ACTA MÉDICA PORTUGUESA
A Revista Científica da Ordem dos Médicos

Acute Aortic Dissection in the Third Trimester of Pregnancy as an Initial Presentation of Marfan Syndrome

Disseção Aguda da Aorta no Terceiro Trimestre de Gravidez Como Apresentação Inicial de Síndrome de Marfan

Marta REIS SANTOS✉[1], Alexandre VALENTIM-LOURENÇO[1], Maria PULIDO VALENTE[1]
Acta Med Port 2025 Aug;38(8):491-495 ▪ https://doi.org/10.20344/amp.22287

ABSTRACT

Aortic dissection is rare and more common in men. In women, it is more frequent during pregnancy, especially in the third trimester. We present the case of a 30-year-old pregnant woman diagnosed with type B aortic dissection at 29 weeks of gestation. Following the diagnosis, fetal maturation was initiated, and the patient was transferred to a tertiary care center. Due to uncontrolled hypertension and persistent pain, and after evaluating the maternal and fetal risk-benefit, a cesarean section was performed at 29 weeks and six days. During the postoperative period, a multi-drug regimen was required for blood pressure control, but the patient recovered and was discharged. Genetic testing, prompted by a family history of sudden death (brother) and aortic dissection (father), identified a mutation in the *FBN1* gene, confirming Marfan syndrome. This case highlights the importance of comprehensive patient history, the challenges of aortic dissection during pregnancy, and the need for a multidisciplinary approach in these cases.
Keywords: Aortic Aneurysm; Aortic Dissection; Marfan Syndrome; Pregnancy Complications, Cardiovascular; Pregnancy Trimester, Third

RESUMO

A dissecção aórtica é rara e mais comum em homens. Em mulheres é mais frequente na gravidez, especialmente no terceiro trimestre. Apresentamos o caso de uma grávida de 30 anos, com 29 semanas de gestação aquando do diagnóstico de uma disseção aórtica tipo B. Perante o diagnóstico, iniciou-se maturação fetal e a paciente foi transferida para um centro terciário. Face à hipertensão de difícil controlo e dor persistente, após avaliação do risco-benefício materno e fetal, foi realizada uma cesariana às 29 semanas e seis dias. No pós-operatório, foi necessário um regime terapêutico com múltiplos fármacos para controlo da tensão arterial, mas a paciente recuperou favoravelmente e teve alta. Testes genéticos, motivados pela história familiar de morte súbita (irmão) e dissecção aórtica (pai), revelaram uma mutação no gene *FBN1*, confirmando síndrome de Marfan. Este caso destaca a importância da história clínica, os desafios da dissecção aórtica na gravidez e a necessidade de uma abordagem multidisciplinar nestes casos.
Palavras-chave: Aneurisma da Aorta; Complicações Cardiovasculares da Gravidez; Disseção Aórtica; Terceiro Trimestre de Gravidez; Síndrome de Marfan

INTRODUCTION

Aortic dissection is rare, being more common in the male population.[1] In women, it becomes more prevalent during pregnancy, ranging from 5.5 to 14.5 per million *versus* 1.2 per million in non-pregnant women, with estimated rates of maternal and fetal mortality of 12% and 28%, respectively.[2-4] In pregnancy, the incidence is higher in the third trimester (50% of cases), followed by the post-partum period (33% of cases).[5,6]

Inherited connective tissue disorders further elevate the risk of dissection to 3% - 8%.[5,7] In these patients, pre-pregnancy risk stratification using the modified World Health Organization (WHO) classification[8] and close echocardiographic surveillance during pregnancy[9] are essential (Fig. 1), although dissection can sometimes be the first manifestation of the condition.

CASE REPORT

We present the case of a 30-year-old pregnant woman, originally from another country and living in Portugal for two years, with an obstetric history of two prior first-trimester miscarriages and a family history of sudden cardiac death (her brother, in his forties, who died in their home country with limited details available) and aortic dissection (her father, in his sixties, who survived after surgical treatment).

An echocardiogram performed a year prior to pregnancy revealed mitral valve prolapse and aortic root dilation (38.5 mm), but she was otherwise healthy and was not taking any regular medication. This echocardiogram was conducted during a consultation with her primary care physician. However, no further investigations were pursued, and her pregnancy was being monitored in a primary care setting. No genetic testing for this family had been performed.

