Aortic surgery in pregnancy

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ABSTRACT

Pregnancy engenders changes in hemodynamics and the aortic wall that make a woman more susceptible to aortic dilatation and dissection. This is particularly true of women with aortic dilatation and an aortopathy, including the inherited fibrillinopathies, bicuspid aortic valve, and Turner syndrome. Women in these risk groups may be served best by undergoing elective aortic surgery before becoming pregnant. However, some women present during pregnancy with significant aortic dilatation, rapid expansion, or aortic dissection, and strategies to deal with these situations, while optimizing maternal and fetal outcomes, change as gestation progresses. This review summarizes the approaches to the management of aortic diseases and the conduct of aortic surgery in pregnancy. (*J Thorac Cardiovasc Surg* 2017;153:S44-8)

Strategy for acute type A dissection in pregnancy.

Central Message

Pregnant women with an aortopathy are more susceptible to AD. Management strategy changes as gestation progresses.

Perspective

Pregnancy engenders hemodynamic and aortic wall changes that make women with aortic dilatation more susceptible to AD, particularly those with an aortopathy. Although elective surgery before pregnancy is preferable, some women present during pregnancy with dilatation, rapid expansion, or acute dissection. This review summarizes management strategies, which change as gestation progresses.

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Cardiac surgery rarely is performed during pregnancy. The Mayo Clinic database yielded only 21 such cases from 1976 to 2009, documenting 1 maternal death but significant fetal complications, including mortality (14%), prematurity (52%), intrauterine growth retardation (5%), respiratory distress syndrome (33%), and development delay (14%).¹ Surgery involving the thoracic aorta during pregnancy is even less common, constituting only 2 cases (10%) in the Mayo series: 1 for a ruptured arch and 1 for rupture of the thoracoabdominal aorta.

PHYSIOLOGY OF PREGNANCY

Throughout gestation, progressive maternal hemodynamic and hematologic changes occur that affect aortic surgery. These include marked increases in heart rate, stroke volume, cardiac output, and left ventricular wall mass.² However, a concurrent decrease in systemic vascular resistance serves to maintain a relatively stable blood pressure. By full term, plasma volume expands by approximately 40%, but red cell mass increases by only 20%, resulting in a dilutional decrease in hematocrit and viscosity. A procoagulant state also occurs, with increases in clotting factors and changes in fibrinolysis that perhaps anticipate the need for hemostasis at delivery.³

The hyperdynamic state of pregnancy affects the significance of obstructive lesions. For example, transvalvular

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Abbreviations and Acronyms

AD = aortic dissection

BAV = bicuspid aortic valve

gradients across bicuspid aortic valves (BAVs) increase throughout pregnancy, peaking in the third trimester⁴; coarctation gradients increase, often causing refractory hypertension⁵; and even the gravid uterus can contribute to hypertension by causing aortoiliac compression.⁶

Particularly relevant to aortic surgery are pregnancy-related changes in aortic dimensions and the structural integrity of the aortic wall. Normally, aortic diameter increases through pregnancy, most strikingly during the third trimester and postpartum period, and the effect is more dramatic in hypertensive and pre-eclamptic women.⁷ It has been hypothesized that circulating angiogenic factors required for the vascular development of the placenta and fetus may simultaneously mediate remodeling of the maternal vasculature, which helps to accommodate the hyperdynamic state of pregnancy. Although aortic dimensions revert toward baseline over the first 6 months postpartum,⁸ with ensuing pregnancies an enduring increase in aortic diameter occurs.⁹ Significantly, pregnancy also engenders a progressive, estrogen-mediated structural weakening of the aortic media caused by reticulin fragmentation and elastin fiber disorganization that peaks in the third trimester and postpartum period.¹⁰

