



Acute Type A Dissection during Pregnancy with Marfan's Syndrome

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Thorac Cardiovasc Surg Rep 2021;10:e18–e21.

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Abstract

Background Marfan's syndrome (MFS) is a connective tissue disorder, caused by a mutation in the *FBN-1* gene and affecting multiple organ systems including the vascular system. During pregnancy, women with MFS have an increased risk of aortic dissection due to changes in the cardiovascular system.

Case Description We present a 39-year-old woman that suffered from an acute aortic dissection in week 33 of her third pregnancy who urgently required lifesaving surgery. Three-dimensional reconstruction of computed tomography scan was performed with unborn child in utero.

Conclusion Monitoring of patients with suspected or confirmed MFS appears essential and interdisciplinary treatment and prevention strategies are required.

Keywords

- ▶ aortic dissection
- ▶ Marfan
- ▶ pregnancy

Introduction

Marfan's syndrome (MFS) is an autosomal dominant disorder caused by mutations in the *FBN-1* gene, encoding for fibrillin 1, a glycoprotein which is a major component of the extracellular matrix.¹ Manifestations of this disease can be variable, but the musculoskeletal, the ocular, and the cardiovascular systems are most commonly affected in patients with MFS.² There is also evidence that the mutation type in the *FBN-1* gene is associated with the degree of severity of cardiovascular manifestations.³ Aortic complications such as dissections occur in more than one-third of MFS patients.⁴

In the state of pregnancy, women experience changes in the cardiovascular system. Hypervolemia due to an increased blood volume leads to cardiac hypertrophy which is considered a physiological change in pregnant women.⁵ Additionally, an increase in steroid hormone levels may lead to a dilation of the aorta, due to a decline of elastic fibers and mucopolysaccharides in the tunica media of the aorta⁶ and

pregnant women with MFS are especially affected by these changes. Despite the previously described correlation between pregnancy and acute aortic dissection in patients with MFS,^{7,8} the implementation of guidelines for the treatment of affected women⁹ in terms of prevention, surveillance, and treatment strategies are often inadequately. Therefore, we want to present a dramatic case of a 39-year-old woman, who suffered from acute type A aortic dissection in week 33 of her third pregnancy.

Case Presentation

A 39-year-old woman with previous diagnosis of MFS was admitted to an external hospital with acute chest pain and shortness of breath in week 33 of her third pregnancy. After computed tomography was performed to confirm the suspected diagnosis of acute type A dissection, the patient was transferred to our university cardiac surgery department. At the same time, the obstetricians unit of our hospital was informed for emergency cesarean section. When the patient

received
November 5, 2020
accepted after revision
November 24, 2020

DOI <https://doi.org/10.1055/s-0040-1722705>.
ISSN 2194-7635.

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Fig. 1 Computed tomography scan confirming suspected aortic dissection with unborn child in utero. The spiraled dissection reached from the aortic valve annulus to the iliac bifurcation.

was prepared for surgery in the operating room, ventricular fibrillation occurred and cardiopulmonary resuscitation (CPR) was performed. After computed tomography was performed to confirm the suspected diagnosis of acute type A dissection (► **Figs. 1** and **2**).

Under continued CPR, median sternotomy was performed to provide access to the heart. Cannulation of the ascending aorta and the right atrium was used for the establishment of cardiopulmonary bypass (CPB) in systemic hypothermia. Subsequently, the proximal part of the dissected brachiocephalic artery was cannulated to provide brain perfusion. When cardioplegia was applied directly through the coronary ostia, an additional rupture of the right coronary artery was noticed. The ascending aorta, the aortic valve, and the left coronary button were excised and a mechanical valve conduit was implanted. Following this, aortocoronary venous bypass was performed to the right coronary artery. Selective antegrade cerebral perfusion was performed to examine the extend of the dissection regarding the aortic arch. After clamping of the brachiocephalic trunk, neuromonitoring showed that cerebral oxygen saturation remained on a stable level. After removing of the aortic cannula, the entire aortic arch showed to be dissected, including the brachiocephalic trunk, the left common

