SECTION OF MATERNAL FETAL MEDICINE
DEPARTMENT OF OBSTETRICS AND GYNECOLOGY
SOUTHERN PHILIPPINES MEDICAL CENTER
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CASE:

Chronic Dissecting Thoracic Aortic Stanford Type B Aneurysm in Pregnancy Treated with Intrapartum Thoracic Endovascular Aortic Repair

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Background:
The intent of this study is to report our first documented case of a Stanford type B dissecting aneurysm in a pregnant patient that was managed with Thoracic Endovascular Aortic Repair, which was the first and only case of its kind in the Philippines as of this writing.

1. INTRODUCTION:

Cardiovascular diseases during pregnancy account for considerable morbidity and mortality, with aortic aneurysms, complicated by dissection or rupture, being one of the top causes of morbidity and mortality. Aortic aneurysms in pregnancy are rare but potentially fatal. Aortic dissections in pregnancy account for 0.1–0.4% of all dissections and represent 0.0004% of all pregnancies affecting 5.5 women per 1 million during pregnancy and puerperium. Among women with aortic aneurysms less than 40 years old, 50% of aortic dissections and aortic aneurysms are associated with pregnancy and was noted to increase the risk of dissection by up to 25-fold (1). In a review of literature from 2003 to 2013, maternal mortality ranges from 21% to 53% (2). This increased risk in pregnancy is likely due to the hemodynamic changes that women experience during pregnancy such as increased heart rate, plasma volume, cardiac output, as well as the development of preeclampsia which is a common complication in pregnant women. In addition to the hemodynamic changes in pregnancy, hormones also induce changes in the aortic wall, and estrogen and progesterone have been proven to change the microstructure of the media and intima, causing fragmentation of reticulum fibers and loss of corrugation of elastic fibers (3). These vascular changes are generally the most pronounced in the third trimester, when 50% of dissections occur, and in the peripartum period, when 33% of dissections occur (4). Most cases of aneurysm in women are associated with underlying disorders such as Marfan syndrome. But there are also rare cases in which there is no underlying cause.

2. CASE PRESENTATION:

A 31-year-old G1, was initially seen at a local obstetrician's clinic for infertility of about 2 years. She had no known medical problems nor surgeries and was not taking any medications. She had a history of hypertension and diabetes on the paternal side of her family. Her father was a diagnosed case of polycystic kidney disease and a known hypertensive and had a sudden death at the age of 47 of a suspected cerebrovascular accident, since no autopsy was done. No other siblings or family members were reported to have other co-morbidities or cardiac problems.

During initial consultation, elevated BP of 160/100 mmHg was noted, and she was further referred to a Medical Internist. Laboratory tests were done, and on CT scan, an incidental finding of thoracic aneurysm was seen. She was initially referred to our institution under the management of Thoracic Cardiovascular Surgery. Eventually, A CT Aortogram with contrast was requested and pregnancy test was done prior to the procedure which turned out to be positive. She was referred to our Maternal Fetal Medicine Section for prenatal care and additional management. A transvaginal ultrasound showed an early live intrauterine pregnancy at 7 weeks and 4 days age of gestation by crown- rump-length, FHT of 156 beats per minute with normal ovaries. All her previous anti-hypertensive medications were discontinued and shifted to Methyldopa 250 mg 1 tablet TID and she was started on Folic acid 5 mg 1 tab once a day and Calcium 600 mg 1 tablet BID. Prenatal course was unremarkable.

At 21 weeks age of gestation, MRI Thoracic Aortogram (Figure 1) revealed a fusiform aneurysm in the descending aorta measuring 8.7 x 7.2 cm and extends over a distance of 11.1 cm, (de Bakey classification III or Stanford type B), Aortic dissection with re-entry of the false lumen at the level of the aneurysm, minimal pericardial effusion, minimal bilateral pleural fluid and adult polycystic kidneys (Figure 2). A multidisciplinary meeting was called to discuss treatment options and surgical planning. TEVAR or thoracic endovascular aortic repair was ultimately recommended.