AORTIC DISSECTION IN PREGNANCY

The hemodynamic and aortic wall changes of pregnancy predispose to aortic dissection (AD), with more than 50% occurring in the third trimester and 33% occurring postpartum,⁶ when these changes are most pronounced. Although pregnancy is the most common risk factor for acute AD in young women, it is still a rare event. From 1998 to 2008, the Nationwide Inpatient Sample database logged 10,550,421 pregnancies and 41,000 acute ADs; only 44 (0.1%) ADs occurred during pregnancy, representing 0.0004% of pregnancies.¹¹ This comprises a yearly incidence of only 3.9 per 10,000, which is nonetheless 7-fold the yearly incidence of AD in the general population (6/100,000).¹² A recent literature review of aortic surgery in pregnancy found only 59 relevant articles, with 56 case reports and 3 series yielding 75 patients; 77% concerned type A dissection.¹³ Although rare, AD during pregnancy causes significant maternal and fetal mortality, as high as 30% and 50%, respectively.¹⁴

AORTOPATHY IN PREGNANCY

Acute AD in young women most commonly occurs during pregnancy in women with a predisposing aortopathy, most commonly Marfan disease. Although the incidence of AD in pregnant women with Marfan disease is only 0%

to 6.4%,¹⁵ it is still tremendously higher than in women without Marfan disease, and a number of studies have sought to identify risk factors for AD in pregnant women with Marfan disease. As in much of aortic surgery, these risks generally are based on aortic size. In a study of 199 pregnancies in nulliparous women with Marfan disease, the aortic diameter expanded more rapidly than in non-Marfan controls, dilating by 3 mm; postpartum the aortic diameter decreased, but not to baseline, and at more than 5 years of follow-up, women with Marfan had significantly larger aortic roots than nulliparous controls (43 vs 37 mm, $P = .002$); multivariate analysis showed that only aortic size and rate of expansion predicted long-term adverse cardiovascular events.¹⁶ In asymptomatic patients with Marfan disease, aortic diameter, normalized by using the cross-sectional area to height ratio, has been found to be a useful indicator for surgical intervention to prevent AD,¹⁷ and in pregnant women with fibrillinopathies, the risk of AD has been shown to increase as this ratio increases.¹⁸ The American College of Cardiology/American Heart Association Guidelines recommend that for women with Marfan disease who are contemplating pregnancy, it is reasonable to prophylactically replace the aortic root and ascending aorta if the diameter exceeds 4.0 cm (Table 1) (4.5 cm by European guidelines¹⁹) and that surgical repair is reasonable if the cross-sectional area/height ratio exceeds 10.²⁰

Similar recommendations have been published for other fibrillinopathies, including vascular type Ehlers–Danlos (≥ 4.5 cm),^{15,19} Loeys–Dietz (≥ 4.2 cm²¹; ≥ 4.5 cm by computed tomography²⁰), and familial thoracic aneurysm and dissection (≥ 4.5 cm),²² as well as aortopathies, such as Turner syndrome (≥ 27 mm/m²)²³ and Turner syndrome with BAV (≥ 25 mm/m²).²⁴ These recommendations are summarized in Table 1. Inactive Takayasu arteritis has a low risk of causing a cardiovascular event during pregnancy and does not require intervention.²⁵

BAV is thought to pose a risk for AD because of its association with ascending aortic dilatation, which is alternatively ascribed to hemodynamic forces, or an inherent aortopathy, and it has long been assumed that the hyperdynamic state of pregnancy accentuates that risk. Although pregnancy dramatically increases the gradient across a stenotic BAV,⁴ in a community-based study of 216 pregnancies in 88 women with BAV, no AD occurred, and during 12.3 years of follow-up, these women did not have an increased risk of aortic dilatation, aortic surgery, or AD.²⁶ However, in general, it is recommended that aortic coarctation should be repaired before pregnancy,¹⁹ and an increased risk may exist when BAV is associated with coarctation, particularly in Turner syndrome.^{27,28}

PERIOPERATIVE MANAGEMENT

Pregnant women with dilated thoracic aortas should undergo monthly or bimonthly echocardiographic measurement

