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Case Report
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"Through the ears of the needle" – secondary hypertension in young women

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Abstract

We present the case of a 37-year-old female patient who was admitted on an outpatient basis for oscillating blood pressure values accompanied by headache and intermittent vertigo. At the time of admission, she was hemodynamically stable. The cardiac auscultation highlighted an apexian systolic murmur (grade 3/6) and as well as a systolic murmur (grade 2/6) under the scapula. The echocardiography revealed a progressive narrowing of the lumen in the proximal descending aorta. Chest radiography outlined rib notches at the level of rib arches 3–9 and a pathognomonic image of "3" secondary to pre- and post-stenotic dilations. The computed tomography scan identified progressive narrowing of the lumen with a minimum caliber of 1.7 mm at the insertion site of the *ligamentum arteriosum* in the proximal descending aorta and significant collateral circulation.

Keywords: coarctation of the aorta, pregnancy, hypertension, HEART team.

Introduction

Coarctation of the aorta (CoA) is diagnosed in 20% of cases in adulthood, often through associated complications. Aortic dissection, hemorrhagic stroke, heart failure, and infective endocarditis are just some of the entities with a poor prognosis for pregnant women. Abortion secondary to placental ischemia is the main fetal complication cited in the literature. Elevated estrogen levels are associated with the remodeling of the aortic walls, which predisposes to acute aortic syndromes.

Case report

We present the case of a 37-year-old female patient who was admitted on an outpatient basis for oscillating blood pressure values accompanied by headache and intermittent vertigo. The patient had a

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previous miscarriage at 5 months without a significant medical history. She was a smoker and did not follow any medical treatment at home. The cardiology evaluation was part of the pre-established assessment as the patient wanted a new pregnancy.

At the time of admission, she was hemodynamically stable, with normal oxygen saturation (94%), normal respiratory rate, and normal body temperature. The patient had a regular heart rate of 75 beats per minute (bpm), high blood pressure at the level of the upper extremities (155/90 mmHg in the left upper limb and 159/88 mmHg in the right upper limb), and normal blood pressure at the level of the lower extremities (124/71 mmHg in the lower left limb and 119/69 in the lower right limb). The clinical examination revealed pale mucous membranes. The cardiac auscultation highlighted an apexian systolic murmur (grade 3/6) and as well as a systolic murmur (grade 2/6) under the scapula. Peripheral pulse was diminished in the right femoral artery, and hydrostatic varicose veins were observed bilaterally.

The electrocardiogram showed sinus rhythm with a heart rate of 60 bpm, intermediate axis, and left ventricular hypertrophy (Figure 1).

The laboratory tests indicated inflammatory syndrome (C-reactive protein – 8.5 mg/dl), iron deficiency anemia (hemoglobin – 11.8 g/dl, serum iron – 19 µg/dl), hypercholesterolemia (total cholesterol – 285 mg/dl, low-density lipoprotein cholesterol – 157 mg/dl and triglycerides – 177 mg/dl) and hyperglycemia (fasting blood glucose – 138 mg/dl).

Echocardiography revealed concentric left ventricular hypertrophy with an interventricular septum and left ventricular posterior wall thickness of 13 mm) as illustrated in Figure 2. The systolic function evaluated with the biplane Simpson's method was normal (left ventricular ejection fraction of 55%), and no cardiac wall motion abnormalities were observed. The suprasternal view allowed us to measure the aortic arch, aortic arch branches, and descending aorta, and we identified a progressive narrowing of the lumen in the proximal descending aorta (distal from the origin of the left subclavian artery). Color Doppler and continuous wave Doppler highlighted the presence of a mild mitral regurgitation and a blood flow in the descending thoracic aorta with a maximum velocity of 3.1 m/s at the level of the narrowing (Figure 2).

Taking into account the clinical and echocardiographic findings suggestive of the presence of CoA, we performed chest radiography which outlined rib notching at the level of rib arches 3–9 and a pathognomonic image of "3" secondary to pre- and post-stenotic dilations.

