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FROM DIAGNOSIS TO TREATMENT: EXPLORING AORTIC COARCTATION AND NOVEL THERAPEUTIC STRATEGIES

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ABSTRACT:

Background: Aortic coarctation, characterized by narrowing of the aorta lumen either preductally or postductally, poses challenges in both diagnosis and treatment. Despite numerous hypotheses, its exact etiology remains elusive.

Methods: This structured abstract assembles data from medical literature to elucidate the clinical presentation, diagnostic challenges, and innovative treatment modalities for aortic coarctation.

Results: Aortic coarctation accounts for 5 to 8 percent of congenital cardiac anomalies and can manifest in both childhood and adulthood. Timely identification is critical, especially in patients under 30 years old presenting with systemic arterial hypertension, to facilitate prompt surgical intervention.

Conclusion: While the etiology of aortic coarctation remains undefined, advancements in diagnostic imaging and surgical techniques have improved outcomes for affected individuals. This abstract underscores the importance of vigilance in identifying this condition and highlights the innovative approaches in its management.

Keywords: Aortic coarctation, Aorta bicuspida, elevated blood pressure, and hypertension.

INTRODUCTION:

At 3/10,000 live births, aortic coarctation (CoAo) accounts for 5-8% of all congenital heart abnormalities. It is described as a narrowing of the aortic lumen, mainly distal to the origin of the left subclavian (postductal) or proximal artery, at the point where the aortic arch and descending aorta meet, at or below the ligamentum arteriosus's insertion. This (initiative) is more common in men (5:1) and is linked to circulatory aneurysms, left chamber hypoplasia, ventricular septal abnormalities (55%), subaortic stenosis (25%), and bicuspid aorta (30–80%). Willis (25 percent). Turner syndrome is linked to preductal coarctation, whereas Williams syndrome is typically characterized by long tubular narrowing. It usually receives collateral circulation from the anterior, vertebral, subclavian, and intercostal arteries to the distal aorta. Blood pressure and femoral pulse palpation typically diagnose this pathology in childhood during the asymptomatic phase. However, symptoms usually manifest in the third and fourth decades of adulthood, necessitating the confirmation or exclusion of this heart disease in patients of this age group who present with systemic arterial hypertension (Wei et al., 2023).

Table 1: Characteristics of Aortic Coarctation (CoAo)

Characteristic	Description
Incidence	3/10,000 live births
Gender Predominance	More common in men (5:1)
Location of Narrowing	Mainly distal to the origin of the left subclavian artery (postductal) or proximal to the left subclavian artery, at the point where the aortic arch and descending aorta meet, at or below the insertion of the ligamentum arteriosus

Associated Abnormalities	- Circulatory aneurysms - Left chamber hypoplasia - Ventricular septal abnormalities (55%) - Subaortic stenosis (25%) - Bicuspid aorta (30–80%) - Willis syndrome (25%)
Syndrome Associations	- Turner syndrome: preductal coarctation - Williams syndrome: Long tubular narrowing
Collateral Circulation	Typically receives collateral circulation from the anterior, vertebral, subclavian, and intercostal arteries to the distal aorta
Diagnosis	Blood pressure and femoral pulse palpation typically diagnosed during the childhood asymptomatic phase
Manifestation	Symptoms usually manifest in the third and fourth decades of adulthood

MEDICAL SUMMARY

Enalapril 5 mg was administered orally every 12 hours to a male patient, 22, who had a medical history of systemic arterial hypertension, which had been identified at the age of 19. He was referred from the first level of care to a cardiology visit four years before admission due to cold hands and feet, sporadic diaphoresis, and hyperhidrosis of the hands. He requested paraclinical services, which revealed the following: glucose 89 mg/dl, urea 20.8 mg/dl, creatinine 0.8 mg/dl, total cholesterol 185 mg/dl, triglycerides 181 mg/dl, Haemoglobin 16.4 g/dl, neutrophils 50%, leukocytes 8,400, and platelets 251,000. The patient's cardiovascular medical examination revealed the following: a mid-systolic murmur in the aortic focus, an expulsive hum, intensity II/VI, irradiating to the suprasternal cava region and interscapular, and a delay between the radial and femoral pulses (DARAGHMA, 2021; Meijs et al., 2021).