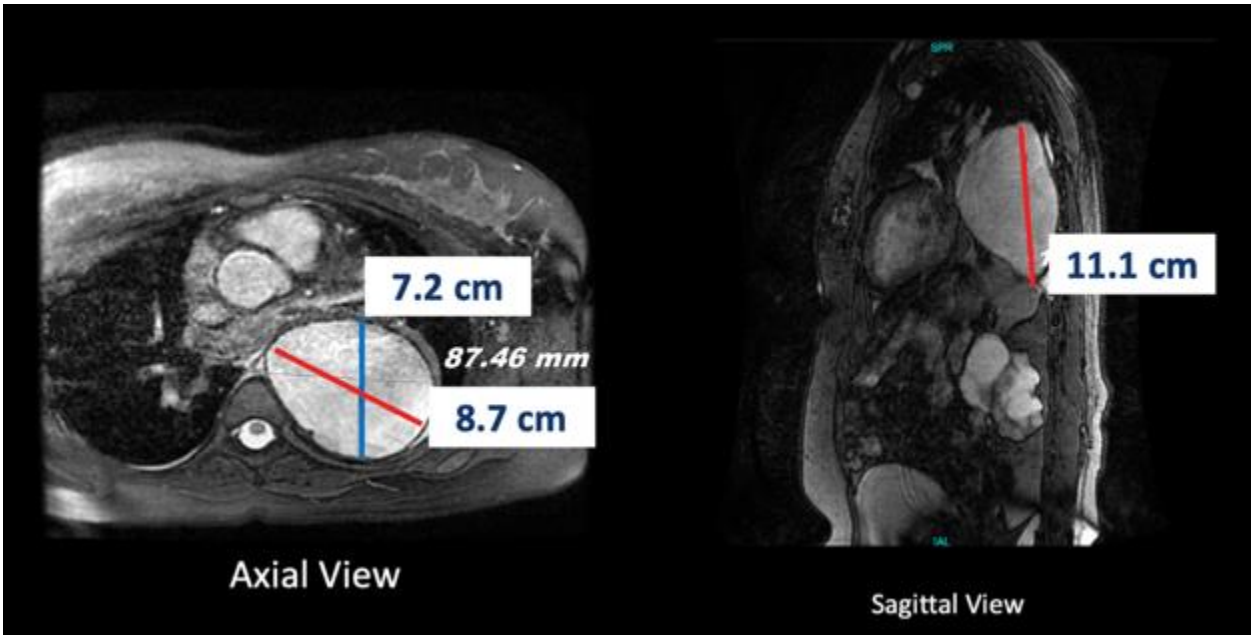


Figure 1. MRI Thoracic Aortogram showing a fusiform Stanford Type B aneurysm in the descending thoracic aorta measuring 8.7 x 7.2 cm and extends over a distance of 11.1 cm.

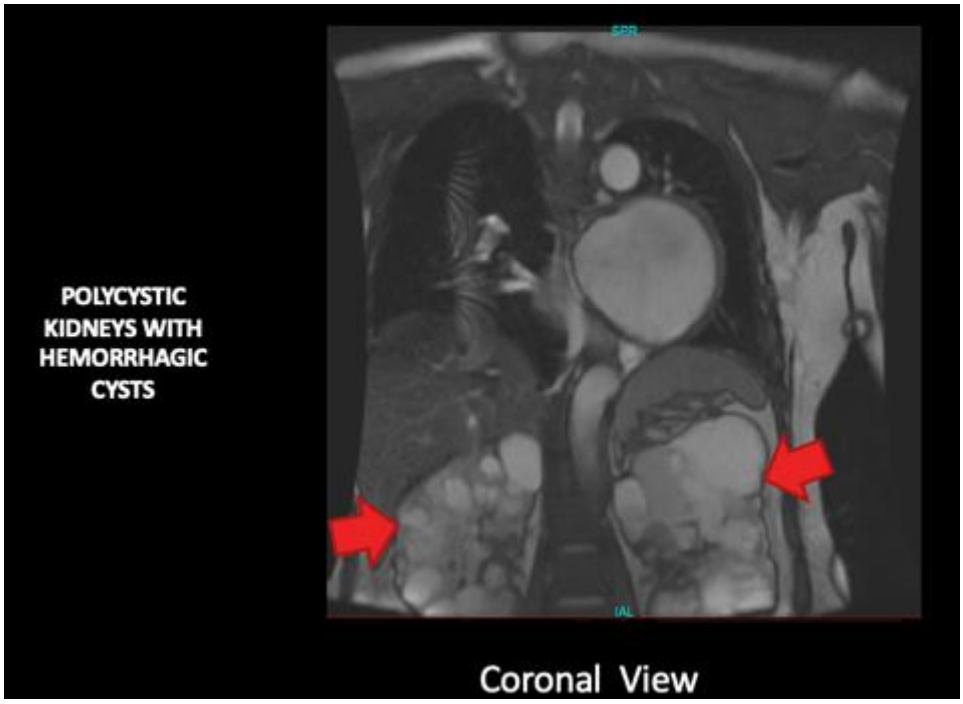


Figure 2. MRI Thoracic Aortogram showing bilateral polycystic kidneys.

