

Pregnant Marfan Syndrome Patients and the Risk of Aortic Dissection

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Objective:

Pregnant Marfan Syndrome (MS) patients face an increased risk of aortic dissection. A dilated aortic root (≥ 40 -45 mm) is considered a relative contraindication for pregnancy. We investigated pregnancy outcome in patients with MS based on aortic root diameter.

Study Design:

Women with MS attending our cardiology high risk pregnancy clinic between 2006-2012 were prospectively followed clinically and with serial echocardiograms by a multidisciplinary MFM-cardiothoracic team. Patients with aortic root dilatation ≥ 40 mm were considered at high risk, while patients with normal aortic root were considered at lower risk.

Results:

Nineteen patients with MS were followed. 6/19 patients had a dilated aortic root (41-46 mm); 5 were managed as inpatients during the third trimester, one is still ongoing. Beta-blockade was offered to all, and taken by 5/6. The one patient without beta-blockers presented with chest pains at 36+6 weeks without echocardiographic changes, an urgent cesarean section was performed, and ascending aortic dissection was diagnosed postpartum. Immediate surgical repair with good results ensued. 3 additional high-risk patients underwent elective cesarean sections at 34-35 weeks following fetal lung maturity confirmation, and one, with an aortic root of 46 mm, had an urgent cesarean section due to preterm labor. 13/19 patients had aortic roots of 27-39 mm and were mostly managed as outpatients. All remained clinically and echocardiographically stable. Most (10/13) delivered vaginally. There was a 31% preterm delivery rate with one antenatal fetal death at 26 weeks. One pregnancy is still ongoing. 1/13 low risk Marfan women was diagnosed postpartum with descending (type B) aortic dissection was diagnosed. The overall dissection rate was 10.5%.

Conclusions:

Pregnant MS patients, especially those with dilated aortic roots, are at a significantly increased risk of aortic dissection in the 3rd trimester and peripartum. Tight control of pregnancy-associated tachycardia and blood pressure by beta blockers may be beneficial in alleviating this risk.