The patient's blood pressure was 150/90 mmHg in the left arm and 140/80 mmHg in the right arm. Following an ECG, the patient's heart rate was recorded at 65 bpm, respiratory sinus arrhythmia was observed, and left ventricular growth data was obtained, revealing a 36 mm Sokolow index (Fig. 1). A transthoracic and echocardiogram, showed a left ventricle of average size and thickness, a left ventricular ejection fraction of 70% at the Simpson biplane, a bivalve aorta with mild stenosis, the detection of CoAo with a maximum gradient of 80 mmHg (Fig. 3 A and B), and computed tomographic angiography (CT angiography) of the chest with 3D

reconstruction. Similarly, a chest x-ray was requested, and the Roesler sign and the «3» sign were visible (Fig. 2). There, the cardiothoracic surgery service assessed postductal CoAo (Fig. 4 A and B) for surgical therapy via coartectomy and end-to-end anastomosis; a favorable outcome was achieved. A circumferential sling of ductal tissue around the aorta at the level of the coarctation platform, extending from the ductus arteriosus, was observed upon histological inspection. Metoprolol 50 mg taken orally every 12 hours was needed for the patient to have ongoing hypertension medication, indicating a simple discharge (Rafiq, 2021; Raza et al., 2023).

DISCUSSION:

The etiology of CoAo is explained by several theories, including the hemodynamic theory, which postulates that abnormalities in blood flow occur at the level of the aortic isthmus during fetal life, and the embryogenic theory, which describes an aberrant pattern of cephalic migration during the development of the aortic arch. Lastly, the most widely recognized theory, put forth by Skoda, is predicated on the aberrant growth of ductal tissue inside the aorta, supporting the tissue's function in the most prevalent form, juxtaductal coarctation (van Staveren et al., 2023).

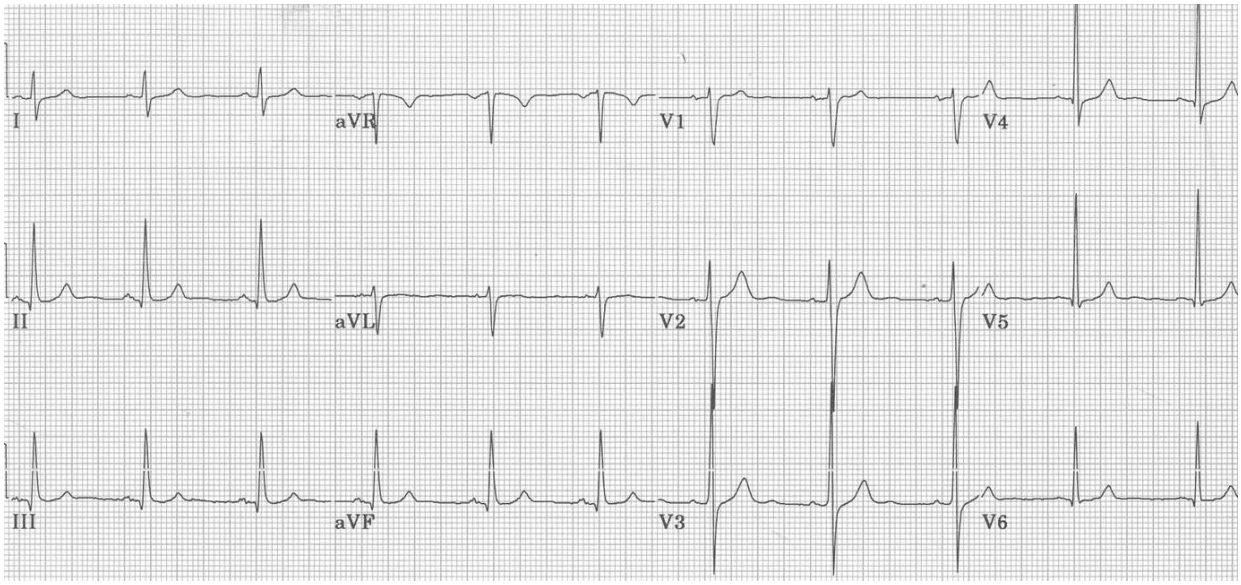


Figure 1: 12-lead electrocardiogram, A sinus arrhythmia with no ischemia, lesion, or necrosis, heart rate of 65 bpm, P wave of 80 ms, PR interval of 200 ms, QRS complex of 80 ms, aQR° + 30°, Sokolow 36 mm, and QTc of 400 ms.