The patient in this case lacked the typical risk factors for aortic dissection such as Marfan's disease, Ehlers-Danlos syndrome, Bi-cuspid aorta and Loeys-Dietz syndrome.
Pertinent physical examination finding included a normal arm span/height ratio, and with negative hyper extensibility of joints. The thumb test for Marfan syndrome was also negative (Figure 3).


Figure 3. Negative thumb test.

In the best interest of the patient (as well as logistics and since at that time that the case was accepted by our MFM Section, it was still at the height of the COVID-19 pandemic), she was admitted at 22 weeks and 5 days age of gestation. The patient was seen at the ER awake, comfortable, with normal vital signs. FHT was 148 bpm at the right lower quadrant.

Covid RT PCR swab was requested and showed negative results. She was then referred and co-managed by the following departments: IM Cardiology, IM Nephrology, Surgery TCVS, Surgery Urology, Pediatric Neonatology, Famed Palliative care and Anesthesia. Plan of the MFM service at this time was for maternal and fetal monitoring, and to standby intraoperatively during TEVAR procedure, followed by postoperative assessment of the mother and the fetus. If with no untoward circumstances after the procedure, long term plan was for primary cesarean section at 37 weeks age of gestation. Bilateral tubal ligation was advised but both the patient and her husband did not give their consents.

Pertinent laboratory results showed mild anemia of 108 mg/dl. Congenital anomaly scan showed no structural abnormalities. The patient was given with the following meds: Multivitamins, Iron and Calcium supplements. Methyldopa 2 tablets every 6 hours and Bisoprolol 2.5 mg tab OD were also given.

CT Aortogram showed Saccular Thoracic Aneurysm, with no dissection of leaking demonstrated. Minimal (Hypodense) pleural effusion, left; bilateral polycystic kidneys, gravid uterus with fetus in breech position. Additional laboratory tests were requested: 2D echo and lipid profile which showed normal results. Hypokalemia, hypomagnesemia and hypocalcemia were noted and subsequently corrected.

Due to the delay and numerous COVID-19 imposed restrictions in the transport of the TEVAR materials from abroad, it was decided that her admission is extended until the said materials arrived.

On the 10th hospital day, BP was maintained at 110/80-120/90 mmHg but there was note of increased in cardiac rate, hence she was started on Digoxin 0.25 mg ½ tab OD PO. And eventually, Verapamil was added. Four doses of dexamethasone were eventually given and completed at 25 4/7 weeks gestation. Course in the ward was unremarkable.

On the 39th hospital day at 28 weeks AOG, the TEVAR stent device arrived, and she was prepared for surgery. Fetal monitoring prior to the operation was done which showed a reactive NST. The patient was placed on general endotracheal anesthesia. Thoracic endovascular aortic repair was done. The TEVAR stent placement was uneventful, with no leakage intraoperatively (Figure 4 and 5).

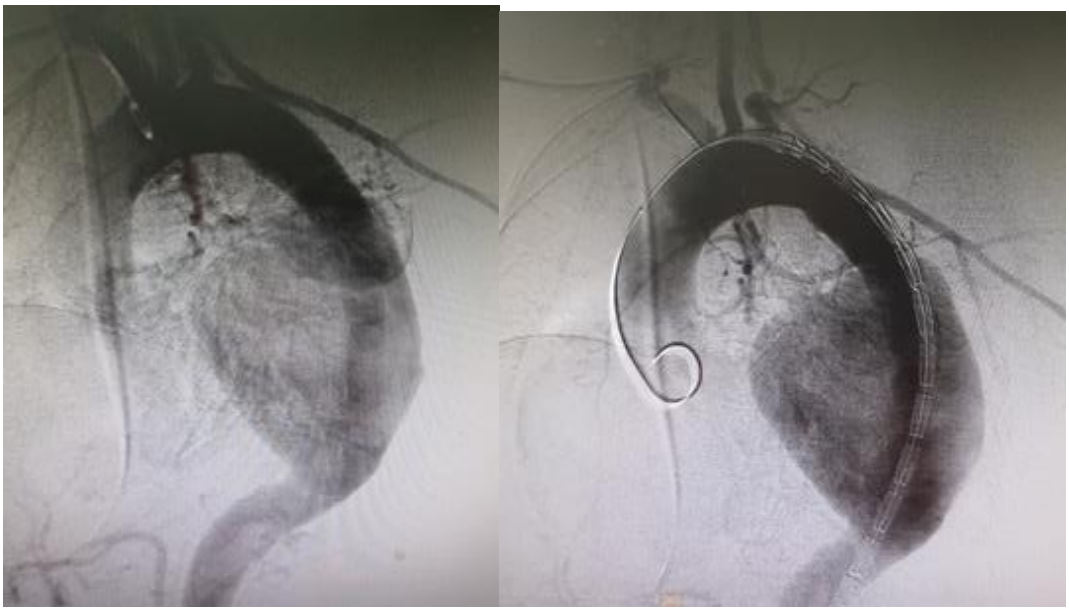


Figure 4. Aortic dissection (3A- before TEVAR stent placement; 3B- during TEVAR stent placement)



Figure 5. Post-TEVAR stent placement.

