

CASE REPORT

Management of Pregnant Woman with Marfan Syndrome and Mechanical Aortic Valved Conduit

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Abstract

We report a case of a 29-year-old patient with Marfan Syndrome and a mechanical aortic valved conduit graft that was referred to cardiologist at seven weeks pregnant. Echocardiogram: aortic root diameter 35 mm, normally functioning mechanical aortic valve graft. She had a specialized cardiologist follow up. The patient was submitted to cesarean at 38 weeks pregnant, with general anesthesia, antibiotic prophylaxis for endocarditis, cardiac monitoring on the surgical center and stayed in the Coronary Care Unit for 24 hours. Pregnancy and puerperium evolved without hemorrhagic events. The clinical and obstetric follow up of pregnant women with Marfan Syndrome is challenging, and the condition of the previously implanted mechanical heart valve enhances the risk for those patients.

Introduction

Marfan Syndrome (MS) is an autosomal dominant disorder with prevalence of 1:10.0000 live births. There is no ethnic, geographic or gender predilection, and the family history of the MS is present in 49% of the affected patients. It is caused by mutations to the fibrillin-1 gene, an important component of the connective tissue. The main alterations of the MS occur in the musculoskeletal, ocular and cardiovascular systems.¹⁻²

Keywords

Marfan Syndrome; Aortic Aneurysm; Thoracic; Pregnancy; High-Risk; Heart Valve Prosthesis Implantation.

Maternal risk increases due to the physiological changes of pregnancy and has a high mortality rate in childbirth, making it a challenge for obstetricians and cardiologists.³

The main cause of death in the MS is the acute aortic insufficiency and aortic dissection. This important finding reinforces the need for preventive measures during pregnancy.³⁻⁶

We report a challenging case of a pregnant woman carrier of the MS, in late postoperative period with ascending aortic aneurysm repair with implantation of aortic valved graft with aortic mechanical prosthesis.

Case Report

T.M.F.B, 29 years old, diagnosis of MS in childhood, with positive family history: mother and sister also carry the disease.

She reports a history of aortic insufficiency and ascending aorta aneurism (Stanford A). In 2011, she was submitted to surgery with implantation of valved graft with aortic mechanical prosthesis.

Pregnant in April 2014, she began prenatal care during the first trimester, and was referred to the cardiology outpatient clinic with seven weeks of pregnancy using oral anticoagulant (warfarin). During the physical examination, she showed atypical facies, arachnodactyly (Figure 1), and bone deformities in the ankles, feet and elbows. Valve prosthetic noise present with systolic murmur in the aortic area ++/6+.

Electrocardiogram showed sinus rhythm and left atrial overload. Echodopplercardiogram: diameter of the aortic root of 35 mm, normofunctioning valved graft and metal aortic prosthesis with peak gradient of the outflow tract

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of the left ventricle of 20 mmHg, mitral valve prolapse with mild/moderate impairment and left ventricular ejection fraction of 66% (Figure 2).

The patient was kept in full anticoagulation with enoxaparin in the first trimester and with warfarin in the second trimester, along with weekly monitoring by the

INR (International Normalized Ratio) and maintenance in the therapeutic range (2.0 to 3.0). Enoxaparin was reintroduced in the 35th week of pregnancy and suspended 12 hours before parturition. Propranolol was prescribed in the 28th week and maintained until the puerperium for control of heart rate following complaints of palpitations.



Figure 1
Arachnodactyly.