TABLE 1. Elective surgery for aortopathy in pregnancy

Pathology	Elective surgery (mm)	Cesarean section (mm)
Aortopathy		
Marfan	$\geq 40^{20}$; $\geq 45^{19*}$	$\geq 45^{19\dagger}$
Ehlers–Danlos IV	$\geq 45^{19*}$	All ¹⁹
Loeys–Dietz	$\geq 42^{21}$ (TEE) ²⁰ ; 44–46 (CT/MRI) ²⁰	$\geq 45^{19}$
FTAAD	≥ 45	-
Turner ($\pm$ BAV)	≥ 27 (mm/m ²) ¹⁹ (≥ 25 [mm/m ²] ^{24,28})	-
Bicuspid valve	$\geq 50^{24\dagger,\S}$	-
Other		
Aortic valve surgery	$\geq 45^{20}$	-
Aneurysm	$\geq 55^{20}$	-
Rapid growth (>5 mm)	Per y or during pregnancy ²⁰	-

TEE, Transesophageal echocardiography; CT, computed tomography; MRI, magnetic resonance imaging; FTAAD, familial thoracic aneurysm and dissection; BAV, bicuspid aortic valve. *Pregnancy contraindicated. Note: tissue frailty may make elective surgery higher risk.¹⁹ †Consider at 40 to 45 mm.¹⁹ ‡Considering pregnancy a risk for dissection. §Pregnancy contraindicated for aortic disease ≥ 50 mm associated with BAV.¹⁹

of the ascending aortic dimensions until delivery.²⁰ If possible, elective surgery should be postponed until after delivery, but, as a general principle, necessary surgery should not be denied because of pregnancy and the surgery should be performed in a facility with capable obstetric and neonatal services, including the availability of fetal monitoring and Cesarean section.¹⁹

CARDIOPULMONARY BYPASS DURING PREGNANCY

The fetoplacental unit depends on properly functioning maternal and fetal circulations. On the fetal side, cardiac output is rate dependent, so it is important to avoid fetal bradycardia. Also, oxygen delivery is precarious. The fetal “oxygenator,” the placenta, drains into the fetal venous circulation so that fetal arterial blood is a venous admixture, with fetal hemoglobin approximately 65% saturated. The oxyhemoglobin dissociation curve is left shifted to improve oxygen uptake, but consequently oxygen delivery is decreased. On the maternal side, to maximize oxygen delivery, it is important to maintain maternal cardiac output, blood pressure, hemoglobin, and pH. It is also critical to avoid uterine contractions, because they severely restrict uterine and placental blood flow and are a major cause of fetal death. Of note, hemodilution should be avoided on cardiopulmonary bypass, because it may decrease progesterone levels and induce contractions.

A number of caveats to observe in the conduct of cardiopulmonary bypass have been recommended.^{1,29} Experimental evidence supports the use of pulsatile perfusion, because it promotes nitric oxide–mediated placental vasodilation,³⁰ but currently available equipment rarely support this mode. Strategies to augment maternal blood supply to the placenta include maintaining high flow, pressure, and hematocrit, whereas hypothermia, which

may induce placental vasoconstriction, should be minimized. Likewise, vasoconstrictors should be avoided. So initially, hypotension should be treated by increasing pump flow, but if necessary, pressors that tend to have less splanchnic vasoconstriction, such as phenylephrine or vasopressin, should be used sparingly.¹ If arch surgery is necessary, moderate hypothermia might be used, using selective antegrade cerebral perfusion, and distal perfusion should be maintained. Retrograde cerebral perfusion probably should be avoided, particularly if the inferior vena cava is snared, because elevated venous pressure may adversely affect the precarious placental perfusion gradient.

Although hypothermia is to be avoided, a recent literature survey included 5 cases in whom deep hypothermic circulatory arrest was used. Fetal (40%) and maternal (20%) mortality were high, but not 100%.¹³

TIMING OF AORTIC SURGERY IN PREGNANCY

The following overview of timing for aortic surgery in pregnancy should be qualified by noting that many of the recommendations are based on case series and a synthesis of current published opinion. There are few issued guidelines, and even these emphasize the want of evidence in this area.

