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Chapter

Aortic Aneurysm in Pregnancy

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Abstract

Aortic aneurysms in pregnancy are rare but often fatal due to the natural physiologic changes of pregnancy and comorbidities specific to pregnancy, which increase the risk for aortic dissection and rupture. These physiologic changes are most pronounced in the third trimester and during the peripartum period, when approximately one third of dissections occur. In patients with known aortic aneurysms or conditions that make them prone to aortic aneurysms, preconception counseling can make pregnancy safer and more manageable. Aortic aneurysms diagnosed during pregnancy are usually due to underlying connective tissue diseases or aortopathies that have not been previously diagnosed. These women require multidisciplinary care including but not limited to obstetrics and gynecology, maternal fetal medicine, neonatology, cardiology, cardiothoracic surgery, cardiothoracic anesthesia, and genetics. Decisions include screening for dissection, when to proceed with surgical management, the best mode and timing for delivery, postpartum care, and contraception.

Keywords: aortic dissection, pregnancy, postpartum, delivery timing, contraception

1. Introduction

Aortic aneurysms in pregnancy are a rare but potentially fatal occurrence. The incidence in the general population is low at 2.6–3.5 per 100,000 person-years [1]. Women present with aortic aneurysms at an average age of 67 years, which is older than the average age of men presenting at 60 years. A significant index of suspicion is necessary to identify aortic aneurysms during pregnancy. Symptoms of aortic aneurysm or aortic dissection include significant chest pain that may or may not radiate to the back, dyspnea, syncope, and dysphagia. Chest x-ray (PA and lateral) should not be withheld for pregnant women with such symptoms. Although the cardiac silhouette may appear mildly enlarged on chest x-ray during pregnancy, persistent symptoms with or without a widened mediastinum should be evaluated with further imaging. In any patient, the highest morbidity and mortality results from an aortic dissection. **Table 1** shows types of aortic dissections, their description, and their associated mortality rates.

The highest mortality risk is a Stanford type A acute dissection, which describes an intimal tear involving the ascending aorta allowing blood to flow into the medial layer, which can ultimately culminate in aortic rupture leading to ischemia, tamponade, aortic regurgitation, or stroke. Stanford type A dissections typically present with a pulse deficit, systolic blood pressure limb differential of over 20 mmHg, focal neurologic deficits, or syncope [2]. Stanford type B dissections, which involve the descending aorta, have a lower mortality rate of 10% after 30 days. These are usually managed medically with strict blood pressure control as opposed to type A dissections, which typically require surgical management [2].

Type of Dissection	Description	Mortality
Stanford type A	Involves ascending aorta	40%
Stanford type B	Involves descending aorta	10%

Table 1.
Types of aortic dissections and associated mortality rates [1].

Risk factors for aortic aneurysms and resulting aortic dissections vary depending on age. For older women in the general population, hypertension and atherosclerosis contribute to the development of aortic aneurysms and aortic dissections. In contrast, aortic aneurysms in reproductive aged women usually occur in the setting of an underlying connective tissue or genetic disorder, such as Marfan's disease, Ehlers-Danlos syndrome, Turner's syndrome, or Loeys-Dietz syndrome, or an underlying aortopathy, such as bicuspid aorta, coarctation of the aorta, familial thoracic aortic dissection syndrome, or aneurysm-osteoarthitis. Among women less than 40 years old with aortic aneurysms, pregnancy has been shown to increase the risk of dissection by up to 25 fold [1]. Fortunately, the incidence of aortic aneurysm in pregnancy is low at 0.05 per 100,000 person-years; however, maternal mortality is high and ranges from 21–53% [3]. Aortic dissections in pregnancy account for 0.1–0.4% of all dissections and represent 0.0004% of all pregnancies [4].

Due to the rare occurrence of aortic aneurysm and dissection in pregnancy, management is largely based on case series and expert opinion. Treatment is nuanced and complex often due to perceived conflicting priorities of maternal and fetal health. The family's wellbeing is absolutely dependent on maternal survival and long-term health so these are not actually separate priorities. This chapter will review the unique physiology and risk factors for aortic dissection in pregnant women with aortic aneurysms, optimal management of known aortic aneurysms in reproductive aged women prior to pregnancy, best practice guidelines for aortic aneurysms during pregnancy, mode and timing of delivery of the pregnancy, and recommendations for postpartum care including prevention of or planning for another high-risk pregnancy.

