Pregnancy-Related Aortic Aneurysm and Dissection in Patients With Marfan's Syndrome: Medical and Surgical Management During Pregnancy and After Delivery

Mile Vranes[1, 2], Milos Velinovic[1, 2], Natasa Kovacevic-Kostic[1], Dragutin Savic[1, 2], Dejan Nikolic[3], Radmila Karan[1]

[1]Cardiac Surgery Clinic, Clinical Center of Serbia, Belgrade, [2]Faculty of Medicine, University of Belgrade, [3]University Children's Hospital, Belgrade, Serbia

Key words: *aortic aneurysm; aortic dissection; pregnancy; Marfan's syndrome; cesarean delivery.*

Summary. *In the current article, 3 cases of aortic aneurysm and dissection in pregnant patients with Marfan's syndrome are reported. It is well known that pregnancy is a risk factor for the development of aortic aneurysm and dissection in women with Marfan's syndrome since it is shown that dissection can develop both before and after labor. Marfan patients with an aortic root diameter greater than 4 cm should undergo preconceptual counseling for surgical aortic repair before pregnancy. Pregnant Marfan patients with an aortic aneurysm should be closely and continuously evaluated by multidisciplinary specialists in order to prevent possible aortic dissection that could be fatal for both the mother and the fetus.*

Introduction

Aortic dissection (AD) is a serious condition with possible lethal outcome (1). Aortic dissection carries a high risk of maternal mortality and, if prepartum, fetal mortality as well (2). One of the important predisposing factors for AD in pregnancy is Marfan's syndrome due to decreased elasticity of the aortic wall (2). Physiological changes in pregnancy accelerate the development of pathological changes, such as hormonally mediated decrease in the amount of mucopolysaccharides and loss of elastic fibers in the aortic wall (3). The risk of AD or aortic rupture increases with gestational age due to increased cardiovascular stress (2). Further, the diameter of the aortic root greater than 4 cm is also considered a great risk for dissection as well as rapid aortic dilatation, especially in Marfan's syndrome (4). Concerning the facts mentioned above, the current study demonstrates our experience in diagnosing and treating an aortic aneurysm and dissection in pregnant patients with Marfan's syndrome.

Case Report 1

A 30-year-old patient with Marfan's syndrome showing prepregnancy echocardiographic findings of the aortic root 3.6 cm in diameter and the aortic valve of normal tricuspid configuration without regurgitation was presented to our clinic at the 26th gestational week with severe chest pain that began 3 hours before admission. The woman was hemodynamically unstable with blood pressure (BP) of 80/40 mm Hg. Transthoracic echocardiography showed a presence of intimal flap 3 cm above the right coronary artery ostium.

After the emergency sternotomy and evacuation of hematopericardium had been performed, cardiopulmonary bypass (CPB) was initiated using femoral right atrial cannulation. The ascending aorta was transected and replaced by a 26-mm Dacron graft using modified "sandwich" technique without circulatory arrest (Fig. 1). Perfusion pressure was maintained at maximal values. Following the surgery, the patient had been receiving assisted ventilation for 10 hours and was hemodynamically stable with BP of 110/60 mm Hg, mean heart rate of 74.23±5.72 beats per minute, and mean SO_2 of 98.42%±1.02%.

Ultrasound examination showed a viable fetus with regular fetal heartbeat and expected movements of the upper and lower limbs. The patient was discharged 10 days after admission with stable vital signs. At the 34th week of pregnancy, the patient underwent an elective cesarean delivery of a healthy baby weighing 3400 g with an Apgar score of 9 at 1 and 5 minutes. The score of 9 was given because of cesarean delivery due to the effects of anesthesia on the newborn's skin color.

Case Report 2

In another case, a 41-year-old Marfan patient was diagnosed with an aortic root aneurysm in her first trimester. She had been closely cardiologically and echocardiographically monitored. Echocardiography performed before pregnancy revealed the aortic root 3.8 cm in diameter and the aortic valve of normal tricuspid configuration with mild (grade 1+) regurgitation present. By cesarean delivery at the 36th gestational week, the patient gave birth to a healthy

Correspondence to R. Karan, Palmoticeva 23, 11000 Belgrade, Serbia. E-mail: karan.radmila@gmail.com

