

Peter Wenaweser
Swiss Cardiovascular Center Bern
Freiburgstrasse Bern
Switzerland

Otto M. Hess
Swiss Cardiovascular Center Bern
Freiburgstrasse Bern
Switzerland

Stephan Windecker
Swiss Cardiovascular Center Bern
Freiburgstrasse Bern
Switzerland

Tel: +41 31 6324497
Fax: +41 31 6324770
E-mail address: stephan.windecker@insel.ch

References


Alfred Hager
Klinik für Kinderkardiologie und angeborene Herzfehler
Deutsches Herzzentrum München
Technische Universität München
Lazarettstr. 36
D-80636 München
Germany
Tel: +49 89 1218 1650
Fax: +49 89 1218 3013
E-mail address: a-hager@web.de

Harald Kaemmerer
Klinik für Kinderkardiologie und angeborene Herzfehler
Deutsches Herzzentrum München
Klinik an der Technischen Universität München
München
Germany

John Hess
Klinik für Kinderkardiologie und angeborene Herzfehler
Deutsches Herzzentrum München
Klinik an der Technischen Universität München
München
Germany

doi:10.1093/eurheartj/ehi547

Online publish-ahead-of-print 23 September 2005

Comment on pregnancy and aortic root growth in the Marfan syndrome: reply

Thank you for the opportunity to respond to the comment from Hager and co-workers. It is an immense tragedy when a young woman dies during pregnancy. We agree with Hager and co-workers that there is no definite safe aortic root diameter for women with Marfan syndrome to get pregnant. Dissections may occur at normal aortic diameters in patients with Marfan syndrome. Should we therefore advise all women with Marfan syndrome against pregnancy? During recent years, a panel of international experts has reached consensus that pregnancy can be tolerated in women with Marfan syndrome with a slightly dilated aortic root. This expert consensus is being validated by our findings, which indicate that pregnancy is relatively safe in women with Marfan syndrome and an aortic root diameter up to 45 mm. However, women with a previous dissection should not get pregnant. We agree with Hager et al. that all patients with Marfan syndrome deserve close and careful monitoring before, during, and after pregnancy. Before pregnancy, all women should undergo a magnetic resonance angiogram to investigate if there is dilatation in other parts of the aorta. Also, frequent echocardiographic imaging should be performed throughout pregnancy and the postpartum period to check for progressive aortic dilatation. In the future other risk factors for aortic dissection, such as aortic elasticity, might become available to


6. Wenaweser et al. performed an analysis to determine the case number necessary to find any differences was not performed. The authors even reported on one woman with an aortic dissection during pregnancy and on an increased growth of the aortic root during long-term follow-up in those patients with an aortic root diameter 40 mm at baseline in a subgroup analysis.

We recently lost one of our patients, a 36-year-old woman with aortic coarctation and bicuspid aortic valve. These patients usually have structural abnormalities in the aortic medial wall predisposing to dilatation, aneurysm, and rupture, which are similar but less pronounced than those described in Marfan syndrome.

This woman died from aortic rupture at the 36th week of her second pregnancy. Unfortunately, the patient was not seen in our centre during pregnancy, and no consecutive imaging was performed.

Summarizing, this study did not provide real evidence for the conclusion that pregnancy in women with Marfan syndrome seems to be relatively safe up to an aortic root diameter of 45 mm. We should recommend to monitor all pregnant women with Marfan syndrome very carefully and closely, as suggested in many previous studies, because aortic dissection does not only depend on aortic diameter progression and may also occur in Marfan patients with a normal aortic diameter.

We believe that it is too early to draw such a conclusion. All statistical tests performed in this study were aimed to find any growth. These tests failed, but a power analysis to determine the case number necessary to find any differences was not performed. The authors even reported on one woman with an aortic dissection during pregnancy and on an increased growth of the aortic root during long-term follow-up in those patients with an aortic root diameter >40 mm at baseline in a subgroup analysis.

We recently lost one of our patients, a 36-year-old woman with aortic coarctation and bicuspid aortic valve. These patients usually have structural abnormalities in the aortic medial wall predisposing to dilatation, aneurysm, and rupture, which are similar but less pronounced than those described in Marfan syndrome.

This woman died from aortic rupture at the 36th week of her second pregnancy. Unfortunately, the patient was not seen in our centre during pregnancy, and no consecutive imaging was performed.

Summarizing, this study did not provide real evidence for the conclusion that pregnancy in women with Marfan syndrome seems to be relatively safe up to an aortic root diameter of 45 mm. We should recommend to monitor all pregnant women with Marfan syndrome very carefully and closely, as suggested in many previous studies, because aortic dissection does not only depend on aortic diameter progression and may also occur in Marfan patients with a normal aortic diameter.