The woman had been consistently normotensive before and during pregnancy, which had progressed uneventfully until the described events. At 29 weeks gestation, she experienced sudden chest pain radiating to her back. The evaluation revealed a normal electrocardiogram, cardiac enzymes, and chest X-ray, but a transthoracic echocardiogram raised suspicions of an aortic dissection. Considering the need to confirm the diagnosis and better characterize the condition, as well as the relatively low risk to the fetus, a computed tomography angiography (CTA) was promptly performed. The scan confirmed an uncomplicated acute type B aortic dissection (Fig. 2).

1. Department of Obstetrics, Gynecology and Reproductive Medicine. Hospital de Santa Maria. Unidade Local de Saúde de Santa Maria. Lisboa. Portugal.
✉ **Autor correspondente:** Marta Reis Santos. mrnsantos21@gmail.com
Recebido/*Received*: 09/09/2024 - **Aceite/*Accepted*:** 23/12/2024 - **Publicado *Online*/*Published Online*:** 26/03/2025 - **Publicado/*Published*:** 01/08/2025

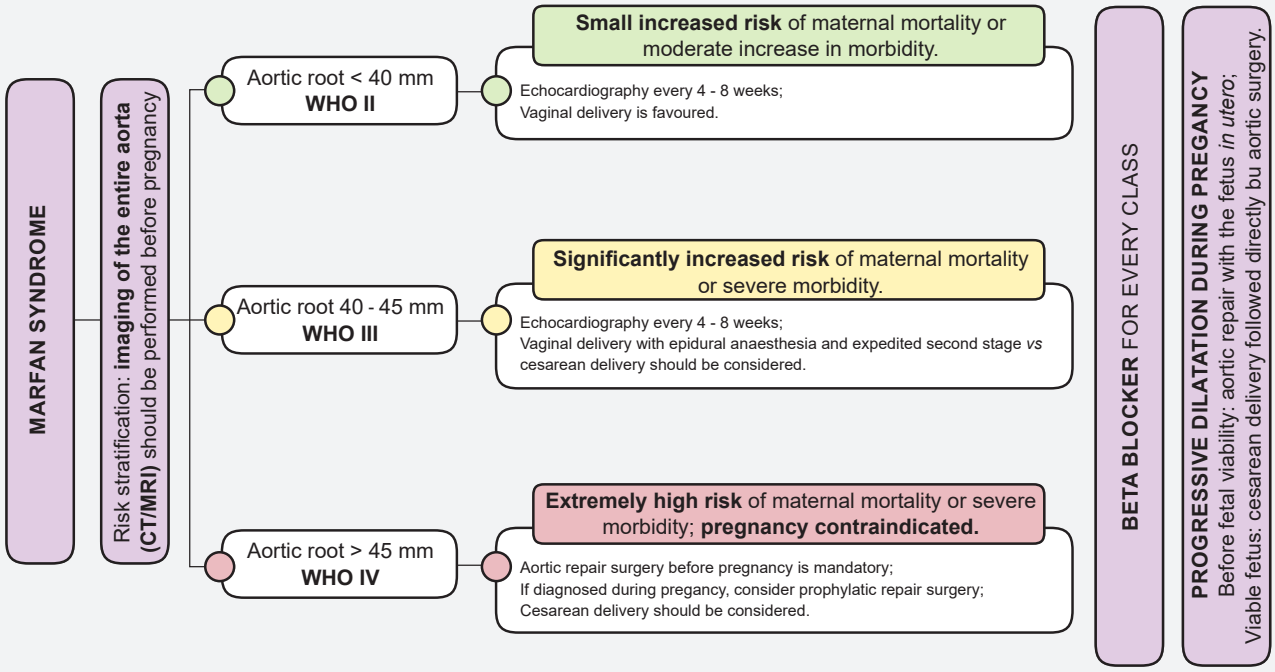

Figure 1 – Risk stratification based on the modified WHO classification of maternal cardiovascular risk and management strategies

The patient was transferred to a tertiary center, where fetal maturation was induced. She developed severe hypertension, requiring intravenous blood pressure control with labetalol and isosorbide dinitrate. Given the family history and current presentation, genetic testing was performed and revealed a mutation in the *FBN1* gene, confirming Marfan syndrome.

On the third day of hospitalization, due to persistent pain and hypertension — signs raising concern for impending hypoperfusion or aortic rupture — a multidisciplinary team comprising obstetrics, intensive care, vascular surgery, anesthesiology, and neonatology recommended a cesarean section at 29 weeks and six days, which the patient accepted.

