at 36 weeks. Delivered alive baby which was kept on high dependency unit (HDU) until it became stable.

Results

These patients require specialized management prior to, during, after delivery. Mode of delivery should be least stressful; however, CS is not an absolute indication. The decision regarding the mode of delivery should be made after counseling. In our case both patients were fully counselled and opted for the pregnancy after understanding the risk for herself and her unborn child. Aorta should be evaluated by echocardiography every three months as aortic complications is increases if aortic diameter exceeds more than 10 percent (4. 0 cm). Woman contemplating pregnancy should have a screening transthoracic echocardiogram, CT or MR for assessment of aortic root, ascending aorta, and possible associated valve or myocardial disease. In addition, consultation with a cardiologist, maternal-fetal medicine specialist, and geneticist is recommended to inform the risks of pregnancy to her and her offspring.

Conclusion

Women with Marfan syndrome who are considering pregnancy should receive counselling regarding the risk of aortic dissection/rupture and aortic regurgitation. Discussion should include potential alternatives to pregnancy (adoption). The 50 percent chance of transmission from an affected parent to her child, phenotypic variability, and the availability of prenatal diagnostic testing should be discussed.