2. Aortic aneurysm and dissection in pregnancy

2.1 Physiology of pregnancy in aortic aneurysms

Aortic aneurysms in pregnancy have a much higher risk of progression to aortic dissection and rupture than aortic aneurysms in the general population. This difference is incompletely understood, but can be mostly explained by physiologic, specifically vascular, changes unique to pregnancy, as well as underlying connective tissue or genetic disorders that predispose reproductive aged women to aortic aneurysms. The clinical features of these diseases are often exacerbated in pregnancy and increase the morbidity and mortality of aortic aneurysms in pregnancy and the pregnancies themselves.

2.1.1 Vascular changes of pregnancy

During pregnancy, specific physiologic changes occur to support the fetus and prepare for childbirth. Cardiovascular changes begin in the first trimester and peak in the third trimester and peripartum period, starting at 28 weeks of gestation up until 4 weeks after delivery. The vascular changes of pregnancy can negatively

impact the progression of an aortic aneurysm and significantly contribute to the increased risk of aortic dissection in aortic aneurysm in pregnancy. Some of these changes include increased cardiac output, heart rate, circulating volume, and left ventricular mass, all of which increase the risk of aortic dissection and rupture [1]. Estrogen and progesterone, both hormones that significantly increase in pregnancy, have been proven to change the microstructure of the aortic media and intima layers, causing fragmentation of reticulum fibers and loss of corrugation of elastic fibers [4]. This weakening of the vessel walls further contributes to the increased risk of aortic dissection and rupture. Some hypothesize that the gravid uterus, as it increases in size, compresses the aorta and thus increases the aortic outflow resistance, also increasing the risk of aortic dissection [1]. All of these vascular changes are most pronounced in the third trimester, when 50% of aortic dissections occur, and during the peripartum period, 4 weeks before and 4 weeks after delivery, when 33% of aortic dissections occur [2]. Certain conditions specific to pregnancy, such as pre-eclampsia, which causes vasoconstriction and sudden severe elevation of blood pressures, significantly increases the risk for aortic dissection and further complicates management.

2.1.2 Connective tissue and genetic disorders in pregnancy

There are several connective tissue and genetic disorders that predispose reproductive aged women to aortic aneurysms and have serious implications during pregnancy, both on the progression to aortic dissection and rupture as well as the pregnancy itself. While not an exhaustive list, the connective tissue and genetic disorders that will be discussed in this section include Marfan syndrome, Ehlers-Danlos syndrome, Loeys-Dietz syndrome, and Turner syndrome.

Marfan syndrome is an autosomal dominant connective tissue disorder that affects approximately 3 in 10,000 people [3]. Among pregnant women with aortic aneurysms, Marfan syndrome causes approximately half of aortic dissections [1]. It is characterized by musculoskeletal abnormalities including pectus excavatum, ocular abnormalities, cardiovascular abnormalities including aortic aneurysm and dissection, pulmonary abnormalities including spontaneous pneumothorax, skin abnormalities including stretch marks, and dura abnormalities including lumbosacral dural ectasia [5]. In the past, life expectancy was extremely limited, usually due to cardiovascular complications related to the disease; however, with current medical and surgical therapy, life expectancy can extend into the 70s. Diagnosis of Marfan syndrome can be made clinically based on criteria within different organ systems based on positive or negative family history. In the absence of family history, 2 major criteria from 2 different organ systems and 1 other major or minor criteria from another organ system must be met. In the presence of family history, only 1 major criterion and 1 major or minor criterion from a different organ system is necessary for diagnosis. Diagnosis can be confirmed by genetic testing, which will show a mutation on the fibrillin gene on chromosome 15.

During pregnancy, women with Marfan disease incur a high risk of morbidity and mortality due to the possibility of aortic dissection. This risk is particularly pronounced when aortic root dilation is over 4 cm. When the aortic root dilation is less than 4 cm, the risk of dissection is 1%; when the aortic root dilation is over 4 cm, the risk of dissection increases to 10%. Additionally, when aortic root dilation is over 4–4.5 cm, pregnant women will likely experience an accelerated rate of aortic root growth as compared to nonpregnant women [5]. In addition to maternal cardiovascular risk, there is likely an increased risk of obstetric and postpartum complications including a higher rate of preterm labor, preterm premature rupture of membranes, maternal urinary incontinence, and pelvic organ prolapse postpartum.

