Canadian Journal of Cardiology 32 (2016) 78–85

Review

Pregnancy and Thoracic Aortic Disease: Managing the Risks

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ABSTRACT

The most common aortopathies in women of childbearing age are bicuspid aortic valve, coarctation of the aorta, Marfan syndrome, Ehlers-Danlos syndrome, Loeys-Dietz syndrome, SMAD3 aortopathy, Turner syndrome, and familial thoracic aneurysm and dissection. The hemodynamic and hormonal changes of pregnancy increase the risk of progressive dilatation or dissection of the aorta in these women. The presence of hypertension increases the risk further. Therefore, appropriate preconception counselling is advised. For women who become pregnant, serial follow-up by a specialized multidisciplinary team throughout pregnancy and postpartum period is required. In this review we discuss risk assessment and management strategies for women with aortopathies.

RÉSUMÉ

Les aortopathies les plus courantes chez les femmes en âge de procréer sont les suivantes : bicuspidie valvulaire aortique, coarctation de l'aorte, syndrome de Marfan, syndrome d'Ehlers-Danlos, syndrome de Loeys-Dietz, aortopathie associée au gène SMAD3, syndrome de Turner et formes familiales d'anévrisme et de dissection de l'aorte thoracique. Les changements hormonaux et hémodynamiques associés à une grossesse augmentent le risque d'une dilatation progressive ou d'une dissection de l'aorte chez ces femmes. Ce risque s'accroît en présence d'hypertension. Par conséquent, des conseils appropriés avant la conception sont de mise. Chez les femmes qui sont enceintes, un suivi périodique par une équipe multidisciplinaire spécialisée tout au long de la grossesse et durant la période postpartum est nécessaire. Dans cette analyse, nous discutons de l'évaluation du risque et des stratégies de prise en charge des femmes présentant une aortopathie.

Thoracic aortic disease (TAD) can present in young women of childbearing age. It is most commonly related to bicuspid aortic valve (BAV), coarctation of the aorta, Marfan syndrome, Ehlers-Danlos syndrome, Loeys-Dietz syndrome, SMAD3 aortopathy, Turner syndrome, or familial thoracic aneurysm and dissection (FTAAD). Some women will have a thoracic aortic aneurysm (TAA) and others have normal aortic dimensions. Aortopathies are a major cause of maternal mortality in pregnancy and can be undiagnosed until the fatal event.[1] In a Dutch nationwide prospective study of cardiac mortality during pregnancy, the overall maternal mortality rate during pregnancy was 3 per 100,000 deliveries and nearly half of the deaths were caused by aortic dissection.[2]

Pregnancy causes characteristic hemodynamic changes, including an increase in blood volume, heart rate, and stroke volume.[3,4] This increased volume overload is at least partly counterbalanced by a decrease of peripheral vascular resistance, diastolic blood pressure, and aortic augmentation index. Furthermore, hormonal changes might lead to less corrugation of the aortic elastic fibres and thus to fragmentation of the aortic reticulin fibres.[5,6]

The increased hemodynamic stress and structural changes of the vascular wall might contribute to progressive aortic dilatation or dissection in pregnant women with aortopathies. The presence of (gestational) hypertension further increases the risk.[7] Appropriate preconception evaluation and serial follow-up by a specialized multidisciplinary team throughout pregnancy and postpartum period is advised. In this review we discuss risk assessment and management of TAD in pregnancy.

Complications During Pregnancy

Maternal risks

Aortic growth rate. Growth of the aortic root is a normal phenomenon in healthy women during pregnancy. The

Received for publication June 1, 2015. Accepted September 11, 2015.

