

## Acute Aortic Dissection in Pregnancy: Cesarean Section Prior to Repair of Acute Type A Dissection

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### Introduction

A 35-year-old G2P0010 with a medical history of Fibromyalgia presented at 32+3wks weeks of gestation with sudden onset of chest and back pain while bending over at her baby shower. On arrival to an outside hospital her vital signs were stable, and labs, and electrocardiogram were all within normal limits. There were no signs of fetal distress either by heart rate monitoring or fetal ultrasound. While at the outside hospital the patient became transiently hypoxemic with oxygen saturations in the high 80%. An arterial blood gas at the time revealed a respiratory alkalosis with hypoxemia on supplemental oxygen. This increased Alveolar-arterial gradient (A-a gradient) indicated a possible acute pulmonary embolism and the patient underwent Computed Tomography of the chest. No pulmonary embolism was seen but a 5cm aortic aneurysm and Type A dissection was diagnosed.

The patient was medically managed for her dissection and then transported to our center for surgical treatment. On arrival, her chest pain was initially well controlled on intravenous labetalol and morphine. While in the intensive care unit, the on call obstetrics team evaluated the patient and again the fetal monitoring was noted to be reassuring. After a few hours however the patient became extremely anxious and her chest pain increased in its severity. At this point, it was decided to proceed with combined cesarean deliver of the baby and surgical repair of the patient's aortic dissection. A multidisciplinary team consisting of cardiothoracic surgery, cardiac anesthesia, high-risk obstetrics, and neonatology were assembled.

The patient was taken to the operating room where using an arterial line for blood pressure monitoring and using a rapid sequence induction general anesthesia was induced while in the left uterine displacement position. Following endotracheal intubation an intraoperative transesophageal (TEE) echocardiogram was performed. The TEE confirmed a 5.5cm aortic aneurysm with a dissection flap in the sinuses of Valsalva extending distally to the sino-tubular junction. The dissection did not involve the coronary arteries, but did extend in to the aortic arch and further into the thoracic aorta. There was minimal aortic insufficiency but a left pleural effusion (assumed to be a hemothorax) was present. It was also noted that the aortic arch and descending aorta were dilated and aneurismal (Figures 1-3).

Once the diagnosis of aortic dissection was confirmed by TEE the patient's chest and abdomen were prepped and draped. The right femoral artery was isolated and a median sternotomy was made in the event that emergent cardiopulmonary bypass was required. A cesarean section was then performed and a live born 2070 gm male was delivered. The infant was electively intubated due to narcotic induced respiratory depression and taken to the Neonatal Care Unit for observation. The one and five minute apgar scores were 4 and 8.

Following adequate uterine hemostasis the patient was fully heparinized and the right femoral artery was cannulated for systemic

perfusion with bicaval venous cannulation for systemic venous drainage and to deliver retrograde cerebral perfusion. Cardiopulmonary bypass was initiated and following systemic cooling to 18°C deep hypothermic circulatory arrest (DHCA) was initiated. Retrograde cerebral perfusion was delivered during the DHCA period. The aortic root was then opened. On examination, the aortic dissection appeared to originate above both coronary arteries, nearly circumferentially dissecting down into both coronary ostia. Using Dacron grafts the aortic root, ascending aorta, and aortic arch were completely reconstructed. Due to preservation of the aortic valve and annulus the aortic valve was resuspended and did not require replacement. The patient was rewarmed and separated easily from cardiopulmonary bypass. There was minimal residual aortic insufficiency.

The patient's hospital course was uneventful and she and her healthy baby boy were discharged home on post operative day number eight.

The patient and baby subsequently tested positive for an identical single point identifiable fibrillin-1 mutation supporting a diagnosis of Marfan syndrome.

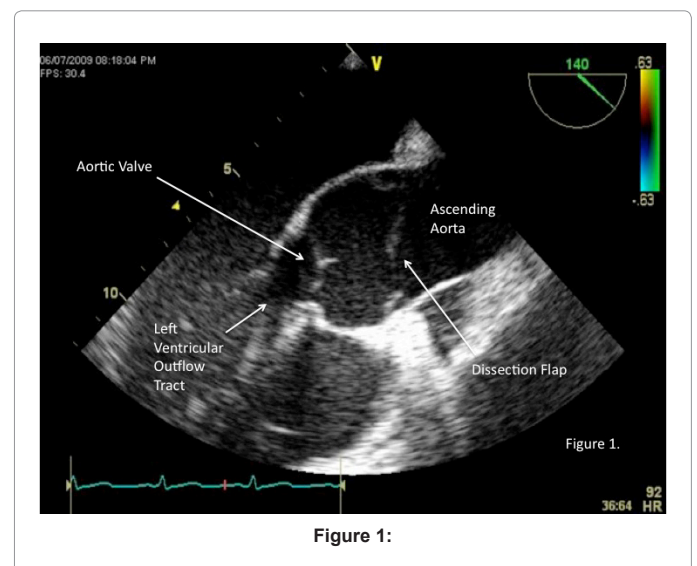


Figure 1:

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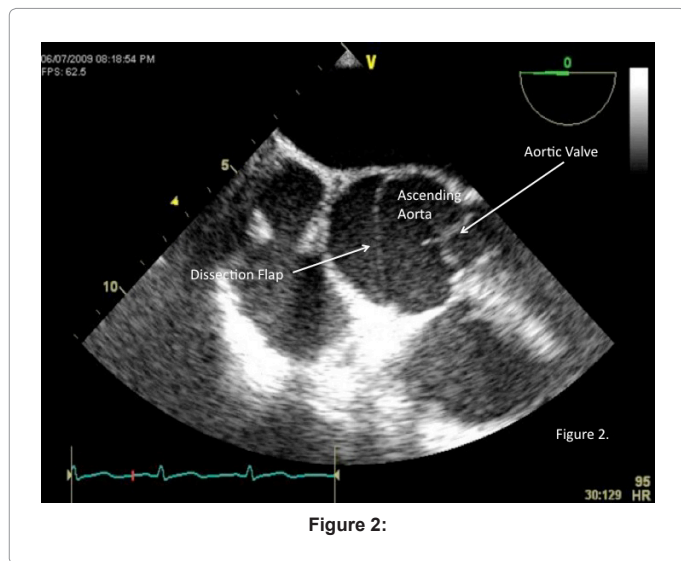


Figure 2:

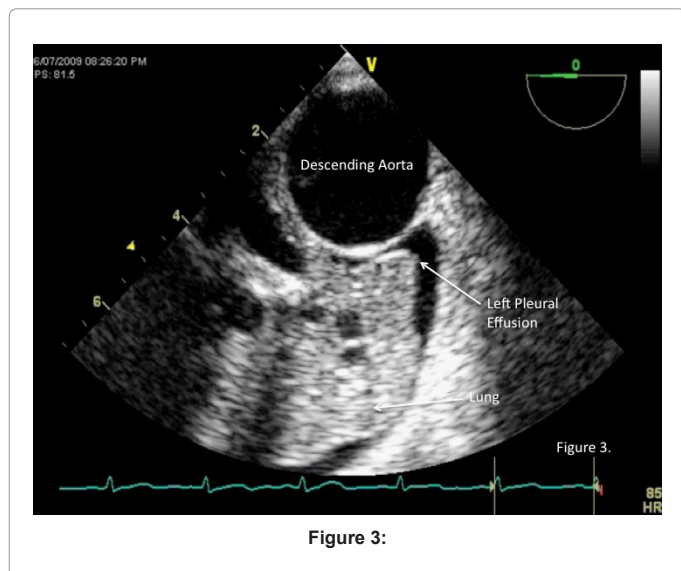


Figure 3:

## Discussion

In women under 40 years of age half of all aortic dissections occur during pregnancy or in the peri-partum period [1]. In pregnancy, the most common risk factors associated with acute aortic dissection include: Marfan syndrome, dilated aortic root (> 4cms), and dilated root with bicuspid aortic valve [2,3], Gestational hypertension, pre-eclampsia, and hormonal changes during pregnancy may also exacerbate or predispose pregnant patients to aortic dissection [4-6]. In addition, high levels of estrogen and progesterone that occur late in pregnancy may in themselves cause degeneration of the aortic wall. This is similar to the cystic medial necrosis seen in cases of aortic dissection in non-pregnant patients. These vascular changes include, smooth muscle hyperplasia, reduced acid mucopolysaccharides, and the loss structural integrity of elastic fibers [7].

The commonly used Stanford classification divides aortic dissections into two groups. Stanford Type A dissections involve the ascending aorta while Stanford Type B dissections primarily involve the descending aorta. (However, in rare cases, Type B dissections may

propagate retrograde and involve the aortic arch and ascending aorta). A more clinically useful approach uses the left subclavian artery to delineate the boundary between Type A and Type B dissections. Using this approach dissections that arise proximal to the left subclavian artery are considered Type A and those distal to the left subclavian as Type B [8]. Given the proximity to the aortic valve, coronary and carotid arteries Type A dissections are at most risk for life threatening events and require early surgical intervention. Type B dissections on the other hand are most often managed medically. The mortality rate for unrepaired Type A dissection is 1% per hour during the first 24 hours after the onset of symptoms and 75% at two week if left untreated. Causes of death include, rupture of the aorta causing sudden death, cardiac tamponade, acute aortic valvular insufficiency, and coronary or carotid artery avulsion or obstruction leading to myocardial infarction or stroke.

