

Acute Aortic Dissection in a Pregnant Patient with Marfan Syndrome: A Case Presentation

Sara Villegas-Keech, MHS, PA-C

Background

Pregnancy-related maternal deaths are on the rise¹ with over 33% linked to cardiovascular disease, making it the leading cause of indirect maternal mortality.² Among the many types of cardiovascular conditions, the most common cause of aortic dissection in women of childbearing age is hereditary thoracic aortic disease, with Marfan Syndrome being the most prevalent underlying pathology.³

Given the complexity and severity of these cases, there is a growing need for multidisciplinary care teams to manage medically high-risk patients, improve outcomes, and reduce mortality. Over the past several decades, the establishment of cardio-obstetric care teams has been increasingly recommended. These specialized teams typically include high-risk obstetricians, cardiologists, anesthesiologists, intensivists, social workers, and other allied health professionals, all working collaboratively to provide comprehensive, life-saving care.

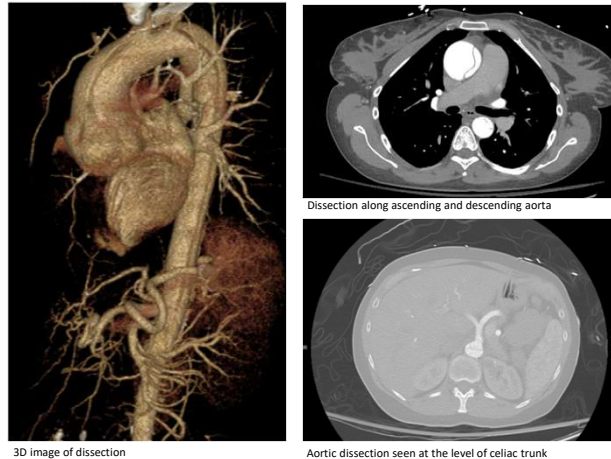
The case discussed today highlights the critical role such teams play in managing high-stakes, time-sensitive situations and the impact this coordinated approach can have on maternal and fetal survival.

Objectives

To highlight the risk of aortic dissection in pregnant patients with Marfan's Syndrome and the critical role of multidisciplinary management to improve outcomes.

Methods

A 41-year-old G7P2042 female with past medical history of Marfan's Syndrome with known heterozygous fibrillin-1 (FBN1) gene mutation, aortic root dilation measuring 55 mm, Borderline Personality Disorder, ADD, OCD, PTSD and housing insecurity presented to establish care for an unplanned but desired pregnancy. Surgical history included two prior cesarean deliveries and four dilation and curettages/evacuations. Medication included prenatal vitamins, low dose aspirin and metoprolol. During early pregnancy care, the multidisciplinary care team – including cardiology and Maternal Fetal Medicine – recommended a valve sparing aortic root repair to reduce the risk of aortic dissection in pregnancy but the patient declined due to risk of pregnancy loss. She was amenable to and was planning an antepartum admission for the start of the third trimester, for close observation until delivery in the case of an emergent cardiac event. However, prior to the planned admission, at 27 weeks and 5 days gestation, the patient presented via ambulance from her temporary living situation to a community hospital with severe back pain. Within 30 minutes of arriving at the Emergency Room, imaging confirmed a Stanford type A aortic dissection.



Results

3:34	Arrived at community hospital via ambulance from hotel
4:07	CT scan with type A dissection identified
4:19	Case discussed and accepted by tertiary care center
5:05	Arrives to tertiary hospital with blood transfusing
	Perfusing well, normal fetal heart tones
5:39	Surgical teams completed assessments
	Decision to move to operating room for urgent cesarean delivery and aortic repair is made
6:00	Arrive to Operating Room
	Intubated
	Central and arterial lines placed
7:13	Skin incision
7:19	Newborn is delivered via classical (vertical) cesarean section
8:30	Sternotomy and heparinization
	Closure of patent foramen ovale
	Hypothermic Circulatory Arrest/Antegrade Cerebral Perfusion Circulation
	Ascending Aortic Aneurysm repaired with 26 mm Dacron graft
	Transverse aortic arch graft
	Total arch replacement
	Elephant trunk procedure
	Arch debranching procedure, individual head vessel re-implantation (aorta to innominate, left common carotid artery bypass)
	Aortic valve replacement with mechanical aortic valve
	Aortic root replacement with a 32 mm Valsalva graft
	Re-implantation of coronary arteries
17:10	Chest closed by cardiac team
17:30	Abdomen closed by obstetric team
17:56	Procedure ends