In nonpregnant patients, beta-blockers significantly slow the growth of the aortic root and decrease the risk of aortic dissection. There is some concern about the use of these medications in pregnancy due to possible adverse effects for the fetus including intrauterine growth restriction and fetal bradycardia. The risk benefit ratio supports the use of betablockers for treatment of aortic aneurysm during pregnancy. Selective beta-1 receptor blockers, such as metoprolol, are usually chosen due to a decreased risk of intrauterine growth restriction [5].

Several medications commonly used in obstetrics require careful use in pregnant women with Marfan syndrome. In cases of threatened preterm labor, tocolytics such as magnesium sulfate and calcium channel blockers can be used without significant adverse events. However, beta-adrenergic agonists, such as terbutaline, may increase the risk for tachycardia and arrhythmias and thus should be used with extreme caution. Additionally, indomethacin, commonly used for tocolysis in threatened preterm labor less than 32 weeks of gestation, can cause fluid retention, which may increase the risk for aortic dissection, and thus should also be used with extreme caution [5].

Ehlers-Danlos syndrome, less common than Marfan syndrome, occurs in approximately 1 in 5000 people. There are 6 different types of Ehlers-Danlos syndrome, which generally causes hypermobility of the joints, poor wound healing, tissue fragility, and hyperelastic skin [3]. The most severe type of Ehlers-Danlos syndrome and the most dangerous during pregnancy is the vascular type of disease. While hypermobility of the joints and hyperelasticity of the skin are less common in this type of disease, there is a significantly increased risk of aortic dissection, which is often not preceded by aortic dilation [3]. Due to tissue fragility, pregnant women with Ehlers-Danlos syndrome vascular type also have an increased risk of peripartum mortality up to 12% due to the risk of arterial or uterine rupture and hemorrhage [3].

Loeys-Dietz syndrome is a connective tissue disorder caused by mutation of the genes TGFBR1 or TGFBR2. Loeys-Dietz syndrome shares many characteristics with Marfan syndrome and Ehlers-Danlos syndrome vascular type. The disease is characterized by pectus excavatum, joint hypermobility, arterial tortuosity, hypertelorism, and bifid uvula [3]. Life expectancy for women with Loeys-Dietz syndrome is short with a mean age of death at 26 years, usually due to vascular complications. Similar to Marfan syndrome and Ehlers-Danlos syndrome vascular type, tissue fragility contributes to an increased risk of aortic dissection and uterine rupture, conferring significant morbidity and mortality in pregnancy.

Turner syndrome is a genetic disorder caused by complete or partial loss of one of the X chromosomes in a female. It occurs in approximately 1 in 2000 female births and is characterized by short stature, delayed puberty, premature ovarian failure, learning disabilities, bicuspid aortic valve, coarctation of the aorta, and aortic arch abnormalities [3]. Due to the high rate of aortopathy in women with Turner syndrome, the risk of aortic dissection in pregnancy is significantly elevated over the general population. While most women with Turner syndrome experience infertility due to premature ovarian failure, women with mosaic Turner syndrome may be fertile and thus capable of pregnancy. The risk of death from acute aortic dissection among pregnant women with Turner syndrome is 2%, and thus pregnancy is not recommended if a cardiac anomaly is present [3].

While knowledge of these connective tissue and genetic disorders may aid in diagnosis, aortic dissection may still occur in the absence of known risk factors and thus should remain on the differential for any pregnant woman who presents with chest pain radiating to her back.

2.2 Management prior to conception

Screening for aortic aneurysm should ideally be performed prior to conception in at-risk women. At-risk women include reproductive aged women with a personal or family history of aortic aneurysm or dissection in the past, known connective tissue or genetic disorder predisposing them to aortic aneurysm, known aortopathy, congenital heart disease, previous cardiac surgery, or significant trauma [4].

Any female with known aortic disease should receive proper family planning and contraceptive counseling as soon as and ideally before they are able to become pregnant. Important aspects of counseling include the significant risk to maternal health and the heritable nature of many underlying diseases that cause aortic aneurysm and dissection. After careful review of a woman's individual risk or based on a woman's family planning desires, surrogacy and adoption may be the safest route for parenthood. Women should be aware that deciding to become pregnant can have serious consequences to their own health.