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See page 83 for disclosure information.

http://dx.doi.org/10.1016/j.cjca.2015.09.003

maximum diameter is reached during the third trimester; but at 6 weeks postpartum the diameter remains enlarged by an average of 1 mm.[8] In nonpregnant women with Marfan syndrome, the average aortic growth rate is higher, approximately 0.38 mm per year.[9] A recent study showed an additional increase in aortic growth rate during pregnancy to 0.3 mm per month in Marfan women. The increase in aortic dilatation rate decreased after delivery, but remained higher than the prepregnancy rate.[10] In this study, patients with an increased aortic dilatation rate during pregnancy were also at increased risk for aortic complications after long-term follow-up.[10] Two smaller studies have reported no difference in aortic growth rate between baseline and during pregnancy.[11,12] However, in Marfan women with an aortic root diameter larger than 40 mm (TAA), pregnancy did influence the long-term dilatation rate.[11] Apart from aortic growth rates in pregnant Marfan patients, the only study on other aortopathies concerns BAV patients. In a comparison of pregnant and nonpregnant BAV patients no increase in TAA risk during 216 pregnancies was found.[13]

Aortic dissection and rupture. The potential life-threatening consequence of aortopathies is aortic dissection and rupture. In the general population, the incidence of aortic dissection outside of pregnancy is 6 per 100,000 individuals per year,[14] and pregnancy increases the incidence of dissection in the normal population 100-fold, to approximately 0.6%.[15]

The incidence of aortic dissection in women with aortopathies is already increased when women are not pregnant, to 31 per 100,000 in those with BAV,[16] at least 36 per 100,000[17] in those with Turner syndrome,[17,18] and 170 per 100,000 in those with Marfan syndrome.[19] In Marfan patients, the probability of aortic dissection, when not pregnant, largely depends on aortic diameter, with an incidence of 0.3% at 45-49 mm (95% confidence interval, 0.00-0.71) and 1.33% at 50-54 mm (95% confidence interval, 0.00-3.93).[19] Aortic diameter is also a predictor for aortic dissection in patients with BAV,[20] but is less predictive for Loeys-Dietz[21] and vascular Ehlers-Danlos syndrome.[22] Other risk factors for aortic dissection are the site of dilatation, the rate of aortic progression, previous aortic dissection, associated lesions, the specific vascular syndrome, and familial history of dissection or sudden death.[10,19,23-25] Although data on aortic complications during pregnancy in the different aortopathies are limited, several studies suggest that pregnancy further increases the incidence of aortic dissection in these disorders (Table 1). Notably, most of the TAD patients with aortic dissection during pregnancy were either not receiving preventative measures[12,26,27] or had a contraindication for pregnancy such as a previous dissection.[11,12]

One of the most extensive studies on this topic was performed in BAV patients, in whom no aortic complications were observed in 82 patients during 216 pregnancies. However, in this study only 8% of the women had an aortic diameter $>$ 40 mm.[13] We could identify 11 case reports on aortic dissection in pregnant BAV women. Of the 11 cases, 3 patients had Turner and 1 had Marfan syndrome.[43]

Several studies have reported the occurrence of aortic dissection in pregnant Marfan patients (Table 1). These studies reported different incidence rates, but overall results of these studies suggest that pregnancy increases the risk of aortic dissection in Marfan women.

There are also a number of reports on pregnancy in other, less prevalent, aortopathies. Two studies on Loeys-Dietz syndrome reported a high incidence of aortic dissection during pregnancy.[34,35] These pregnancy cohorts were small in size and it is not clear whether preventative measures were taken. In a study on pregnancy in patients with SMAD3 aortopathy, often seen as a subtype of the Loeys-Dietz syndrome, there were no cases of aortic complications.[36]

In vascular Ehlers-Danlos syndrome,[22] also a rare and severe connective tissue disorder characterized by frail vascular tissue, vascular rupture during pregnancy has been reported as high as 50%,[37] with mortality rates between 5%[38,39] and 50%.[37] In a recent study on 256 vascular Ehlers-Danlos patients who experienced 565 pregnancies, a high mortality rate of 6.5% was reported during pregnancy, without an increase in the long-term mortality risk of this patient group.[39]