The patient's post-operative recovery was complicated by acute pain, atrial fibrillation and heart failure with reduced ejection fraction responsive to medical optimization. She was discharged to temporary housing one month following surgery.

Results (continued)

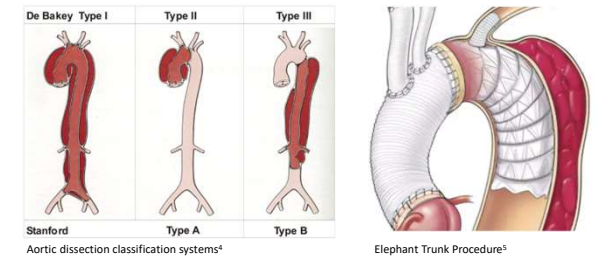


TABLE 1 Recommended Delivery Management for Aortopathy¹⁸

Condition	Aortic Diameter	Delivery Mode
Aorta ^a not significantly dilated	<4 cm	Vaginal ^{b1}
Aortic enlargement ^c	4-4.5 cm	Vaginal reasonable ^e
Syndromic (Marfan syndrome, Ehler-Danlos, Loeys-Dietz syndrome) or rs-HTAD	4-4.5 cm	Cesarean reasonable ^d
Aortic diameter	≥4.5 cm	Cesarean reasonable
Chronic aortic dissection	NA	Cesarean recommended
Acute type A dissection T1-T2		Urgent aortic surgery ^f
Acute type A dissection T3		Urgent cesarean delivery followed by aortic surgery
Acute type A dissection 24-28 wk		Individualized decision may be required
Type B dissection		Medical therapy with endovascular repair if possible and required. Cesarean mode delivery
Progressive aortic growth		Prophylactic aortic surgery may be considered.

American College of Cardiology expert panel recommendations for management of aortopathies in pregnancy⁶

Conclusion

This case underscores the importance of patient education, rapid diagnosis and coordinated multidisciplinary care in pregnant patients with connective tissue disorders or risk factors for aortic dissection. As maternal mortality rises, especially with cardiovascular disease as the leading cause, the need for multidisciplinary care teams and specialty cardio-obstetric care centers is growing. Diverse teams and care centers can provide early diagnosis and interventions that lead to favorable maternal and neonatal outcomes despite high-risk circumstances.

References

- 1) Maternal Mortality in Georgia & the U.S. By the Numbers. (n.d.). www.wabe.org/maternal-mortality-in-georgia-by-the-numbers
- 2) Nair, M., Nelson-Piercy, C., & Knight, M. (2017). Indirect maternal deaths: UK and global perspectives. *Obstetric Medicine*, 10(1), 10–15.
- 3) Curtis, S. L. (2025). Aortopathy in pregnancy: Unanswered questions. *JRSM Cardiovascular Disease*, 14, 1–8.
- 4) Nienaber, Christoph A., and Kim A. Eagle. "Aortic Dissection: New Frontiers in Diagnosis and Management." *Circulation*, vol. 108, no. 5, 5 Aug. 2003, pp. 638–635. <https://doi.org/10.1161/01.cir.0000087009.16755.e4>
- 5) <https://consultqd.clevelandclinic.org/simplified-frozen-elephant-trunk-repair-midterm-outcomes-underscore-safety>
- 6) Lau, Emily, and Niti Aggarwal. "Recommendations for the Management of High-Risk Cardiac Delivery." Nih.gov, Apr. 2024, pmc.ncbi.nlm.nih.gov/articles/PMC11198580/pdf/main.pdf. Accessed 15 July 2025.