Postpartum, the patient experienced worsening blood pressure control requiring combined intravenous therapy

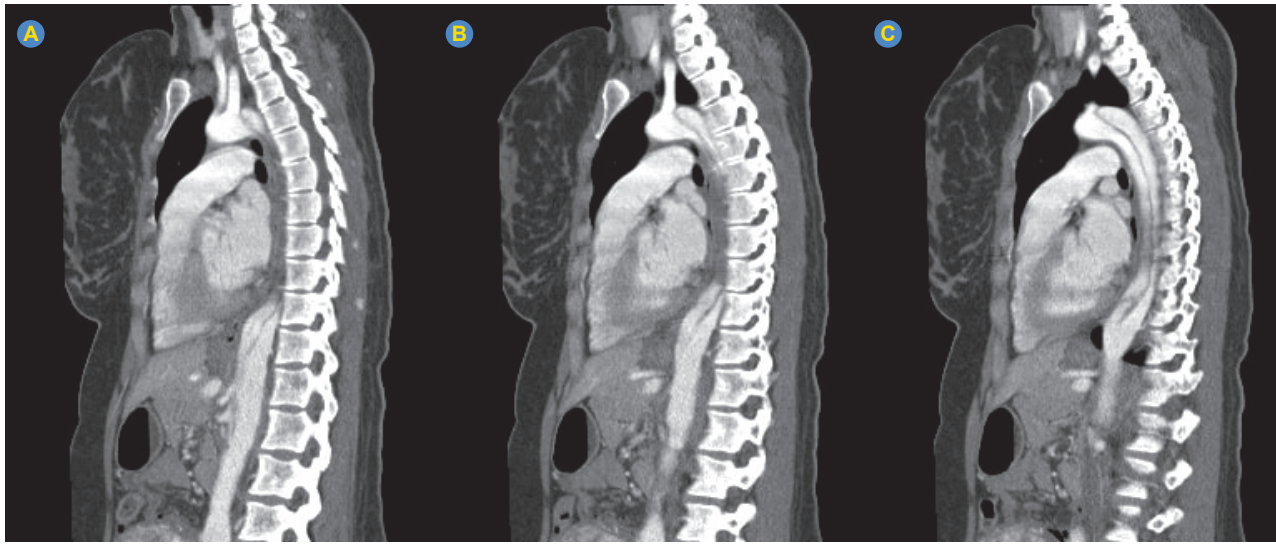

Figure 2 – Angio-CT scan of the patient at admission showing type B aortic dissection

with labetalol and isosorbide dinitrate and oral therapy with methyldopa, captopril, amiloride, hydrochlorothiazide, carvedilol and nifedipine. There were concerns that renal hypoperfusion could be causing such severe hypertension, but the reevaluation CTA did not confirm this hypothesis. Therefore, treatment was conservative, with no need for surgical intervention. Gradually, it became possible to titrate the antihypertensive medication and discontinue the intravenous treatment. She was discharged on day 20 postpartum on losartan 50 mg id, carvedilol 25 mg bid and acetylsalicylic acid 100 mg id.

However, the patient presented again on day 27 postpartum with new onset neck pain. The diagnostic tests revealed a progression of the dissection to the left carotid artery. The patient acknowledged poor adherence to antihypertensive therapy, which may have contributed to this complication. Following admission for conservative treatment and surveillance, she was discharged on day 40 postpartum, with the addition of spironolactone 12.5 mg once daily to her previously prescribed medicines.

The newborn required non-invasive ventilation and surfactant therapy but improved steadily, transitioning to ambient air by day 26. The neonate was discharged on day 55 postpartum. At this time, the patient had fully recovered obstetrically and was successfully breastfeeding.

DISCUSSION

Aortic dissection in pregnancy

In pregnant women, aortic dissection accounts for high rates of maternal and fetal mortality (12% and 28%, respectively). However, the rarity of this condition (14.5 per million pregnant women) creates considerable gaps in understanding how to prevent, manage, and monitor patients at risk for pregnancy-associated aortic dissection or those of childbearing age who have experienced an aortic dissection.[4]

Pregnancy itself is a risk factor for aortic dissection, with studies reporting a 23-fold increased risk during pregnancy.[4] This is mostly due to hormonal (elevated estrogen and progesterone levels disrupt elastic fibers, increase MMP-2 activity, and reduce elastin deposition, weakening the aortic wall) and hemodynamic changes (increased circulating volume and cardiac output leading to increased hemodynamic stress on the aortic wall).[4,9] Inherited connective tissue disorders such as Marfan syndrome and Loeys-Dietz syndrome are significant risk factors. Additional risk factors include smoking, hypertension, cocaine use, and autoimmune diseases like Takayasu's arteritis.[6,10]