The computed tomography scan confirmed our clinical suspicion of diagnosis by identifying progressive narrowing of the lumen with a minimum caliber of 1.7 mm at the insertion site of the *ligamentum arteriosum* (21 mm from the origin of the

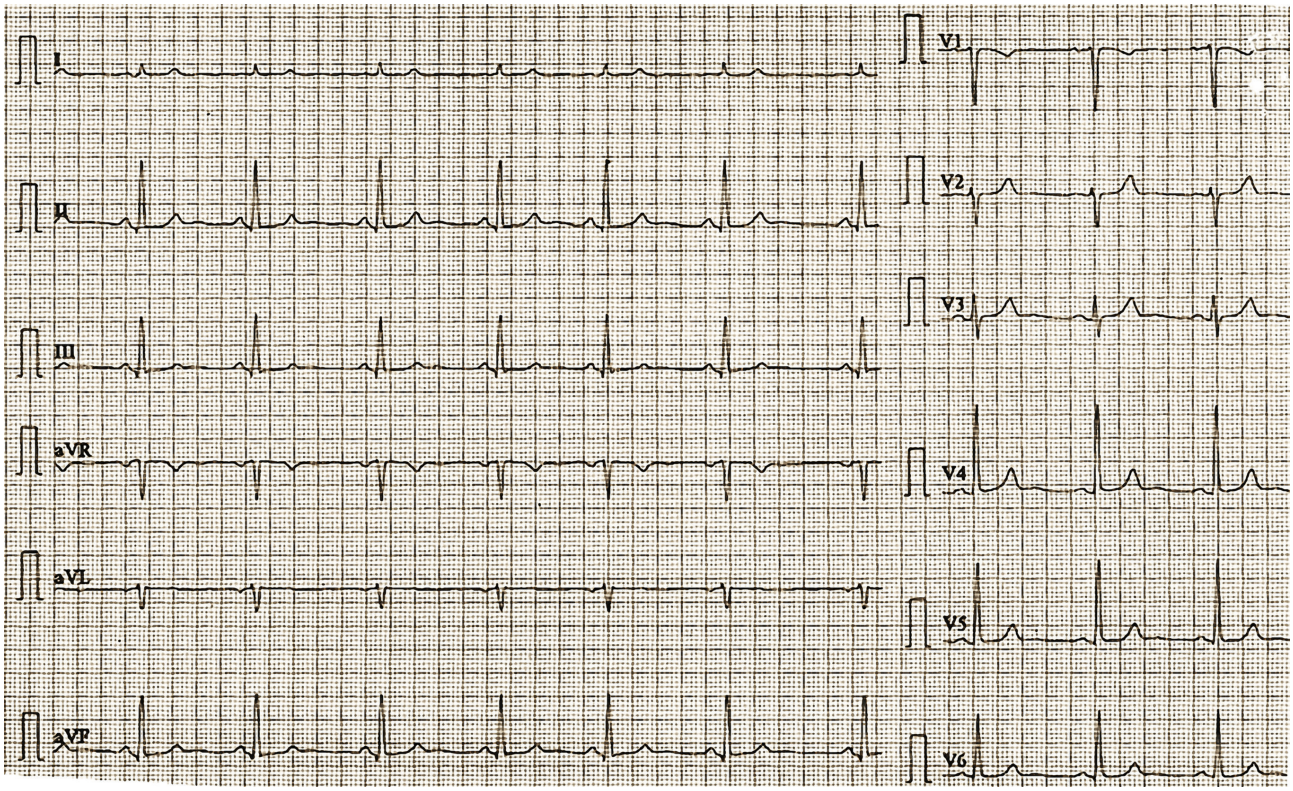

Figure 1. Electrocardiogram showing sinus rhythm with a heart rate of 60 beats per minute (bpm), intermediate axis and left ventricular hypertrophy.