In one of the least prevalent aortopathies, namely Turner syndrome, a retrospective study reported an incidence of 2% for aortic dissection during pregnancy.[40] BAV, aortic coarctation, and hypertension in the Turner population[44] are believed to further increase the risk of aortic complications during pregnancy. In a study of pregnant women with aortic coarctation,[41] the only aortic dissection occurred in a woman with Turner syndrome and BAV. However, there are also case reports on aortic complications in pregnant women with aortic coarctation who do not have Turner syndrome.[45]

A heterogeneous group of aortopathies, collectively called FTAAD, are caused by mutations in different genes. It is known that patients with a mutation in the α-actin 2 gene are at substantial risk of aortic dissection during pregnancy.[42] Further research is needed to determine the risk of aortic complications in FTAAD patients with other known or unknown mutations.

Next to aortic dissection or rupture, the different aortopathies might also lead to additional complications, such as aortic insufficiency and dissection and/or rupture of smaller arteries such as the carotid artery. In that light, the opposite is seen in fibromuscular dysplasia (FMD), a disorder that causes carotid or renal artery stenosis or dilatation, and occasionally aortic dissection is observed.[46,47] Although there are case reports on pregnant patients with FMD who have experienced complications because of either renal or carotid artery stenosis,[48,49] there are no reports of aortic dissection or rupture during pregnancy in FMD patients.

Aortic dissection might occur at any time during gestation, but is more frequent in the third trimester and postpartum.[42,50] Nicely reviewed, dissections occur for approximately 5% in the first trimester, 10% in the second trimester, 50% in the third trimester, and 20% postpartum.[50] The high incidence of aortic dissection in the third trimester might be because of the increase in cardiac output, which peaks toward the end of pregnancy.[4] Most dissections originate in the ascending aorta (type A dissection), but dissections in the descending part of the thoracic aorta (type B) might also occur.[51]

Obviously, aortic dissection during pregnancy can have devastating consequences. Overall, maternal mortality from

Table 1. Studies on pregnancy in patients with thoracic aortic disease

Reference	Study description			Cases of aortic dissection or rupture			
	Type of study	Pregnant patients included, n	Pregnancies, n	Aortic dissection or rupture	Preventative measures	Diagnosis before pregnancy	Additional remarks on aortic dissection and/or rupture cases
BAV							
McKellar et al.[13]	Retrospective	82	216	0	–	–	
MFS							
Donnelly et al.[10]	Retrospective	69	199	0	–	–	
Meijboom et al.[11]	Prospective	23	47	1	1 of 1	1 of 1	The patient experienced an aortic dissection before pregnancy
Rossiter et al.[12]	Prospective	21	45	2	1 of 2	2 of 2	One of 2 patients had a prepregnancy descending aortic dissection and used intravenous cocaine during pregnancy
Pacini et al.[26]	Retrospective	85	160	7	0 of 7	5 of 7	
Lind and Wallenburg[27]	Retrospective	44	78	5	0 of 5	3 of 5	
Meijboom et al.[28]	Retrospective	63	142	1	1 of 1	1 of 1	The patient had a type A dissection prepregnancy
Curry et al.[29]	Retrospective	21	29	1	NR	NR	
Hassan et al.[30]	Retrospective	339	339	6	NR	NR	
Pyeritz[31]	Retrospective	26	105	0	–	–	
Lipscomb et al.[32]	Retrospective	36	91	4	1 of 5	1 of 5	
Omnes[33]	Prospective	18	22	1	0 of 1	1 of 1	
LDS							
Loeys et al.[34]	Prospective	12	21	4	NR	NR	
Tran-Fadulu et al.[35]	Retrospective	31	93	1	NR	NR	
SMAD3							
Van de Laar et al.[36]	Retrospective	13	23	0	–	–	
vEDS							
Rudd et al.[37]	Retrospective	10	10	3	NR	NR	Two cases of rupture of another major artery
Pepin et al.[38]	Retrospective	81	183	0	–	–	Seven cases of vascular rupture
Murray et al.[39]	Retrospective	256	565	0	–	–	Seven cases of arterial dissection/rupture
TS							
Karnis et al.[40]	Retrospective	200	200	4	4 of 4	4 of 4	Two of the 4 patients had a history of hypertension
Coarctation							
Beauchesne et al.[41]	Retrospective	50	118	1	0 of 1	1 of 1	The only patient who had Turner syndrome. She also had BAV
FTAAD							
Regalado et al.[42]	Retrospective	53	137	8	0 of 8	0 of 8	Five of the 8 women had hypertension