Baseline workup includes referral to and establishment of care with a cardiologist if not already done, echocardiography, electrocardiogram, and genetic consultation if necessary. Routine medications must be reviewed to make sure they are not teratogenic as many patients are stable on warfarin, ACE inhibitors, and ARBs, all of which carry significant fetal risk with exposure in utero [4].

Among women with Marfan syndrome, elective surgery is generally recommended prior to conception when the baseline aortic root dilation is greater than 4.7 cm [5]. This is due to the maternal risk of death exceeding 10% [4]. Preconception care should include echocardiography to assess the proximal and distal aortic diameters and valvular and cardiac function. In women with significant ventricular enlargement, Holter monitoring is recommended to evaluate for arrhythmias.

Elective surgery should also be recommended prior to pregnancy in women with bicuspid aortic valve and aortic root dilation greater than 5 cm due to the maternal risk of death exceeding 10% [4]. Due to the high risk of aortic dissection, elective surgery prior to pregnancy is also recommended in the following situations: history of aortic valve surgery and aortic root dilation greater than 4.5 cm, Loeys-Dietz syndrome and aortic root dilation greater than 4.2 cm, any aortic root dilation greater than 5.5 cm regardless of underlying connective tissue disease, and symptomatic, traumatic, or syphilitic aneurysms [4].

Importantly, while physicians may make recommendations one way or another, women have autonomy over when and if they wish to become pregnant and thus care teams must be willing to support women's choices or refer to a team that can provide proper clinical care.

2.3 Management during pregnancy

After thorough counseling or if a woman desires pregnancy regardless of the risks, management of aortic aneurysm in pregnancy requires close follow up, multidisciplinary care, and a heightened index of suspicion for aortic dissection or need for surgical intervention.

2.3.1 Screening during pregnancy

After pregnancy is established, strict blood pressure control and serial imaging are necessary to evaluate the aortic root diameter. Depending on the baseline aortic root diameter and the underlying cause of aortic aneurysm, imaging can

be obtained as frequently as every 4–8 weeks or as infrequently as every trimester throughout pregnancy and then postpartum [3]. Blood pressure control and heart rate control are recommended using beta-blockers to slow the growth of the aortic root and help prevent aortic dissection [5]. Prompt recognition of rapid growth of aortic aneurysm or aortic dissection is essential due to the high risk of progression to aortic rupture, maternal death, and poor fetal/neonatal outcomes. If the aortic root diameter is increasing too rapidly, surgery may be necessary during pregnancy to decrease the risk of aortic dissection.

2.3.2 Imaging during pregnancy

Physicians who are unfamiliar with imaging in pregnancy may worry about fetal risks with different imaging modalities. Echocardiogram is safe and sufficient to monitor stability or progression of known aortic aneurysm in pregnancy. CT or MRI should be used to evaluate the aortic arch or thoracic aorta if these areas are known or suspected to be involved during pregnancy. In the workup of aortic aneurysm, the benefit of an accurate diagnosis from a CT angiogram highly outweighs the radiation exposure risk for the maternal-fetal dyad and should not be withheld. The radiation exposure from a CT angiogram is 0.01–0.66 mGy, which is well below the threshold for fetal injury (50 mGy) [6].

In an acute situation, such as when an aortic dissection is suspected, it is important to select the correct and most accurate imaging modality. CT angiography imaging is considered the gold standard and accuracy of diagnosis of aortic dissection approaches 100% with the newest machines available at most hospitals [7]. Transesophageal echocardiography is usually sufficient for diagnosing dissection; however, studies have shown significant inter-observer variability. Magnetic resonance angiogram is able to properly evaluate left ventricular dysfunction; however, most imaging units refer acute patients to CT angiography due to the time sensitive nature of diagnosis of aortic dissection [8].