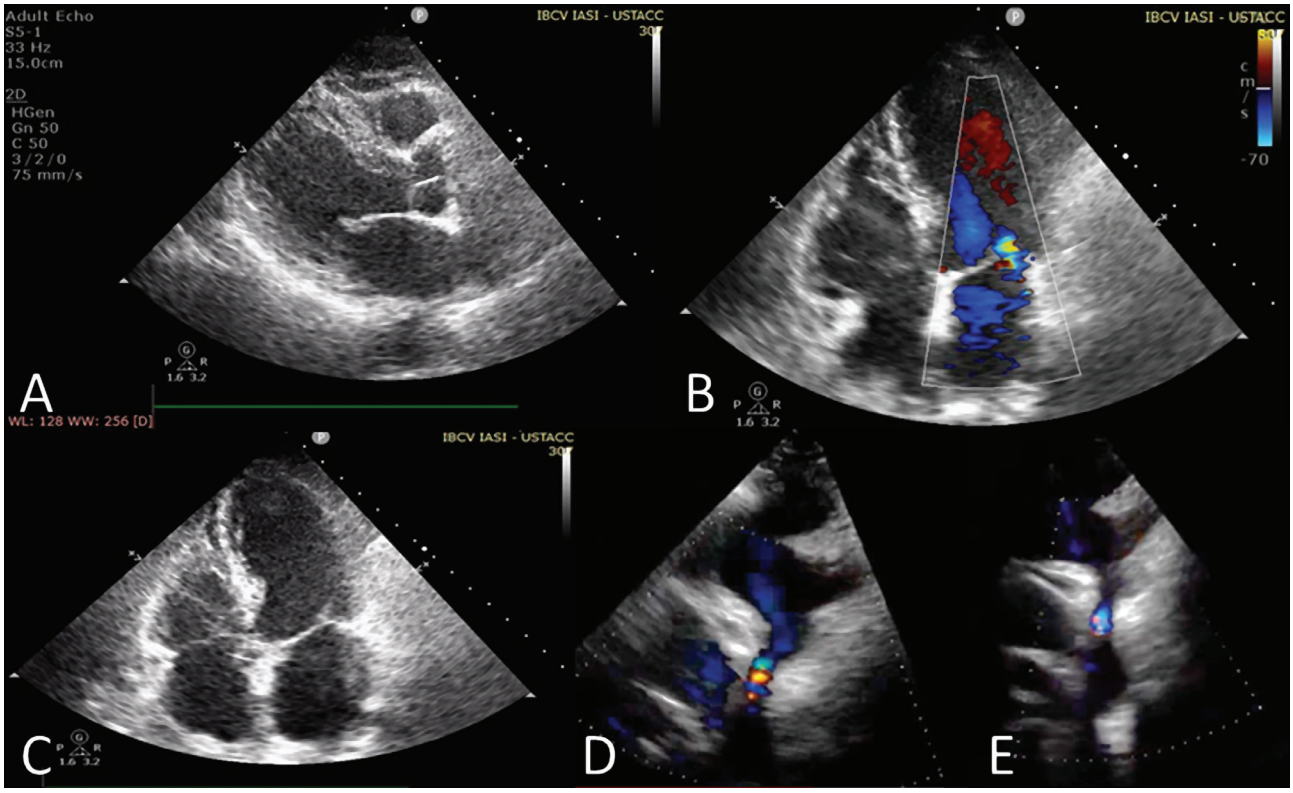

Figure 2. Echocardiography. A – parasternal long axis view. Left ventricular hypertrophy without left ventricular enlargement; B – Apical 4-chamber (A4) view; C – Mild mitral regurgitation; D, E – Suprasternal view - progressive narrowing of the lumen in the proximal descending aorta.

left subclavian artery) in the proximal descending aorta and significant collateral circulation, with dilatation of intercostal and internal mammary arteries up to 6 mm (Figure 3).

After being explained the treatment options, with the associated risks and benefits, the patient refused both interventional and surgical treatment. Amlodipine 10 mg/day was started for blood pressure control.

Discussion

Congenital heart disease affects approximately 1% of the global population [1]. CoA is a secondary cause of hypertension, usually discovered in early childhood. With a 6–8% prevalence in women with congenital heart disease, this condition can be challenging if it is a pregnancy discovery [2, 3].