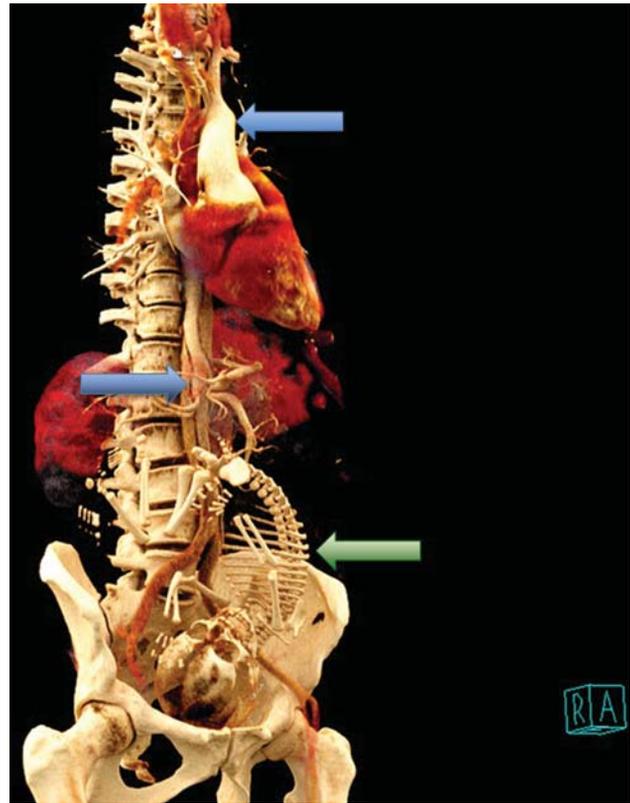


Fig. 2 Three-dimensional reconstruction showing aortic dissection reaching from ascending aorta (light blue arrow) to the infrarenal aorta (dark blue arrow). The unborn child was in utero (green arrow) at this moment and was delivered via cesarian section during cardiopulmonary resuscitation of the mother.

carotid artery, and the left subclavian artery. Brachiocephalic vessels were controlled by tourniquets. After that, the affected vessels as well as the aortic arch were resected. Subsequently, a graft-to-descending aorta anastomosis with a hybrid prosthesis was performed. After clamping of the prosthesis, the neo-aorta was cannulated for CPB. Following this, a graft-to-graft anastomosis, reconstruction of the brachiocephalic trunk, the left common carotid artery, and the left subclavian artery and the corresponding anastomoses to the hybrid prosthesis were performed. Despite intensive inotropic support, termination of CPB was not possible; therefore, a central venoarterial extracorporeal membrane oxygenation (ECMO) was implanted. Under these conditions, the patient was transferred to the intensive care unit (ICU). Despite this extensive surgical procedure and an elaborated postoperative ICU stay, it was possible to discharge the patient after ECMO weaning into neurological rehabilitation. The clinical course of the newborn child was uneventful.

Discussion

Untreated aortic aneurysm in women with MFS, who are especially vulnerable in pregnancy can be fatal as aortic dissection is associated with high rates in morbidity and mortality.¹⁰ Additionally, the fetal mortality rate is up to 50% in case of an aortic dissection.¹¹ Nevertheless, it was possible to save both