2.3.3 Surgery during pregnancy

When aortic aneurysm or aortic dissection occurs in pregnancy, management is largely based on case series and expert opinion. Withholding indicated surgery from a pregnant woman as a result of concern for teratogenesis, pregnancy loss, preterm birth, or litigation is unfounded, and may significantly contribute to both maternal and neonatal morbidity [9]. Recommendations for aortic aneurysm treatment and surgery triage are based on nonpregnant individuals. **Table 2** shows differing society guidelines for evidence-based timing of surgical intervention. In general, if the aortic root diameter exceeds 5 cm, rapid progression in size is noted, or there is aortic valve regurgitation, surgical intervention during pregnancy is recommended and the benefits absolutely outweigh the risks [3]. While there is no standardized definition of rapid progression, most agree that an increase of more than 3–5 mm is significant. Pregnant women with aortic aneurysms are more likely to have an underlying aortopathy, which should decrease thresholds for surgical repair as risk for aortic dissection, aortic rupture, and maternal death are increased. Preeclampsia also increases risk for dissection and rupture. Studies have also shown an increased risk of mortality for patients with shorter stature, independent of size of aneurysm, and thus some experts recommend basing clinical decisions on the maximal cross-sectional area, which is calculated by dividing the square centimeters of the maximal aortic root diameter by the patient's height in meters, rather than the absolute aortic root diameter. When this measurement is used, surgical intervention during pregnancy is recommended if the ratio is above 10 [4].

Society	Recommendation
Canadian society for vascular surgery	Aortic root diameter at or above 5 cm, less if growing faster than 10% per year
Japanese circulation society	Aortic root diameter at or above 6 cm, 5 cm if accompanied by pain
European society of cardiology	Aortic root diameter greater than 5.5 cm, less if there is an indication for surgery on the aortic valve to combine surgeries
American college of cardiology	Aortic root diameter at or above 5.5 cm, less if growing faster than 0.5 cm/year, 4-5 cm or maximal cross sectional area greater than 10 with aortopathy or bicuspid aortic valve

**Symptomatic patients or patients with aortic dissection require urgent surgical intervention regardless of aortic root diameter.*

Table 2.
Society guidelines for surgical intervention of asymptomatic thoracic aortic aneurysms [3, 10].*

Contrary to popular belief, cardiopulmonary bypass (CPB), which is necessary for cardiothoracic surgical intervention, is not strictly contraindicated during pregnancy. The optimal mode and timing of delivery in relation to cardiothoracic surgery is discussed in the next section, but for some women, continuing pregnancy during CPB may be considered if they have compelling surgical indications and a previable or extremely preterm fetus. During CPB, several modifications can improve the fetal mortality rate below 20%, including performing the procedure under normothermic conditions in the left lateral decubitus position to maintain placental perfusion and decrease the risk of uterine contractions and preterm labor, high pump flow rate over 2.5–2.7 $L/min^{-1}/m^{-2}$ to maintain placental perfusion, short CPB and aortic cross-clamp time, perfusion above 70 mmHg, and hematocrit over 28% [5, 11].

For some women, particularly in the late second or third trimester, cesarean delivery immediately before cardiothoracic surgical intervention is the preferred management option. A common misconception in this scenario is excessive uterine bleeding if CPB and full anticoagulation are performed soon after cesarean delivery. Post cesarean bleeding is controlled physiologically by myometrial contraction and a two-layer suture closure. Preventative and early detection strategies include leaving the laparotomy open during the cardiothoracic surgery to directly evaluate and manage any intra-abdominal bleeding, placement of a prophylactic intrauterine balloon to provide uterine tamponade, or active monitoring of vaginal bleeding with the use of Allen stirrups [5]. In a case series, twenty-one mothers who had CPB initiated immediately after cesarean delivery had an average blood loss of 800 mL. None had excessive bleeding requiring abdominal packing or hysterectomy, and none used the additional preventative strategies mentioned above [11].

2.3.4 Fetal diagnosis of Aortopathy

Genetic screening or testing of the fetus is recommended for autosomal dominant aortopathies due to the high risk of transmission in the offspring. This information can be used for delivery and neonatal care planning, such as what level of care is needed, or it can be used by parents to determine whether or not to continue this particular pregnancy. These conditions include but are not limited to Marfan syndrome, Ehlers-Danlos syndrome, and Loeys-Dietz syndrome. This can be accomplished by chorionic villus sampling or amniocentesis. Unfortunately, even after obtaining fetal or placental cells, diagnosis of Marfan syndrome in the fetus

may be missed as there are hundreds of mutations that have been identified on the fibrillin 1 gene, and not all of them have been definitely linked to Marfan syndrome [7]. Fetal sonographic findings may assist in fetal diagnosis of Marfan syndrome, such as cardiomegaly, dilated ascending aorta, dilated pulmonary artery, or dysplastic atrioventricular valves [7].