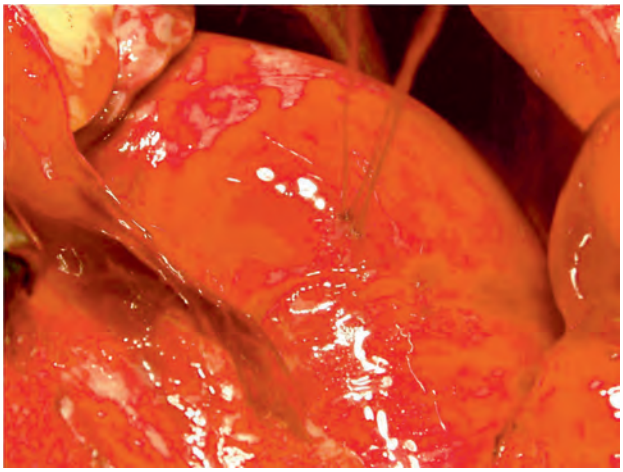

Fig. 1. Aortic dissection in a pregnant patient with Marfan's syndrome

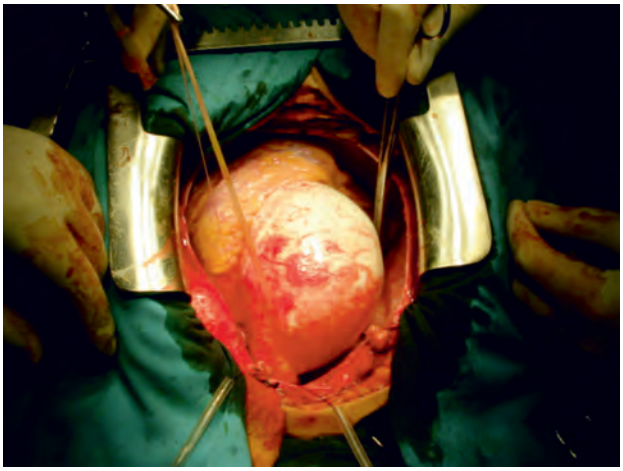

Fig. 2. Aortic aneurysm of a pregnant patient with Marfan's syndrome

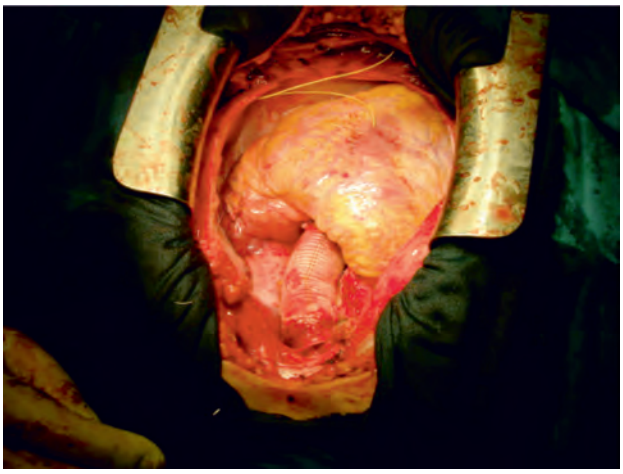

Fig. 3. The Bentall procedure replacing the aortic root, the ascending aorta, and the aortic valve

male newborn weighing 3500 g. Four months after the delivery, she was admitted to our clinic because of type I aortic dissection. Two days before admission, she complained of a sudden onset of chest pain and shortness of breath. Her BP was 140/75 mm Hg, and HR was 100 beats per minute. Transthoracic echocardiography revealed the aortic root 4 cm in diameter and a dissection located 1.5 cm above the aortic root. Cardiopulmonary bypass was initiated using right subclavian artery cannulation. The ascending aorta was replaced by a 26-mm Dacron graft. The postoperative course was uneventful. The patient was discharged 12 days after the operation.

Case Report 3

A 25-year-old patient with Marfan's syndrome from a remote rural area was admitted to our clinic for a surgical repair of an aortic aneurysm, diagnosed during her pregnancy, without any echocardiographic findings before pregnancy.

Three and a half months earlier, at the 38th week of gestation, the patient had given birth to a healthy male newborn weighing 3200 grams with an Apgar score of 9/10 at 1 and 5 minutes by cesarean delivery under general anesthesia. The postoperative course was uneventful, and the patient was scheduled for surgical aortic repair. Transthoracic echocardiography showed the aortic root of 56 mm and the ascending aorta of 81 mm in diameter (Fig. 2). The Bentall procedure was performed replacing the aortic root, the ascending aorta, and the aortic valve with reimplantation of the coronary ostia (Fig. 3). Cardiopulmonary bypass was initiated using femoral right atrial cannulation. An operation was performed with a brief period of circulatory arrest in deep hypothermia (13 minutes, 17°C). The patient was extubated after 9 hours and was discharged 8 days after the operation.

Discussion

In this paper, our experience in the treatment of pregnant patients with Marfan's syndrome who were diagnosed with either aortic dilatation or dissection is presented as a possible contribution to better understanding of such pathology. Aortic dissection associated with pregnancy is a rare condition, which is usually fatal for the mother and, especially, for the fetus (1). It is associated with the hyperdynamic and circulatory changes that occur in pregnancy, such as an increase in heart rate, stroke volume, cardiac output, and in end-diastolic dimensions subsequently (5). These changes are most prominent in the third trimester, and most dissections occur during this period of pregnancy (5). The gravid uterus causes compression of the aorta and iliac arteries, and this possibly increases outflow resistance to blood ves-

sels. In addition, an increase in cardiac blood ejection into the upper aorta may initiate an intimal tear causing AD (6). Fragmentation of reticulin fibers with a decrease in the amount of acid mucopolysaccharides as well as loss of normal corrugation of the elastic fibers is associated with hormonal changes that occur in pregnancy (7).