Diagnosing or excluding aortic dissection relies heavily on clinical suspicion, as it is a rare condition and may not be an immediate consideration in the differential diagnosis of chest pain during pregnancy, where more common causes are often prioritized.[4] The initial risk assessment can be categorized using the Aortic Dissection Detection Risk Score.[6,8,11] Beyond the clinical risk score, a series of initial simple tests are usually conducted, including an electrocardiogram, chest X-ray, transthoracic echocardiogram (TTE), and cardiac biomarkers.[12] They help rule out other conditions such as pulmonary embolism, myocardial infarction or pericarditis, and TTE can potentially identify aortic dissection by revealing an aortic intimal flap, aortic wall thickening or dilation, significant aortic valve regurgitation with or without cusp prolapse, and pericardial effusion. However, a CTA scan of the entire aorta is the primary imaging method for confirming aortic dissection.[12] This test should not be delayed when clinically indicated based on concerns for fetal harm, as their risks are minimal and much lesser compared to maternal risk in this setting.[13] Also, the CTA is important for accurate classification of the type of aortic dissection, as management strategies differ significantly based on whether the ascending aorta is involved.

Managing aortic dissection during pregnancy requires a multidisciplinary team and specialized care. Initial treatment focuses on reducing pulse pressure and heart rate to minimize aortic wall stress and prevent complications like rupture or malperfusion. Intravenous beta-blockers, particularly labetalol, are the preferred choice. For type A dissections, where surgical treatment is always required, cesarean delivery is performed before aortic repair if the fetus is viable; otherwise, surgery proceeds with the fetus *in utero*. In uncomplicated type B dissections, conservative medical management with close monitoring of both mother and fetus is advised.[6]

Marfan syndrome and pregnancy

Marfan syndrome, caused by *FBN1* mutations, increases susceptibility to aortic aneurysm and dissection. In this case, there was no prior diagnosis of Marfan syndrome. The fact that the patient and her family were originally from another country may have hindered the suspicion of Marfan syndrome, as details regarding the clinical history of her family members – particularly her brother – were not readily accessible.

When the diagnosis is known, women with Marfan syndrome require pre-pregnancy risk stratification using the modified WHO classification,[8] with close monitoring during pregnancy. Beta-blocker therapy, although controversial in the past,[14] may slow aortic dilation, with recent studies showing reduced aortic root growth in pregnant women with Marfan syndrome.[15] Though evidence is not conclusive, the European Society of Cardiology (ESC) recommends beta-blockers to prevent dissection in pregnancy.[8] Highlights of the surveillance of pregnancy in Marfan syndrome are summarized in Fig. 1.

Multidisciplinary management

This case highlights the importance of a multidisciplinary approach involving obstetrics, intensive care, vascular surgery, anesthesiology, neonatology, and genetics. Timely blood pressure control and fetal monitoring allowed for successful delivery at 29 weeks + six days, balancing maternal risks and fetal maturity. Postpartum, coordinated care was critical to managing the patient's hypertension and preventing further complications.

Challenges in blood pressure control

Postpartum hypertension presented considerable challenges. The immediate postpartum period is characterized by significant physiological changes, including uterine contraction, relief of aortocaval compression, and catecholamine release, leading to a 60% - 80% increase in cardiac output.[16] Additionally, the reabsorption of third-space fluid during this phase may have further contributed to elevated blood pressure.[17] In a later stage, the patient's confirmed poor adherence to outpatient antihypertensive therapy may have contributed to the progression of the dissection into the left carotid artery. This highlights the critical importance of strict adherence to therapy and patient education in preventing complications in individuals with Marfan syndrome.

Neonatal outcomes

The premature neonate required initial ventilatory support and surfactant therapy but steadily improved, being discharged without significant complications. The infant's progression reflects advances in neonatal care for preterm infants born to high-risk mothers. At the time of this report, the child was healthy and attending follow-up pediatric appointments at the hospital. Genetic testing for Marfan syndrome was planned within the first year of life to determine the need for further evaluations.

Implications for future pregnancies

Patients with a history of aortic dissection, especially with Marfan syndrome, face a high risk of recurrence in future pregnancies. According to ESC guidelines, women with type B dissection should be advised against pregnancy.[8] If pregnancy occurs, it should be managed in a specialized center with strict blood pressure control and regular cardiovascular imaging.