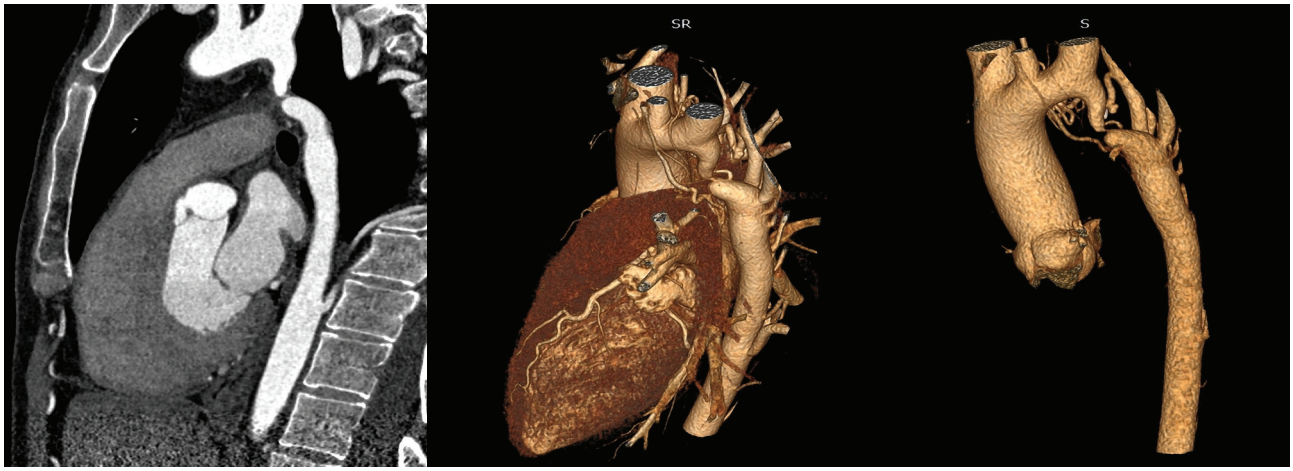

Figure 3. Computed tomography scan – 3D reconstruction. CoA in the proximal descending aorta and significant collateral circulation.

Pregnant women with CoA have moderate cardiovascular and obstetric risk and are classified as mWHO II-III according to the modified World Health Organization (mWHO) classification [3]. With a cardiac event risk of 10–19% during pregnancy and an increased risk in terms of mortality and morbidity, these patients require close cardiological supervision and control of all associated risk factors [4].

The presence of pregnancy in patients with CoA is associated with both maternal and fetal risks. Although hypertensive disorders of pregnancy are not more prevalent than in the general population, uncontrolled blood pressure values can lead to the neonatal appearance of growth retardation, *abruptio placentae*, and premature birth. Renal failure and hypertensive crisis are maternal complications with prognostic values [5–7].

Blood pressure control decreases the risk of aortic rupture or dissection by reducing hemodynamic stress. Physiological (variance of blood pressure levels throughout pregnancy) or pathophysiological changes include coarctation-associated aortopathy, hormonally mediated vessel wall weakness, and increased thrombogenicity [8]. In the middle trimester of pregnancy, the blood pressure levels have a decreasing tendency towards normal levels at preterm. Pregnancy is also associated with increased plasma volume, heart rate, stroke volume, cardiac output, and decreased systemic vascular resistance.

Before pregnancy, patients with CoA require extensive cardiology examination to identify associated anomalies. Intracranial aneurysms are more frequent in these patients, and therefore, blood pressure control becomes imperative to prevent hemorrhagic complications. Stenotic bicuspid aortic valve or endocarditis have hemodynamically significance at the time of delivery. Young girls diagnosed with CoA should also be investigated for Turner syndrome [2]. The optimal choice of antihypertensive medication can be challenging as some of the most used antihypertensive agents are teratogenic.

In terms of obstetrical outcomes, patients with CoA are advised to choose vaginal delivery, with indications for cesarean delivery in case of significant systemic hypertension, a dilated aorta, or significant residual coarctation. Unrepaired CoA is also associated with higher cesarean section rates, low birth weight, preterm delivery, or postpartum hemorrhage. Cesarean sections have a negative or at least neutral effect as they do not offer maternal protection with the cost of an increased adverse fetal outcome [9, 10].