BAV, bicuspid aortic valve; FTAAD, familial aortic aneurysms and dissections; LDS, Loeys-Dietz Syndrome; MFS, Marfan syndrome; NR, not reported; TS, Turner syndrome; vEDS, vascular Ehlers-Danlos syndrome.

aortic dissection during pregnancy is described to be as high as 30% and fetal mortality as high as 50%.[52]

Obstetric and fetal outcome

During pregnancy in the general population, the incidence of major obstetrical complications is 400-600 per 100,000 for postpartum hemorrhage,[53] and 60 per 100,000 individuals per year for uterine rupture.[54] The incidence of these complications is not increased in pregnant women with Marfan syndrome,[27,50] yet are more common in women with vascular Ehlers-Danlos[55-57] and Loeys-Dietz syndrome,[34,36] which are the most severe connective tissue disorders. Increased fetal complications are reported in women with TAD. In the general population the incidence of intrauterine growth retardation is approximately 5%,[58] premature rupture of the fetal membranes (PROM) 8%,[59] preterm birth 12%-13%,[60] and the miscarriage rate in women who are aware of their pregnancy is 12%-15%.[61] In 8 studies, 3 prospective[11,12,33] and 5 retrospective,[10,27-30] fetal outcome has been reported in women with Marfan syndrome. Overall, the studies suggest that pregnancy in the Marfan patient is associated with a higher incidence of fetal complications. In 5 of 6 studies on the incidence of intrauterine growth retardation and small for gestational age, higher rates of these complications were observed in women with Marfan pregnancy.[27-30,33] Three[10,28,33] of the 6 studies mention that adverse fetal outcome was associated with a higher rate of β-adrenergic blocker usage.[62-64] However, in one study, β-blockers were not administered in any of the pregnancies that led to adverse fetal outcome,[27] which suggests that not only β-blockers but other unknown factors also contribute to the higher risk of fetal growth retardation.

Three[11,28,30] of 8[10-12,27-30,33] studies showed a greater incidence of premature birth, which might be because of PROM.[28] A relationship between connective tissue disorders

and PROM has been suggested.[65,66] Some studies reported a higher rate of miscarriages[11,28,33] or recurrent abortions in Marfan women.[28,31] Environmental factors, other genetic variants, epigenetic programming, and perhaps differences in the type of fibrillin-1 gene mutation, might explain why certain women with Marfan syndrome are at higher risk for miscarriages than others. Pregnancy in vascular Ehlers-Danlos patients is also associated with PROM, miscarriages, and prematurity.[55,57,65,66] The highest incidence observed for spontaneous abortions was 29% and for premature birth 23%.[55] Further studies are required to assess the fetal and neonatal outcome in other TAD patients.