CONCLUSION

This case underscores the critical importance of early diagnosis, multidisciplinary care, and strict adherence to therapy in managing aortic dissection during pregnancy, especially in women with Marfan syndrome. The positive outcomes for both mother and child highlight the effectiveness of coordinated care, and the necessity of vigilant monitoring for future pregnancies. Strict outpatient follow-up and patient education are vital to prevent recurrence and manage complications.

ACKNOWLEDGMENTS

We express our sincere gratitude to the Intensive Care Department, the Vascular Surgery Department, the Anesthesiology Department, the Genetics Department, and the Neonatology Department of ULS Santa Maria for their critical contributions to the management of this case. Their expertise and commitment were pivotal in addressing the challenges presented, and their clinical and scientific support was of the highest standard.

AUTHOR CONTRIBUTIONS

MRS: Data acquisition, analysis and interpretation, writing and critical review of the manuscript.

AVL, MPV: Data analysis and interpretation, critical review of the manuscript.

All authors approved the final version to be published.

PROTECTION OF HUMANS AND ANIMALS

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the Helsinki Declaration of the World Medical Association updated in October 2024.

DATA CONFIDENTIALITY

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

PATIENT CONSENT

Obtained.

COMPETING INTERESTS

The authors have declared that no competing interests exist.

FUNDING SOURCES

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

REFERENCES

1. Bossone E, Carbone A, Eagle KA. Gender differences in acute aortic dissection. J Pers Med. 2022;12:1148.
2. Yuan S. Aortic dissection during pregnancy: a difficult clinical scenario. Clin Cardiol. 2013;36:576-584.

3. Meng X, Han J, Wang L, Wu Q. Aortic dissection during pregnancy and postpartum. J Card Surg. 2021;36:2510-7.
4. Russo M, Boehler-Tatman M, Albright C, David C, Kennedy L, Roberts AW, et al. Aortic dissection in pregnancy and the postpartum period. Semin Vasc Surg. 2022;35:60-8.
5. Rimmer L, Heyward-Chaplin J, South M, Gouda M, Bashir M. Acute aortic dissection during pregnancy: trials and tribulations. J Card Surg. 2021;36:1799-805.
6. Mazzolai L, Teixido-Tura G, Lanzi S, Boc V, Bossone E, Brodmann M, et al. 2024 ESC guidelines for the management of peripheral arterial and aortic diseases. Eur Heart J. 2024;45:3538-700.
7. McNally E. Aortic dissection with pregnancy—anticipating prepartum and postpartum risk. JAMA Cardiol. 2021;6:66-7.
8. Regitz-Zagrosek V, Roos-Hesselink JW, Bauersachs J, Blomström-Lundqvist C, Cífková R, De Bonis M, et al. 2018 ESC Guidelines for the management of cardiovascular diseases during pregnancy. Eur Heart J. 2018;39:3165-241.
9. Smith K, Gros B. Pregnancy-related acute aortic dissection in Marfan syndrome: a review of the literature. Congenit Heart Dis. 2017;12:251-60.
10. Kano HD, Sun W. Risk factors for aortic dissection. US Cardiol. 2023;17:e17.
11. Rogers AM, Hermann LK, Booher AM, Nienaber CA, Williams DM, Kazerooni EA, et al. Sensitivity of the aortic dissection detection risk score, a novel guideline-based tool for identification of acute aortic dissection at initial presentation. Circulation. 2011;123:2213-8.
12. Czerny M, Grabenwöger M, Berger T, Aboyans V, Della Corte A, Chen EP, et al. EACTS/STS guidelines for diagnosing and treating acute and chronic syndromes of the aortic organ. Ann Thorac Surg. 2024;118:5-115.
13. Committee Opinion No. 723: guidelines for diagnostic imaging during pregnancy and lactation. Obstet Gynecol. 2017;130:e210-6.
14. Gersony DR, McClaughlin MA, Jin Z, Gersony WM. The effect of beta-blocker therapy on clinical outcome in patients with Marfan's syndrome: a meta-analysis. Int J Cardiol. 2007;114:303-8.
15. Roberts EA, Pistner A, Osobamiro O, Banning S, Shalhub S, Albright C, et al. Beta-blocker use during pregnancy correlates with less aortic root dilatation in patients with Marfan's syndrome. Aorta. 2023;11:63-70.
16. Guimarães T, Magalhães A, Veiga A, Fiuza M, Ávila W, Pinto FJ. Cardiopatia e gravidez – o estado da arte. Rev Port Cardiol. 2019;38:373-83.
17. Sibai BM. Etiology and management of postpartum hypertension-preeclampsia. Am J Obstet Gynecol. 2012;206:470-5.