2.4 Mode and timing of delivery

Optimal management for the health of the mother and fetus depends on the size of the aortic aneurysm, presence or absence of aortic dissection, presence of underlying aortopathy, fetal gestational age, and desires for the pregnancy. Multidisciplinary counseling and informed consent are essential and complicated. This section will review best practices for the mode of delivery, including assisted vaginal or cesarean, and timing of delivery in relation to cardiothoracic surgical repair.

2.4.1 Mode of delivery

In pregnant women with aortic aneurysms, the mode of delivery depends on the presence of aortopathy, size, and stability of the aortic aneurysm. Vaginal delivery is recommended for women with aortic root diameters less than 4 cm. Epidural for adequate analgesia to help maintain blood pressure and heart rate control is recommended. Among pregnant women with Marfan syndrome, dural ectasia, or dural sac dilation, should be ruled out prior to epidural placement as they may often be asymptomatic in up to 90% of patients with Marfan syndrome [7]. Strict blood pressure control with antihypertensive medications and an assisted second stage of labor with either vacuum or forceps is recommended to decrease the risk of dissection [3].

Cesarean delivery is recommended for women with an aortic root diameter over 4 cm, severe aortic valve regurgitation, significant progression of aortic aneurysm, or history of a previous dissection or repair [7]. Detailed planning and communication with anesthesia, cardiology, and cardiothoracic surgery are necessary regardless of the mode of delivery.

2.4.2 Timing of delivery

Prior to fetal viability, providers must have a discussion with women about the option of or recommendation for pregnancy termination. For example, Marfan syndrome and other aortopathies with an aortic root size greater than 4 cm is an indication for pregnancy termination due to the significant risk of maternal death if the pregnancy is continued. If cardiothoracic surgical repair is indicated before fetal viability, termination of pregnancy should be strongly considered due to the fetal loss rate of up to 33% and the increased potential for long term neurologic impairment after CPB [12]. The definition of fetal viability is nuanced and depends on the woman's personal beliefs, the accuracy of gestational age dating, a particular state's legislation, and institutional guidelines. The American College of Obstetricians and Gynecologists defines the periviable period as between 20 and 25 weeks of gestation. Deliveries under 20 weeks 0 days are defined as spontaneous abortions. With accurate gestational age dating, there is no survival of neonates delivered at 20 and 21 weeks. Although there have been significant advances in neonatal intensive care, individualized and evidence-based conversations about neonate survival and life without significant neurologic impairment are essential.

After fetal viability, the decision analysis shifts to decisions about CPB and cardiac surgery with the fetus remaining in utero or delivery of the fetus just before the cardiac surgery. Risks of prematurity include but are not limited to neonatal death, low birthweight, respiratory distress, and neuro-developmental disabilities. Before 32 weeks of gestation, maternal administration of intravenous magnesium sulfate is recommended for fetal neuroprotection to reduce the risk of cerebral palsy. Before 34 weeks of gestation, corticosteroids are recommended for fetal lung maturation [13]. While there is no consensus, most experts recommend cesarean delivery immediately followed by CPB starting at 28–32 weeks of gestation as it is thought that at this gestational age, the benefits of delivery without exposure to maternal CPB outweigh the risks of neonatal prematurity [4, 5]. Advances in neonatal intensive care have improved neonatal survival and a lower gestational age for delivery may be reasonable (above 25 weeks) if the patient chooses to avoid the fetal risk of CPB and accept the risks of prematurity.

Before approximately 25 weeks of gestation, surgical repair via CPB with a viable fetus in utero may be the best management plan. The obstetrician and patient must then decide whether to monitor the fetal heart rate during cardiac surgery and what actions should be taken in the event of nonreassuring fetal heart rate, such as perfusion adjustments, intrauterine resuscitation, or delivery. This discussion requires shared decision making and multidisciplinary planning [5]. If neither urgent cesarean delivery nor intrauterine resuscitation are available or desired, intraoperative fetal monitoring is not necessary and may cause more harm than benefit. At all time points throughout the surgery, maternal life should be prioritized as maternal cardiac arrest or severe hypoxemia will most certainly lead to fetal death or extreme fetal morbidity. After 20 weeks gestation during a maternal cardiac arrest, the uterus should be emptied by hysterotomy no later than 4–5 minutes after arrest to maximize venous return and efficacy of maternal resuscitation efforts. If the fetus is viable, delivery no later than 4–5 minutes after maternal cardiac arrest and resuscitation also results in good neonatal outcomes. Tocodynamometry, to monitor uterine contractions, is generally recommended intraoperatively (when feasible) and postoperatively. Preterm contractions are common during and after CPB and may or may not result in preterm labor [5].