Table 2. Key recommendations for the management of pregnancy in the TAD patient

Stage of pregnancy	Recommendations
Preconception evaluation	• TAD patients with a pregnancy wish should be counselled on the maternal and fetal risks • Prepregnancy imaging of the entire aorta with MRI is advised
Pharmacologic treatment	• Start β-blockers before pregnancy for MFS, LDS, vEDS, FTAAD, and BAV or Turner patients with aortic dilatation • ARBs should be replaced by β-blockers as soon as contraceptive measures are stopped
Evaluation during pregnancy	• Monitoring of the ascending aorta using ultrasound is recommended every 4-12 weeks • For TAD patients with a dilatation of the aortic arch, descending thoracic aorta and/or abdominal aorta, an MRI without gadolinium is advised • Routine ultrasound examinations for fetal monitoring is recommended with special attention to fetal growth in pregnant patients with β-blockade
Aortic surgery	• Elective aortic surgery should be performed at the ascending aorta diameters mentioned in Table 3 • If type A aortic dissection occurs during pregnancy, immediate surgery is required - If the gestational age is > 28 weeks, cesarean delivery is performed - For a gestational age < 28 weeks, one should consider cesarean delivery or fetal monitoring during cardiac surgery • For type B dissections pharmacologic treatment with regular monitoring of the aorta with MRI is indicated
Delivery	• In patients with vEDS or LDS, Marfan patients with an aortic diameter > 45 mm and BAV patients with a diameter > 50 mm, cesarean delivery is advised • In patients with a low or intermediate risk, vaginal delivery is relatively safe

ARB, angiotensin-II receptor 1 blocker; BAV, bicuspid aortic valve; FTAAD, familial aortic aneurysms and dissections; LDS, Loeys-Dietz syndrome; MFS, Marfan syndrome; MRI, magnetic resonance imaging; TAD, thoracic aortic disease; vEDS, vascular Ehlers-Danlos syndrome.

Table 3. Thresholds for surgery of women with aortopathies considering pregnancy

Disorder	Ascending aorta diameter
Marfan syndrome, FTAAD, SMAD3	45 mm
Loeys-Dietz syndrome	40-45 mm
Vascular Ehlers-Danlos syndrome	Contraindicated*
Bicuspid aortic valve	50 mm
Turner syndrome	27 mm/m^{2†}
Others	50 mm

Preconception ascending aortic replacement should be performed at the diameters mentioned in this table.

FTAAD, familial aortic aneurysms and dissections.

*Vascular Ehlers-Danlos syndrome is a contraindication for pregnancy.

† Indexed aortic diameter.

Managing the Risks

Preconception counselling

To assess the risks of pregnancy and to determine appropriate preventative measures, preconception counselling is needed.[67] Key recommendations for the management of pregnancy in the TAD patient are listed in Table 2. A multidisciplinary medical team needs to be involved in the preconception assessment including a cardiologist, geneticist, and obstetrician. Assessment should include a detailed history including family history, a physical examination, and a review of all aortic imaging.

Before pregnancy, imaging of the entire aorta using magnetic resonance imaging (MRI) is warranted. The need for elective aortic root replacement should be considered before conception.[51,68] The aortic diameter at which one should undergo elective surgery differs according to the condition (Table 3) and is discussed in more detail in the *Surgery Before Pregnancy* section. Pregnancy is contraindicated in patients with vascular Ehlers-Danlos, in Marfan patients with an aortic root diameter > 45 mm, and in patients with a history of aortic dissection.[11,67] In women who get pregnant despite this advice, a multidisciplinary team should be involved including an anaesthesiologist and cardiac surgeon to discuss the time and mode of delivery. Frequent evaluation during pregnancy is needed.

The maternal and fetal risks should also be discussed with the patient. The patient should be aware that elective aortic surgery does not eliminate the risk for aortic complications because dissection or rupture can occur at other sites in the aorta. The potential need for aortic surgery during pregnancy should also be addressed in high-risk women. It is important to educate the pregnant TAD patient about the symptoms of aortic dissection and that one should seek immediate medical attention if these symptoms occur.