Marfan's syndrome is an important predisposing factor for the development of AD in pregnant women. Our experience with these 3 patients shows that pregnancy is a risk factor in women with Marfan's syndrome for the development of AD since it is shown that dissection can develop both before and after labor. According to previous findings, female Marfan patients with a normal aortic root diameter have a 1% chance for AD during pregnancy (8). These patients have a connective tissue disorder that increases the predisposition to dilatation and dissection of the aortic root (9). Besides, according to one study, there is an additional aortic wall stress that may favor dissection during pregnancy in patients with Marfan's syndrome (9).

Recent recommendations for the management of aortic diseases during pregnancy in patients with Marfan's syndrome point out that the aortic root diameter greater than 4 cm or an increase in the aortic root diameter during pregnancy is considered as a risk factor for AD in these patients; meanwhile, possible pregnancy should be discouraged in case of the aortic root diameter greater than 4.5 cm (8). In pregnant Marfan patients with a diameter of the ascending aorta greater than 4.5 cm, prepregnancy surgery is recommended (8).

Lind and Wallenbrurg pointed out in their study that less than 25% of patients with Marfan's syndrome underwent prepregnancy counseling (10). Three patients described in the current study did not undergo counseling before pregnancy. All pregnant Marfan patients who are diagnosed with an aortic enlargement should undergo regular echocardiographic monitoring at 4–12-week interval as well as monitoring of fetal development (8).

If AD occurs before 30 weeks of gestation, immediate aortic root repair is indicated; meanwhile, if it happens after 30 weeks of gestation, close monitored cesarean delivery followed by cardiac surgery is the recommended modality of treatment (1). During 6 months after delivery, regular cardiologic and echocardiographic follow-up is mandatory, because late dissections may occur up to 3 months after delivery (1, 8), as described in the case report of one of our patients.

In the case of the first patient, AD occurred at the 26th week of gestation, so immediate AD surgery was necessary. The surgery was done successfully with high-flow, high-pressure perfusion without circulatory arrest, which decreased a risk of fetal mortality or fetal physiological damages.

Conclusions

The presentation of these 3 cases underscores the importance of continuous and thorough medical evaluation of pregnant patients with Marfan's syndrome by a multidisciplinary team leading to an adequate and timely surgical procedure to prevent possible aortic dissection that could be fatal for both the mother and the fetus.

Statement of Conflicts of Interest

The authors state no conflicts of interest.

References

1. Sakalauskas J, Kinduris S, Benetis R, Giedraitis S, Jakuska P, Tamošiūnas V, et al. Surgical treatment of acute type A aortic dissection. Medicina (Kaunas) 2009;45:192-6.
2. Immer FF, Bansi AG, Immer-Bansi AS, McDougall J, Zehr KJ, Schaff HV, et al. Aortic dissection in pregnancy: analysis of risk factors and outcome. Ann Thorac Surg 2003;76: 309-14.
3. Goland S, Elkayam U. Cardiovascular problems in pregnant women with marfan syndrome. Circulation 2009;119;619-23.
4. Shores J, Berger KR, Murphy EA, Pyeritz RE. Progression of aortic root dilatation and the benefit of long-term beta-adrenergic blockade in Marfan's syndrome. N Engl J Med 1994;330:1384-5.
5. Barrett JM, Van Hooydonk JE, Boehm FH. Pregnancy-related rupture of arterial aneurysms. Obstet Gynecol Surv 1982;37:577-66.
6. Wheat MW. Intensive drug therapy. In: Dorghazi S, editor. Aortic dissection. New York, NY: McGraw-Hill; 1983. p. 55-60.
7. Manallo-Estrella P, Barker AE. Histopathologic findings in human aortic media associated with pregnancy. Arch Path 1967;83:336-41.
8. Regitz-Zagrosek V, Blomstrom Lundqvist C, Borghi C, Cifkova R, Ferreira R, Foidart JM, et al. ESC Guidelines on the management of cardiovascular diseases during pregnancy: The Task Force on the Management of Cardiovascular Diseases during Pregnancy of the European Society of Cardiology (ESC). Eur Heart J 2011;32(24):3147-97.
9. Dietz HC, Pyeritz RE, Hall BD, Cadle RG, Hamosh A, Schwartz J, et al. The Marfan syndrome locus: confirmation of assignment to chromosome 15 and identification of tightly linked markers at 15q15-q21.3. Genomics 1991; 9:355-61.
10. Lind J, Wallenburg HC. The Marfan syndrome and pregnancy: a retrospective study in a Dutch population. Eur J Obstet Gynecol Reprod Biol 2001;98:28-35.

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