Immediately post op, fetal monitoring was done. Fetal bradycardia was noted, initially at 70-75 beats per minute. Resuscitation was done and oxygenation maintained via endotracheal tube. However, while ongoing extubation, there was note of edema of the upper extremities, face and proptosis of both eyes, hence airways were again secured via endotracheal tube. Diphenydramine IVTT was given, with impression of hypersensitivity reaction probably secondary to the contrast dye. Despite resuscitative measures, the fetal heart rate was at 80-90 beats per minute with minimal to absent variability. With these ominous findings, the decision was to do a stat cesarean section. Intraoperative findings were unremarkable and baby was delivered abdominally, via breech extraction with neonatal APGAR score of 5,7, birth weight of 891 grams (AGA) and Ballard score of 29 weeks (Figure 6). Initial assessment of the baby showed bradycardia of 110-120 beats per minute thus the baby was immediately transferred to NICU and prophylactic intubation was done (Figure 7). Surfactant was given at the 15th hour of life.



Figure 6. The neonate after Stat CS delivery.



Figure 7. The neonate at the NICU.

The patient stayed at the Surgical ICU for 4 days with no adverse complications and was eventually extubated on the second post operative day, and on the 4th post-op day, she was cleared by all services for transfer to the ward. During this time, the baby was still bradycardic at 100-115 bpm hence dopamine drip was started and maintained at the lowest setting and once the heart rate was maintained at 150-160 beats per minute it was eventually discontinued. CXR showed pneumothorax and CTT was inserted. On the 9th day of life, the CTT was removed, and baby was

extubated and maintained on oxygen supplementation via nasal cannula with no note of desaturations. Milk feeding via OGT was started.
The patient was discharged improved on the 11thpost-op day. Home medications were Aspirin 80 mg – 1 tablet OD, Clopidogrel 75 mg 1 tablet OD, Verapamil 80 mg 1 tablet every 6 hours, Bisoprolol 10 mg tablet OD, Clonidine 150 mg tablet every 6 hours.

3. DISCUSSION

Diagnosing aneurysm in pregnant women proves challenging especially if asymptomatic, but many cardiac disorders can be identified by taking a careful personal and family history, particularly cardiomyopathies, the Marfan syndrome, congenital heart disease to name a few. It is also important to ask specifically about possible sudden deaths in the family. A thorough physical examination considering the physiological changes that occur during pregnancy is mandatory, including auscultation for new murmurs, changes in murmurs, and looking for signs of heart failure.

The most commonly used classifications for aortic dissections are based on the affected anatomical portion. The Stanford group considers: type A (dissection involving ascending aorta) and type B (descending aorta only). Stanford Type A is the more common type, which accounts for 60% of aortic dissections and surgical management is recommended in these cases. On the other hand, Stanford Type B aneurysms accounts for 40% of dissections and is usually managed medically. In our patient, she was initially managed medically under optimized antihypertensive therapy with Methyldopa and Beta blockers, which eventually expanded during her admission to a quadruple regimen of Methyldopa, Verapamil, Bisoprolol and Digoxin.

The gold standard for aneurysm evaluation is computed tomography angiography (CTA), which allows precise diameter assessment and accurate preoperative planning. In pregnant patients, the effects of radiation on the fetus depend on the radiation dose and the gestational age at which exposure occurs. If possible, procedures should be delayed until at least the completion of the period of major organogenesis (12 weeks after menses). A CT scan has a 0.3 mGy radiation exposure to the fetus, and 7 mGy on the mother. There is no evidence of an increased fetal risk of congenital malformations, intellectual disability, growth restriction, or pregnancy loss at doses of radiation to the pregnant woman up to 50 mGy (5). In our case, a Chest/Thorax CT scan with contrast was initially done before her pregnancy and followed by MRI Thoracic aortogram at 21 weeks age of gestation. Both procedures allowed further diagnostic confirmation, as well as a better detailing of the dissection extent and involvement of the aortic vascular branches, fundamental data for the dissection classification and therapeutic planning.