Parents should also be aware of the transmission risk of their disorder to the child. Marfan,[69] Loeys-Dietz,[21] and vascular Ehlers-Danlos syndrome[22] have an autosomal dominant inheritance with a 50% transmission risk. For BAV and coarctation of the aorta, the mode of inheritance is less defined.[70] Some studies reported a 10% risk of transmission if one of the parents has BAV. In Turner syndrome, a

chromosomal disorder, the transmission risk is less clear. According to a study of 160 spontaneous pregnancies in Turner syndrome women, only 58% resulted in a live birth of which 20% had Turner syndrome and 14% Down syndrome.[71]

Possibilities for prenatal diagnosis should be discussed. Chorionic villus sampling or amniocentesis for detection of the parental mutation is possible, if the mutation in the parent is known. These tests are only useful and indicated if a positive result will lead to termination of the pregnancy. The patient should also be aware of the 1% risk of miscarriages associated with these tests.[72] An alternative to prenatal diagnosis is preimplantation genetic diagnosis. When the mutation of the parent is known, preimplantation genetic diagnosis allows selection of unaffected embryos before implantation.[73] This procedure, however, also has its disadvantages related to in vitro fertilization therapy.[74]

Evaluation during pregnancy

During pregnancy, regular assessment of the aortic diameter is needed. There is no available study on the optimal follow-up during pregnancy, but in the American College of Cardiology/American Heart Association 2010 TAD guidelines, monthly or bimonthly echocardiographic measurement of the ascending aorta diameter is advised.[75] The European Society of Cardiology guidelines recommend monitoring of the ascending aorta using echocardiography every 4-12 weeks during pregnancy and at 6 months postpartum.[67] MRI without gadolinium contrast is advised for pregnant women with aortopathies who have a dilatation of the aortic arch, descending aorta, and/or abdominal aorta.[67] This is preferred over computed tomography to avoid exposure of the fetus to radiation. The authors also recommend an initial MRI scan without gadolinium of the entire aorta with subsequent regular echocardiographic monitoring in women with aortopathies who have never had prepregnancy imaging. In addition to routine ultrasound examinations, advanced ultrasound scans can be performed for prenatal detection of congenital abnormalities associated with a connective tissue disorder. Although rare, cases of prenatal diagnosis of ascending aorta aneurysms using fetal echocardiography have been reported.[76,77]

Pharmacologic treatment

Drug treatment to prevent progression of aortic dilatation has been studied mainly in the nonpregnant Marfan population. In some studies, β-blockers have been reported to reduce aortic growth rate, and others have not observed this effect.[78,79] β-Blockers, however, are recommended in pregnant women with Marfan syndrome.[67] These drugs are also recommended in pregnant women with Loeys-Dietz Syndrome[21] and pregnant women with BAV or Turner syndrome with aortic dilatation. Celiprolol, a selective β1-receptor antagonist and a partial β2-receptor agonist, was shown to protect against arterial dissection and rupture in nonpregnant vascular Ehlers-Danlos patients.[80] For pregnant vascular Ehlers-Danlos patients, however, metoprolol is routinely administered.

Although rare, β-blockers are associated with fetal complications such as fetal growth retardation, small for gestational age, prematurity, and perinatal mortality.[62-64] The maternal benefits, however, usually exceed the fetal risks. Because β-blockers are indicated in most women with aortopathies, routine ultrasound examinations to monitor fetal growth is advised.

Other agents such as angiotensin-II receptor type 1 blockers, which have a beneficial effect in nonpregnant Marfan patients, are contraindicated because of their teratogenic effects.[81] Angiotensin-II receptor type 1 blockers should therefore be replaced by β-blockers in patients with TAD as soon as contraceptive measures are stopped.