Table 1 International recommendations summarized from the following studies

| Counseling and management of patients with MFS during pregnancy ^{9,16,18} | |
|--|--|
| Risk assessment prior to or during pregnancy | <p>Advise patients with MFS <i>not to become pregnant</i>, if⁹</p> <ul style="list-style-type: none"> • Ascending aorta >45 mm or • Ascending aorta >40 mm and family history of dissection <p>Counseling about the risk of aortic dissection prior to and during pregnancy^{9,16}</p> <ul style="list-style-type: none"> • No dilatation, intermediate risk of maternal mortality (maternal cardiac event rate, 10–19%)¹⁸ • 40–45 mm, significantly increased risk of maternal mortality; requires expert counseling (maternal cardiac event rate, 19–27%)¹⁸ <ul style="list-style-type: none"> • >45 mm, extremely high risk of maternal mortality; <i>termination should be sensitively discussed</i> if pregnancy occurs (maternal cardiac event rate, 40–100%)¹⁸ |
| Diagnostic management | <ul style="list-style-type: none"> • <i>Echocardiographic measurements every 4–12 wk</i> of the ascending aortic dimensions are recommended to detect aortic expansion until birth and 6 months postpartum^{9,16} <ul style="list-style-type: none"> • For imaging of dilated distal ascending aorta, aortic arch, or descending aorta, MRI (without gadolinium) is recommended¹⁶ |
| Therapeutic management | <p>Medical</p> <ul style="list-style-type: none"> • Strict blood pressure control¹⁶, <i>β-blocker therapy</i> throughout pregnancy⁹ <p>Cardiac</p> <ul style="list-style-type: none"> • Consider <i>prophylactic surgery</i> when aortic dilatation and/or advancing aortic valve regurgitation are documented^{9,16} • Delivery where cardiothoracic surgery is available¹⁶ • Ascending aorta <40 mm, vaginal delivery is feasible⁹ • Ascending aorta >45 mm or (history of) dissection, <i>cesarean delivery should be considered</i>⁹ <p>Type A aortic dissection during pregnancy</p> <ul style="list-style-type: none"> • Multidisciplinary care including obstetricians, fetal care, cardiothoracic/vascular surgeon, and anesthesiologists. Optimal perioperative management including full maternal and fetal monitoring, attention to cardiopulmonary bypass, pulsatile perfusion, etc. <ul style="list-style-type: none"> • Viable fetus: cesarian section followed by aortic surgery • Nonviable fetus: aortic surgery with <i>fetus in utero</i> |

Abbreviation: MFS, Marfan's syndrome.

mother and child, despite the increased perioperative risk.¹² This case shows dramatically that a high alertness regarding cardiovascular surveillance in the state of pregnancy is crucial for these patients. Despite the fact that the women in our case was pregnant for the third time and MFS as well as aortic dilation was diagnosed previously, there was a lack in education about the risks accompanied by pregnancy regarding aortic dissection. Periodically monitoring changes in the aortic diameter could have been performed during pregnancy. This shows that a multidisciplinary approach, involving gynecologist, cardiologist, and cardiac surgeons is mandatory to monitor affected patients. Medical therapy can be essential to decrease the risk for aortic dissection in pregnant women. While some drugs are contraindicated in pregnancy (angiotensin-converting enzyme inhibitors), some β and α adrenergic blockers can be prescribed safely and are associated with a decline in dissection rates.¹³ Additionally, if a critical aortic diameter is reached during pregnancy, planned termination or elective aortic surgery can be discussed.¹⁴ Hereby, the risk for acute aortic dissection could be decreased, which in turn would decrease mortality rates for the mother and the child at the same time.

In terms of women with MFS contemplating pregnancy, prepregnancy counseling is advised to evaluate the specific risk for each patient.¹⁵ Genetic analyses of the mutation type may be helpful to assess the risk for cardiovascular complications as studies have shown that certain mutation

types are associated with an increased risk in cardiovascular events.³ Magnetic resonance imaging to detect preexisting aneurysms can be a useful tool to further create an individual risk profile for women with MFS. If aortic dilation is diagnosed, prophylactic aortic replacement should be discussed. Current guidelines recommend that aortic replacement should be considered in patients with an aortic diameter of 5.5 cm even if they have no symptoms¹⁶ and studies show that aortic replacement can be performed safely, especially in young patients with no preexisting comorbidities.¹⁷

Conclusion

Acute type A dissection in pregnant women with MFS is associated with high rates regarding morbidity and mortality. Despite a detailed understanding of changes in the cardiovascular system in the state of pregnancy and the well-known association of aortic events in patients with MFS, there are yet no universal recommendations for affected women. ► **Table 1** summarizes international recommendations regarding counseling and management of patients with MFS prior to and during pregnancy. Close monitoring for pregnant women with MFS and a detailed education prior to pregnancy are crucial measures. Prophylactic aortic replacement may be discussed and can be performed safely in young patients without comorbidities.

Authors' Contribution

Concept/design and critical revision of article were done by C.H. and M.W. Also, C.H. contributed in funding. Data interpretation and drafting article were done by F.H. and P.P.M., respectively.

Institutional Review Board Waiver and Patient Consent Statement

Patient signed informed consent related to clinical course, therefore and due to its retrospective nature of the educational case report, the Institutional Review Board was waived.

Funding

This publication was supported by Deutsche Forschungsgemeinschaft and University of Erlangen Foundation within the funding programme Open Access Publishing.

Conflict of Interest

None declared.

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