In previously published cases and reviews of pregnancy-associated aortic dissections most cases are Stanford type A dissections, varying from 79% to 89%, with only 11% to 21% being type B dissections (6,7,8). For acute pregnancy-associated Type A dissections, open surgical repair is currently recommended and performed either before or after delivery, depending on the fetal gestational age. For pregnancy-associated Type B dissections, treatment recommendations are still not available. Based on previous reports, treatment with Type B dissection in pregnant women with either conservative or medication only, or surgical repair have mortality rates from 30.8% to 42.8% (6,9,10). Since the discovery and approval of newer treatment for aneurysm, such as the Thoracic endovascular aortic repair (TEVAR), it has now been included as an option in the management of Type B aortic dissection. Although there are no randomized controlled trials directly comparing TEVAR to open surgery, numerous studies suggest that TEVAR is associated with decreased morbidity compared with open repair (11,12,13,14,15).

According to the Gore TAG pivotal trial in 2015 (11), TEVAR is indicated for false lumen aneurysm with a maximum diameter of 55 mm or more. Increased risk of aneurysmal dilation and arterial rupture during pregnancy are well known and we therefore anticipated an increase of these risks in our patient because of the hemodynamic and endocrine changes during pregnancy and the puerperium. There are several areas of concern that we need to address if TEVAR is undertaken. One of this is the use of fluoroscopy during the said procedure in a pregnant patient and its effect on the fetus. The most vulnerable period of an exposed fetus to radiation for intellectual disability is at 8 to 15 weeks gestation but is significantly lesser in fetuses aged 16-25 weeks and negligible after 25 weeks AOG and if exposed to more than 50 mGy (16).

Based on the European Journal of Vascular and Endovascular Surgery, the minimum computed whole-body radiation of TEVAR is at 0.034 or 34 mGy (17), and since the procedure was planned at 22 weeks, but was done at 28 weeks age of gestation and does not exceed 50 mGy, the risk of the fetus having intellectual disability is negligible. And recent absolute risk estimates for fatal cancer risk for ages 0-15 year after in utero irradiation have been estimated to be 6% per Gy or 0.6% risk in our case. For the fatal cancer risk for the whole life span is 15% per Gy or 1.65% risk in our case.

By online search of previous published cases, we have come across only two reported cases of chronic aortic dissection in a pregnancy managed by TEVAR (1, 18), since cases like these as of 2016 has not yet been reported (19), and current guidelines for the treatment of aortic dissection do not provide recommendations for this difficult situation. Since we are presented with a patient still in her early second trimester on admission with plans for TEVAR procedure, fetal monitoring is of utmost importance. Patients with thoracic aneurysm have poor or decreased blood flow to the lower extremities and can result in hypoperfusion of the placenta and decreased blood flow to the fetus as well. This in turn can result to a growth restricted fetus, oligohydramnios, and even fetal death in utero. Fetal biometry every 2 weeks was done with intermitted FHT monitoring by Doppler every hour to optimize fetal monitoring in our patient. Weekly intrapartum monitoring was also recommended.

The type of delivery in patients with heart disease remains controversial. A vaginal delivery results in less bleeding, faster recovery, and decreases the risk of postoperative complications. In the absence of acute dissection, normal delivery can be performed with an aortic diameter < 40 mm in cases of Marfan Syndrome, with a cesarean section being indicated above this reference value (5). The carrying out of the delivery should include interventions to reduce aortic wall stress, strict blood pressure control, adequate analgesia, and instrumental delivery when indicated (1).

In our index patient, the aorta had a maximum diameter of 8.7 centimeters, which supported our choice of delivery by caesarean section. Bilateral tubal ligation is generally indicated as a definitive contraceptive method and was based on the high risk of maternal death in the event of a new pregnancy (20). But the patient and her husband did not consent to this form of contraception despite being advised and settled on barrier method as their choice of contraception.

For the timing of delivery, we planned to deliver the patient via elective caesarean section at 37 weeks if TEVAR proved to be a success. Given that the TEVAR was done at 28 weeks age of gestation, with latest pre-op estimated fetal weight of 878 grams by ultrasound, plan was then to monitor the baby intraoperatively and immediately postoperatively and to do emergency caesarean section anytime there is maternal or fetal indication. In our case, there was note of persistent fetal bradycardia with minimal to absent variability post operatively, hence a decision to do a stat caesarean section was made.

The vast diversity as well as the small number of cases reported in the literature do not allow establishing guidelines for the management of aortic dissection during pregnancy. Multidisciplinary management and individualized treatment aim to provide survival opportunities for the mother and fetus, considering maternal hemodynamic stability, fetal viability, and the best opportunity for surgical intervention, when indicated.

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