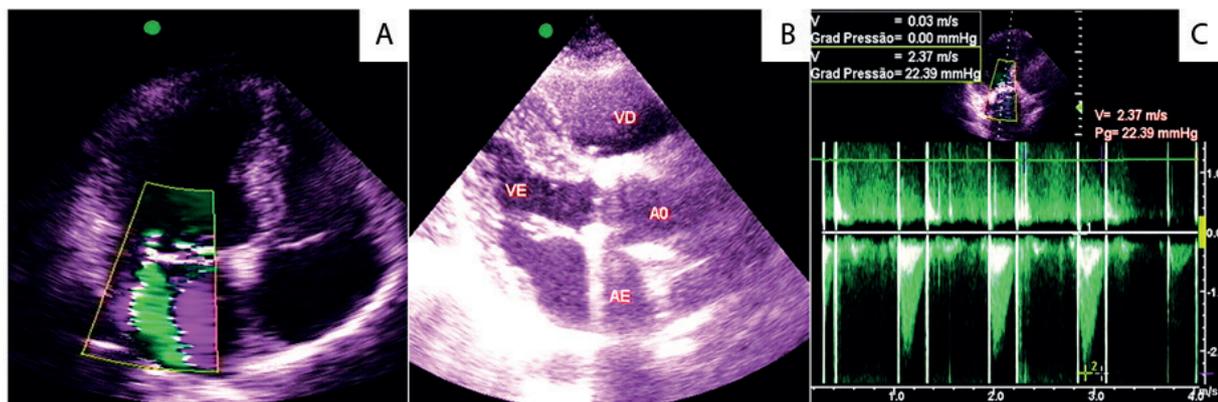


Figure 2
Echodopplercardiogram showing mild/moderate mitral valve insufficiency (a), aortic valve conduit (b) and residual gradient of outflow tract of the left ventricle after implantation of mechanical prosthetic valve in the aortic position (valved graft) (c).

In the 38th week of pregnancy, elective caesarean delivery with general anesthesia was performed. Prophylaxis for bacterial endocarditis with ampicillin and gentamicin was conducted an hour before the procedure, with a new dose of ampicillin 6 hours after the procedure. Cardiac monitoring in the surgical center and postoperative period in the coronary unit were maintained for 24 hours. Pregnancy and puerperal period went by without hemorrhagic events, and warfarin was reintroduced on the first day following childbirth.

Three months after the birth, the infant remains asymptomatic, without hemorrhagic events, need for unscheduled surgical interventions or major adverse cardiac events. Maintained in clinical monitoring, oral anticoagulation and clinical follow up with a cardiologist.

Discussion

The MS and the prior complications displayed by the patient grant high risks to the pregnancy and childbirth, and it is not uncommon for the woman to be discouraged from getting pregnant. The risk of hemorrhagic events due to anticoagulation, linked to the risk of thrombotic events involving the mechanical valve prosthesis require an rigorous and extremely cautious monitoring from the pregnant woman and from the multidisciplinary medical team.

Guidelines by the Brazilian Society of Cardiology on heart diseases and pregnancy classifies the patient of our report as a case of high-risk and with contraindications to pregnancy, citing therapeutic abortion as a conduct to consider.⁷ According to guidelines by the European Society of Cardiology on the same theme, the patient at hand would be deemed to be of high-risk; however, in this case, it suggests that the conduct must be individualized, and specialized monitoring is an alternative.⁸

It is noticed that the classification of maternal risk varies among the references and even among services from the same country. The conduct adopted in our case, in consonance with the patient's wish, was to keep the pregnancy and perform specific cardiological and obstetrical monitoring.

It is important to stress that the patient was referred with the pregnancy in progress, without previous planning, an unfortunately common fact in our reality, which made it impossible to carry out genetic counseling and other clarifications prior to the conception.

The performance of cesarean delivery under general anesthesia was indicated aiming to decrease the risk of aortic rupture during labor, although the pregnant woman had previously had surgery on her aorta and aortic valve with implantation of metallic aortic valved graft. Possible points of fragility and aortic rupture could exist, due to the weakening of connective tissue caused by the MS.⁸

Propranolol was used during the pregnancy up to the puerperal period. The use of beta-blockers in patients with MS is recommended during pregnancy, birth and puerperal period to prevent aortic dissection,⁷ a complication that may occur suddenly even without large aortic dilatation. Oral anticoagulation was suspended during the first trimester of the pregnancy following the diagnosis of pregnancy due to the risk of development of warfarin embryopathy in this period.⁹

In a recent observational study, it was proven that only 58% of the pregnancies of mechanical heart valve prostheses have no type of complication, showing the dimension of the challenge in conducting these patients.¹⁰ In the reported case, the improbable favorable and uncomplicated evolution was due to the early perception of the high-risk pregnancy and rigorous perinatal clinical accompaniment.

This case illustrates a serious and challenging situation of MS and pregnancy, in which the prenatal care, cardiological monitoring and advice and the presence of the multidisciplinary team were also fundamental to prevent maternal and fetal morbidity and mortality.

Author contributions

Conception and design of the research: Mario ADS, Calil OA, Jacques TM, Serpa RG, Barbosa LFM. Acquisition of data: Mario ADS, Calil OA, Jacques TM. Analysis and interpretation of the data: Barbosa RR, Mario ADS, Calil OA, Serpa RG, Barbosa LFM. Writing of the manuscript: Barbosa RR, Mario ADS. Critical revision of the manuscript for intellectual content: Barbosa RR, Barbosa LFM.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Study Association

This study is not associated with any thesis or dissertation work.

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