Blood pressure regulation is important in women with aortopathies, especially during pregnancy, because of the increased risk of aortic dissection. In patients with coarctation of the aorta[82] and in Turner syndrome[44] there is already an increased risk of hypertension, which should be examined in these patients. Attention should be paid to pregnancy-related hypertension and pre-eclampsia, which might further increase the probability of aortic dissection during pregnancy.[83,84]

Aortic surgery

Surgery before pregnancy. The threshold for intervention before pregnancy varies according to the type of aortopathy (Table 3). According to European and the 2009 Canadian guidelines, previous aortic surgery should be performed at an aorta diameter $>$ 45 mm in Marfan patients.[67,85] In women with Marfan syndrome with aortic diameters of 40-45 mm, additional factors should be taken into account, such as family history of dissections and aortic growth rate. The 2014 Canadian Cardiovascular Society guidelines, however, recommend prepregnancy aortic replacement at an aorta diameter $>$ 40 mm with or without additional risk factors.[86] A threshold diameter of 45 mm, however, is generally used for Marfan patients. In women with other aortopathies and a high risk of aortic dissection such as FTAAD, European Society of Cardiology guidelines also suggest surgery before pregnancy at an ascending aortic diameter $>$ 45 mm.[67] For Loeys-Dietz women with a pregnancy wish, aortic replacement should be performed at an aortic diameter between 40 and 45 mm.[21]

In Turner patients specifically, an indexed aortic diameter is used, corrected for body surface area, because these women have a shorter stature and dissections can occur at smaller aortic diameters. For patients with acquired aortopathies (hypertension, atherosclerosis) prepregnancy surgery is recommended at an aortic diameter of $>$ 50 mm.[67] Coarctation of the aorta should be repaired before pregnancy.[67]

Surgery for complications during pregnancy. When a type A aortic dissection occurs early during pregnancy, immediate surgery is required,[67] and one should consider terminating the pregnancy. If the gestational age is $>$ 28 weeks, cesarean delivery can be performed with subsequent aortic surgery. For a gestational age $<$ 28 weeks, it is more difficult because of extreme prematurity. In that case, aortic surgery might be performed with fetal monitoring and modifications in anaesthesia.[87] High-pressure perfusion with pulsatile flow, normothermia, and continuous fetal monitoring during cardiac surgery are recommended. For type B dissections, aggressive pharmacologic therapy is usually appropriate to achieve hypotension which is, however, a risk for the fetus. In these patients regular monitoring of the aorta using MRI is advised. The overall maternal mortality rate of cardiothoracic

surgery during pregnancy is 3%,[88] and the fetal fatality rate is up to 30% and neonatal morbidity is 3%-6%.[88,89] Risk factors for fetal fatality during cardiovascular surgery are hypothermia and anoxic time.[89] More research is needed to determine the complications associated with cardiac surgery during pregnancy.

Management during delivery

During delivery, preventative measures are taken to decrease excessive aortic wall stress. There are no studies on the benefits and risks of cesarean delivery compared with vaginal delivery in women with hereditary aortopathies. Patients with vascular Ehlers-Danlos or Loeys-Dietz syndrome, Marfan patients with an aortic diameter > 45 mm, and BAV patients with dilatation > 50 mm are considered high risk patients and should undergo cesarean delivery mainly for optimal time management. In lower-risk women, vaginal delivery is considered relatively safe. During vaginal delivery, one should maintain a low threshold for epidural anaesthesia, forceps, and vacuum extraction. In patients with Marfan, Loeys-Dietz, or Ehlers-Danlos syndrome, epidural anaesthesia might be complicated because of dural ectasia or scoliosis. During contractions, the mother should be positioned on her left side or in a half sitting position to prevent aortocaval compression syndrome.[90]

Conclusions

Pregnancy in women with aortopathies is associated with a risk for maternal, fetal, and neonatal complications. Most studies on the risk of pregnancy in women with aortopathies have examined Marfan syndrome patients. Current guidelines are mainly based on experiences in Marfan syndrome and on expert opinion. Continued research on pregnancy and TAD is required to better define risks and management strategies.

Acknowledgements

The authors thank Romy Franken for her careful reading of the manuscript and her helpful suggestions.

Funding Sources

Interuniversity Cardiology Institute of The Netherlands 2013 Incentive.

Disclosures

The authors have no conflicts of interest to disclose.

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