Initial management of Type A dissections includes: aggressive blood pressure control using invasive arterial monitoring and pain control. Intravenous  $\alpha$ -blockers or calcium channel blockers are the antihypertensive agents of choice as they act rapidly to decrease systemic blood pressure quickly. Labetolol which is commonly used to manage hypertension in pregnancy is particularly well suited for use in aortic dissections. The  $\alpha$ 1 antagonism lowers peripheral vascular resistance decreasing blood pressure. While the  $\beta$  sympathetic receptor antagonism decreases the rate of rise of ventricular force on the aorta (dP/dt). This decrease shear forces on the aortic wall and dissection flap reducing further dissection propagation. Labetolol also has the advantage of intravenous bolus dosing and can be administered as a rapidly titratable infusion.

Once a decision to perform a cesarean section in a patient with a Type A dissection is made the choice of anesthetic must be weighed carefully. Neuraxial blockade in the form of a spinal or epidural have been used successfully for cesarean section in patients with aortic dissection [9]. These techniques avoid the potentially life threatening hypertension and tachycardia that often accompany endotracheal intubation that can cause aortic rupture and death. As in our case, general anesthesia has the advantages of a secured airway in the event of hemodynamic instability and if combined repair of the aortic dissection is planned. The fact that this patient demonstrated a Mallampatti Class 3 airway raised the possibility of a difficult intubation. This was compounded by the known difficulty of the pregnant airway. It was felt that given her adequate thyromental distance and mouth opening as well as having advanced airway devices available a rapid sequence induction and intubation was an acceptable option. In the event that this patients airway was deemed too difficult to intubate other options would have included asleep fiberoptic intubation or other advanced techniques. Fortunately, these were not necessary.

In the operating room, the sequence of isolation of the femoral artery and performance of sternotomy, prior to cesarean section was chosen as the safest course of action in the event of acute maternal decompensation. The decision to perform the cesarean section and delivery prior to aortic repair include: the (1) use of profound hypothermia (18°C) and deep hypothermic circulatory arrest which cause a decrease in placental blood flow with accompanying fetal hypoxia and ultimately intrauterine death, (2) by delivery of the fetus the effects of aortocaval compression and maternal hypotension are averted, and (3) a decrease in maternal oxygen consumption. In the event of maternal hemodynamic instability or cardiac arrest delivery of the baby would give the mother the greatest chance of survival [10].

This is due to the gravid uterus compressing the diaphragm impairing ventilation as well as decreasing venous return to the heart and impeding cardiac output.

Following surgery, during discussion with a genetic counselor, it was discovered that the patient's father had died suddenly in his 40s from what had been thought to be either an acute myocardial infarction or aortic dissection. In addition, the patient disclosed a vague history of ocular complaints. Based on this history suggestive of connective tissue disease the patient underwent a transthoracic echocardiogram early in her pregnancy that did not show any aortic anomalies. This history was not known at the time of her presentation to the emergency room with chest pain and we must assume that her dissection occurred late in pregnancy.

As part of postoperative genetic counseling of the patient and her baby, a variation in one copy of the fibrillin-1 (FBN1) gene was discovered. The variant c.1147G>T was found in the heterozygous state in both mother and baby which leads to a nonsense mutation that supports the diagnosis of Marfan syndrome.

Marfan syndrome is a variable autosomal dominant disorder of connective tissue with a worldwide incidence of 1 in 5000 people. Prior to the advent of genetic testing the diagnosis of Marfan syndrome was based on a family history of physical signs such as long limbs, or aortic aneurysms [11]. Molecular genetic testing now allows for detection of mutations in the fibrillin-1 gene (FBN1). This gene codes for the glycoprotein protein fibrillin-1. Multiple Fibrillin-1 molecules crosslink together to form microfibrils which provide a matrix for the structural integrity of many parts of the body. A mutation in FBN1 causes the production of abnormal fibrilins and increases the activity of transforming growth factor (TGF- $\beta$ ). This likely causes weakened and abnormal elastic fibers and the known complications of Marfan syndrome. Most notably, aortic aneurysms and dissections, ocular lens dislocation, and abnormal connective tissue. In animal models, over activity of TGF-  $\beta$  has been linked to aortic dilation and can be prevented by administration of anti-TGF- $\beta$  antibodies [12,13]. Currently, a mutation in this gene supports the diagnosis of Marfan syndrome.

Following the diagnosis of Marfan syndrome both mother and child will need lifetime surveillance to serially follow aortic size and screen for the many other problems associated with Marfan syndrome.

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