For women seeking counseling before becoming pregnant, the European guidelines advise that pregnancy “should be discouraged” in patients with Marfan disease with aortic root dilatation 4.5 cm or greater, and, although recently called into question,³¹ pregnancy is contraindicated in Ehlers–Danlos syndrome. It is recommended that women in various risk groups who have aortic dilatation exceeding recommended size thresholds (Table 1) undergo elective surgery before pregnancy. During pregnancy, however, if significant aortic dilatation is discovered or rapid aortic expansion (≥ 5 mm) occurs, particularly in a member

of a high-risk group, a number of options need to be weighed in regard to the timing of surgery. Early in pregnancy, maternal risk may warrant termination of pregnancy. During the first trimester, surgery carries a significant risk of causing fetal developmental anomalies, whereas during the third trimester, significant risks of maternal and fetal circulatory collapse are incurred. The safest period to undertake semielective surgery is during the second trimester, because fetal organogenesis is largely completed, but maternal hemodynamic changes are not yet fully established.^{27,32}

Emergency surgery, such as repair of acute type A AD, should be undertaken regardless of trimester. During the first trimester and much of the second trimester, if continuation of pregnancy is desired, obstetric consultation is warranted. Fetal monitoring may provide useful feedback for managing cardiopulmonary bypass, but nonetheless fetal loss is common (Figure 1). During the late second and early third trimesters, strategy depends on fetal viability, which in turn may depend on the experience of the local neonatal service. Currently, the prospects for independent fetal survival is very poor at less than 23 weeks gestation, but becomes increasingly likely over subsequent weeks. During this period, strategy involves balancing at least 3 risks: (1) the risk of fetal loss, if pregnancy is continued; (2) the risks of neonatal prematurity, if Cesarean section is performed; and (3) the risk of maternal mortality. Uterine smooth muscle contraction compressing the spiral arteries is a major component of hemostasis after delivery. In concert, however, anesthesia, smooth muscle relaxation, hypothermia, and obligatory full anticoagulation may pose a risk for fatal hemorrhage should Cesarean section be undertaken during cardiopulmonary bypass. Thus, if Cesarean section is part of the operative strategy, it should be undertaken before anticoagulation and cardiopulmonary bypass. With the increasing likelihood of independent fetal survival, particularly after 32 weeks gestation, Cesarean section followed by aortic repair offers the best chance for fetal and maternal survival.

Type B AD should be managed medically unless complicated by malperfusion, which should be managed by an endovascular approach if possible. Of note, the European Guidelines do not consider a previous type B dissection to be a contraindication to pregnancy.¹⁹

CONCLUSIONS

Maternal accommodations for pregnancy include the development of a hyperdynamic state and aortic wall changes that, although benefiting fetal development, increase the risk of AD, particularly for women with predisposing aortopathies. These factors must be considered in the perioperative management of pregnant patients and in the conduct of cardiopulmonary bypass. Appropriate surgical strategy involves choosing not only the best procedure but also the optimum timing for surgery. Although aortic

FIGURE 1. Strategy for acute type A dissection in pregnancy. FM, Fetal monitoring; CS, Cesarean section.

surgery rarely is required during pregnancy, an understanding of the progressive developmental and hemodynamic circumstances that occur throughout gestation is crucial to achieving the best fetal and maternal outcomes.

Conflict of Interest Statement

Authors have nothing to disclose with regard to commercial support.

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EDITORIAL COMMENTARY

In pregnancy, aortic tissue is the issue

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In this elegant report, Lansman and colleagues¹ provide an interesting review of a complex topic: Management of aortic disease and aortic dissection in pregnancy. The available literature offers little evidence concerning this

topic, so this review should be applauded. The authors concisely describe the hyperdynamic state and aortic wall changes that occur during

CT images of a type A dissection that occurred 1 week postpartum in a patient with Marfan syndrome.

Central Message

A multidisciplinary team can optimize management and determine whether pregnancy is inadvisable or aortic repair needed before pregnancy.

See Article page S44.