Ramlakhan *et al.* demonstrated in a study of 303 women with CoA that there are various predictors of cardiac events, such as moderately abnormal systolic dysfunction, previous heart failure, a New York Heart Association (NYHA) class of at least II, and the chronic use of cardiac drugs before pregnancy. Women from undeveloped countries have more comorbidities, a higher rate of pre-pregnancy cardiac morbidity, as well as higher rates of unrepaired CoA [4, 11, 12].

The ZAHARA II study demonstrated that uteroplacental Doppler flow (UDF) parameters were altered in women with congenital heart diseases and correlated with maternal cardiac function before pregnancy [11, 12].

Utero-placental resistance influences both left and right ventricular dysfunction. Siegmund *et al.* demonstrated that right ventricle function correlates with impaired UDF parameters. It was previously demonstrated that there is a statistically significant association between function of the right ventricle and UDF parameters in women with congenital heart diseases or right-sided heart disease. Siegmund *et al.* demonstrated that the value of tricuspid annular plane systolic excursion (TAPSE) at 32 weeks of gestation in women with CoA was lower compared to that of healthy pregnant women. The decreasing values of TAPSE between 20 and 32 weeks indicated subclinical right ventricle dysfunction, which may raise the question of a prior subclinical dysfunction with uncertain mechanisms until now. The interaction between the left and right ventricle systolic function is explained by the increased left ventricle mass and afterload persistence after successful repair of CoA [12, 13].

The therapeutic management of patients with CoA is controversial, especially during pregnancy. The main concern for these patients is the tissue integrity of the para-coarctation region. The choice of percutaneous catheter intervention or surgical repair must be made by the HEART team, including the cardiologist, interventional cardiologist, cardiovascular surgeon, and obstetrician. During pregnancy, women are advised to choose surgical repair with excision of the para-coarctation tissue, leaving interventional procedures in cases of recurrence (up to 35% of cases). Balloon angioplasty can be used as an alternative to surgical repair in case of discrete narrowing. After surgical or interventional correction, patients should have a lifelong follow-up, including echocardiography, to identify aortic dilatation or onset aneurysm. In terms of medical management, thiazides diuretics and angiotensin-converting enzyme inhibitors are ruled out due to the teratogenic effect. Labetalol is a non-selective beta-blocker that can be successfully prescribed to control blood pressure during pregnancy, with no effect on the uteroplacental blood flow [14–16].

Conclusion

CoA can be challenging when discovered during pregnancy, with significant cardiovascular, maternal and fetal risks. In case of an unrepaired CoA,

the management should be discussed within the HEART team. Controlling blood pressure levels decreases the risk of aortic rupture or dissection. We presented the case of a 37-year-old female who has a poor obstetrical prognosis after refusing interventional or surgical treatment.

Conflict of Interest

The authors confirm that there are no conflicts of interest.