2.5 Postpartum care

Equally as important as managing acute complications of aortic aneurysm in pregnancy is planning for or prevention of the next high-risk pregnancy. This section discusses optimization of care in the postpartum period after delivery and options for contraception should the patient desire prevention of another pregnancy.

2.5.1 Postpartum surveillance

The postpartum period represents a particularly vulnerable timeframe for women as many of them will lose insurance coverage, neglect to care for themselves due to focus on their newborn, and are recovering from their delivery and potentially cardiothoracic surgery as well. The physiologic changes of pregnancy do not immediately return to pre-pregnancy after delivery, and many of the cardiovascular changes persist beyond the traditional 6 week postpartum period, when most women's insurance companies will terminate their coverage. In a recent cohort-crossover study, researchers found that the risk of aortic dissection and rupture remain increased as compared to the general population as long as 1 year after delivery [14]. This elevated risk of morbidity and mortality emphasizes the need for

careful coordination of postpartum care and surveillance, including regular follow up with cardiology and obstetrics and gynecology, careful maintenance of medication, and routine imaging and lab work.

2.5.2 Contraception

The postpartum period is a critical time to discuss contraception and plans for childbearing in the future. Unfortunately, due to the medical complexity of these patients and the effort required to coordinate care effectively, contraception is often forgotten in the care of the recently postpartum woman with an aortic aneurysm or aortic dissection during pregnancy. As mentioned earlier in this chapter, the discussion of family planning and contraception has ideally already occurred before or as soon as women with known aortic aneurysms are able to become pregnant. If this has not occurred, it is beneficial to discuss contraception during the prenatal course so that a plan is made for after delivery. If this has not occurred, this discussion can happen during the postpartum period. Primary care providers, cardiologists, and cardiothoracic surgeons may not be equipped to discuss the safest and most reliable contraceptive options with patients, and thus this discussion may be best suited for the obstetrician/gynecologist. Ideally, cardiologists and surgeons should become familiar with selective contraceptive methods (such as the progestin-only injectable) to provide short term so that women with cardiovascular disease do not conceive before seeing an obstetrician/gynecologist.

The Centers for Disease Control and Prevention developed the US Medical Eligibility Criteria (US MEC) for Contraceptive Use, which gives standardized recommendations for each type of contraceptive method in the setting of certain medical conditions [15]. In patients with complicated valvular heart disease, estrogen containing hormonal contraceptive methods are contraindicated due to the increased risk of heart attack and stroke. However, all other methods including the copper and levonorgestrel IUDs, the etonogestrel implant, the progestin-only injectable, and progestin-only pills, are considered safe and effective for these women. Contraceptive counseling should include discussion of a woman's priorities in different aspects of the contraceptive method, such as efficacy, side effects, patient autonomy, and future pregnancy plans.

3. Conclusion

Aortic aneurysm in pregnancy is a rare occurrence that leads to increased maternal, fetal, and neonatal morbidity and mortality. Specific changes to women's physiology and complications unique to pregnancy predispose pregnant women with aortic aneurysm to aortic dissection; therefore a high index of suspicion is lifesaving. Pregnant women with aneurysms are likely to have an underlying collagen disorder or aortopathy and are at even higher risk for aortic root diameter growth and progression to aortic dissection, rupture, and death. Such underlying conditions also increase the risk for poor obstetrical outcomes such as uterine rupture, premature delivery, and maternal hemorrhage. In women who are known to be at-risk for aortic aneurysm or aortic dissection, management and counseling prior to conception is essential and should include discussion of highly effective contraception, surrogacy, and adoption. Management of aortic aneurysm in pregnancy requires multidisciplinary care, imaging, and medication management. Indicated surgery should not be withheld simply because of the pregnancy or provider discomfort as absence of treatment results in worse outcomes. Decisions regarding surgery and mode and timing of delivery are nuanced, complicated, and require shared patient

centered decision making, effective communication, and cooperation from multiple specialists in a tertiary care center. With careful planning, adequate facilities, and skilled providers, favorable outcomes for mother and baby are possible for patients who experience aortic aneurysm and dissection in pregnancy.

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Conflict of interest

The authors declare no conflict of interest.

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