The aorta's circumferential ring would form due to smooth muscle cell infiltration in the coarctation area, as shown by histopathological studies at this level. Despite this, no explanation has accounted for the vast range of concomitant abnormalities.
It is typically asymptomatic; however, when symptoms do arise, they are typically vague and include headaches, dizziness, palpitations, colds, nosebleeds, dyspnea with exertion, claudication in both legs, and, in rare circumstances, evidence of heart failure. During physical examination, there were no changes in diastolic blood pressure. Still, systolic blood pressure was higher in the upper and lower in the lower extremities (systolic pressure differential gradient > 20 mmHg). Moreover, patients with well-developed collateral circulation may not exhibit this modification. There is a delay between the radial impulse and the arrival of the femoral pulse (Mitchell et al., 2024).

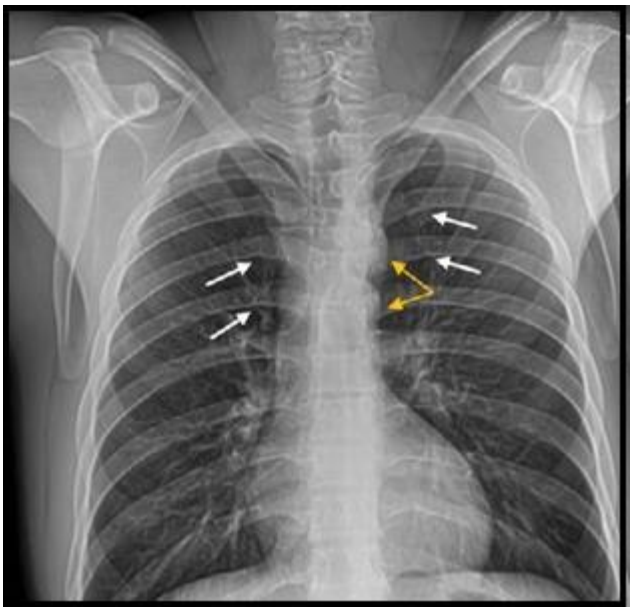


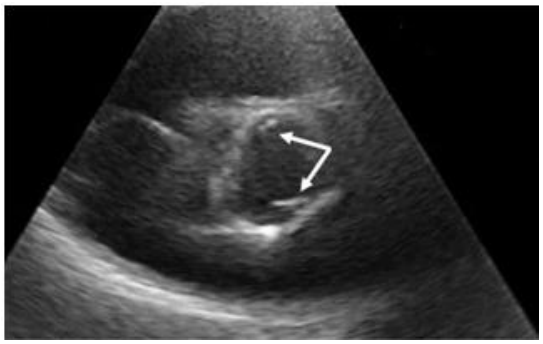
Figure 2: Anterior-posterior chest x-ray, Roesler indicator (white points). The «3» symbol has yellow arrows.

A nondisplaced apex with a prolonged systolic rise is felt at the level of the chest, and the aorta may also be perceptible in the suprasternal cavity. Due to the reinforcement of A2, there may be an enhanced S2. Usually, a mid-systolic murmur is audible in the interscapular and left paravertebral regions; in a bicuspid aorta, there may also be a proto-systolic click and a diastolic leak—instances of inadequate aortic flow. The patient in the case study had physical test results

that pointed to COAo. Although it was not audible in this patient, collateral flow in the internal mammary and intercostal arteries can occasionally be heard as a murmur. The left atrium and ventricle are growing, and the electrical axis has a leftward deviation, according to the nonspecific electrocardiogram (HALL & DENVER; Saugel et al., 2020).

Roesler's sign, which is highly specific but not very sensitive, is visible radiologically as notches developed in the lower half of the posterior costal arches (3-8°) where the intercostal arteries with higher flow are located (Adamo et al., 2022). This sign is only seen on the right side in cases of preductal CoAo. However, it is shown in the lower ribs in cases of abdominal coarctation. The "3" sign is another well-known observation. In this situation, the dilated distal aorta forms the inferior curvature, while the superior curvature is formed by the aortic arch and dilated left subclavian artery. Echocardiography is necessary to detect symptoms of aortic stenosis or insufficiency and determine the architecture, hypertrophy, and left ventricular function of the aortic valve. Adult coarctation sites can be challenging to locate; however, Doppler technology makes it possible to assess the acceleration of blood flow in the aorta as it descends from the suprasternal notch (Ushakov et al., 2021).

The patient in the case study had a trans coarctation gradient that reached a maximum of 80 mmHg. However, excellent anatomical characterization is provided by nuclear magnetic resonance angiography and CT angiography, which makes it possible to localize and describe the extent of constriction precisely. Angiography should only be used when there is doubt about the diagnosis, as a peak-to-peak gradient more significant than 20 mmHg suggests a hemodynamically severe aortic coarctation. However, because of the solid diagnostic suspicion, the patient opted to obtain a CT angiography (Atlinkson, 2023).