References

1. Wadey CA, Weston ME, Dorobantu DM, Pieles GE, Stuart G, Barker AR, Taylor RS, Williams CA. The role of cardiopulmonary exercise testing in predicting mortality and morbidity in people with congenital heart disease: a systematic review and meta-analysis. Eur J Prev Cardiol. 2021 Aug 18:zwab125. doi: 10.1093/eurjpc/zwab125. Epub ahead of print. PMID: 34405863.
2. Beckman RH III. Coarctation of the aorta. In: Allen HD, Gutgesell HP, Clark EB, Driscoll DJ, editors. Moss and Adams' Heart Disease in Infants, Children and Adolescents: Including the Fetus and Young Adult. Vol. 2, 6th ed. Philadelphia, PA: Lippincott, Williams & Wilkins, 2001:988–1010. https://doi.org/10.1016/S0735-1097(01)01617-5.
3. Cluver CA, Herbst P, Griffith-Richards S, Hassan M, Allwood B. Coarctation of the aorta: a rare cause of severe hypertension in pregnancy. Lancet. 2020 Jul 4;396(10243):62. doi: 10.1016/S0140-6736(20)31312-X. PMID: 32622398.
4. Ramlakhan KP, Tobler D, Greutmann M, Schwerzmann M, Baris L, Yetman AT, Nihoyannopoulos P, Manga P, Boersma E, Maggioni AP, Johnson MR, Hall R, Roos-Hesselink JW; ROPAC investigators group. Pregnancy outcomes in women with aortic coarctation. Heart. 2020 Oct 29;107(4):290–8. doi: 10.1136/heartjnl-2020-317513. Epub ahead of print. PMID: 33122301; PMCID: PMC7873427.
5. Beauchesne LM, Connolly HM, Ammash NM, Warnes CA. Coarctation of the aorta: outcome of pregnancy. J Am Coll Cardiol. 2001 Nov 15;38(6):1728-33. doi: 10.1016/s0735-1097(01)01617-5. PMID: 11704388.
6. Rosenthal L. Coarctation of the aorta and pregnancy: report offivecases. BMJ 1955;1:16–8. doi: 10.1136/bmj.1.4904.16. PMID: 13209181; PMCID: PMC2060735.
7. Goodwin JF. Pregnancy and coarctation of the aorta. Clin ObstetGynecol 1961;4:645–64. doi: 10.1097/00003081-196109000-00004. PMID: 13707069.
8. Mortensen JD, Ellsworth HS. Coarctation of the aorta and pregnancy: obstetric and cardiovascular complications before and after surgical correction. JAMA 1965;191:596–8. doi: 10.1001/jama.1965.03080070080017. PMID: 14239034.
9. Manalo-Estrella P, Barker AE. Histopathologic findings in human aortic media associated with pregnancy. Arch Pathol 1967;83:336–41. PMID: 4225694.
10. Rodger M, Sheppard D, Gándara E, *et al.* Haematological problems in obstetrics. Best Pract Res Clin Obstet Gynaecol 2015;29:671–84. doi: 10.1016/j.bpobgyn.2015.02.004. Epub 2015 Mar 4. PMID: 25819750.
11. Nora JJ, Nora AH. The evolution of specific genetic and environmental counseling in congenital heart diseases. Circulation 1978;57:205–13. doi: 10.1161/01.cir.57.2.205. PMID: 338196.
12. Siegmund AS, Kampman MAM, Bilardo CM, Balci A, van Dijk APJ, Oudijk MA, Mulder BJM, Roos-Hesselink JW, Sieswerda GT, Koenen SV, Sollie-Szarynska KM, Ebels T, van Veldhuisen DJ, Pieper PG; ZAHARA investigators. Pregnancy in women with corrected aortic coarctation: Uteroplacental Doppler flow and pregnancy outcome. Int J Cardiol. 2017 Dec 15;249:145-150. doi: 10.1016/j.ijcard.2017.09.167. Epub 2017 Sep 22. PMID: 28966042.
13. A. Balci, K.M. Sollie, B.J. Mulder, *et al.*, Associations between cardiovascular parameters and uteroplacental Doppler (blood) flow patterns during pregnancy in women with congenital heart disease: rationale and design of the Zwangerschap bij Aangeboren Hartafwijking (ZAHARA) II study, Am. Heart J. 161 (2) (2-2011) 269–275. doi: 10.1016/j.ahj.2010.10.024. PMID: 21315208.
14. P.G. Pieper, A. Balci, J.G. Aarnoudse, *et al.*, Uteroplacental blood flow, cardiac function, and pregnancy outcome in women with congenital heart disease. Circulation 128 (23) (12-3-2013) 2478–2487. doi: 10.1161/CIRCULATIONAHA.113.002810. Epub 2013 Nov 5. PMID: 24192800.
15. V.W. Li, Y.F. Cheung, Right ventricular myocardial motion and deformation in adolescents and young adults after repair of coarctation of the aorta. Echocardiography 32 (5) (5-2015) 797–804. doi: 10.1111/echo.12868. Epub 2014 Dec 8. PMID: 25483708.
16. O.H. Toro-Salazar, J. Steinberger, W. Thomas, A.P. Rocchini, B. Carpenter, J.H. Moller, Long-term follow-up of patients after coarctation of the aorta repair, Am. J. Cardiol. 89 (5) (3-1-2002) 541–547. doi: 10.1016/s0002-9149(01)02293-7. PMID: 11867038.