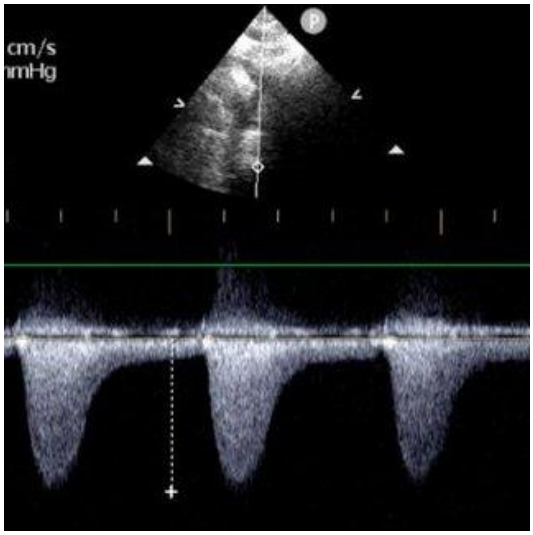


Figure 3: Transthoracic echocardiography, A: The bivalve aorta is visible on the short axis at the level of the large vessels. B: transcoarctation gradient of 80 mmHg; suprasternal axis .

Patients with a trans coarctation pressure gradient larger than 30 mmHg should be considered for surgical repair (class I recommendation, level of evidence C). This procedure involves removing the stenotic segment and either end-to-end anastomosis or replacing the stenotic segment (Liszewski & Kurian, 2020) aortic section with an impacted prosthesis. Due to the presence of significant intercostal aneurysms and atheromatous alterations at the aortic level, correcting CoAo surgically in individuals older than 15 years can be challenging8. It is crucial to remember that even in children, this surgical restoration is palliative and needs ongoing observation. Since they were first used in the 1950s, surgical repair techniques have changed (Antsiperov et al., 2022; Auersperg & Trieb, 2020).

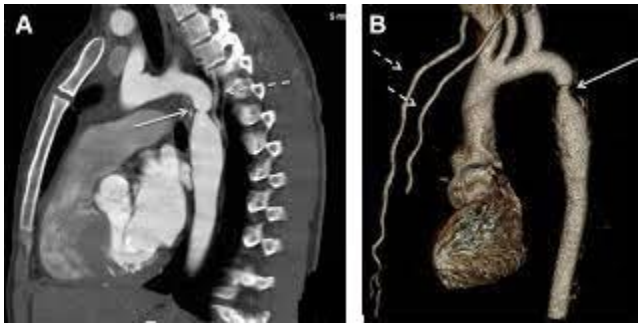


Figure 4: Chest computed tomography angiography. Section Sagittal (A). B: 3D reconstruction of the sagittal segment. At the postductal level, an aortic coarctation site is visible (arrows) (Leo et al., 2023).

Since the use of balloon-expandable prostheses has decreased the rate of complications, endovascular repair has gained popularity over the past ten years. Nevertheless, compared to surgical repair, it has a higher incidence of coarctation, the need for reoperation, and persistent arterial hypertension, so the patient decided to undergo surgical management. Adults should undergo routine follow-up MRI or CT aortography after repair, regardless of the technique utilized, mainly if endoprostheses are utilized (Kim et al., 2020; Marquis-Gravel & MD).

CONCLUSION:

Adults should be evaluated for CoAo because complications can arise between 15 and 40. These complications can manifest as heart failure, infective endocarditis in bicuspid aortic valves, endarteritis at the coarctation site, aortic dissection (which is more common in pregnant women), cerebral hemorrhage due to aneurysm rupture, and persistent systemic arterial hypertension, which affects one-third of patients who have had surgery. More than 90% of CoAo patients pass away from heart or cerebrovascular illness before turning 50 if surgical correction is not performed. Despite significant advancements in clinical diagnostic techniques in medicine, particularly in cardiology, CoAo remains a pathology for which a thorough and sufficient cardiovascular history and objective examination are necessary for a clinical diagnosis. As a result, in the case of a young patient, secondary etiology should be seriously suspected in the reported case and with data about systemic arterial hypertension until the opposite is demonstrated. It should be directed to the cardiology service instead of being treated only with antihypertensive medications, as in primary arterial hypertension. If this pathology is verified, immediate surgical intervention will be required to prevent potentially deadly follow-up consequences.

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