Jordan Medical Journal

ORIGINAL ARTICLE

Unveiling Coarctation: A Case of Early Onset Hypertension Unravelling a Late Diagnosis of an Aortic Defect

Tala A Haddad[1*], Hashim M. AlHammouri[1], Zaid Abdulraheem Al-Abed[1], Nora AbuAmmouneh[1], Toleen Haddad[1], Mahmoud Qeshta[1], Iyad Al-Ammouri[1], Hanna Makhamreh[1]

[1] Faculty of Medicine, University of Jordan

***Corresponding author:**

talahaddad73@gmail.com

Received: January 31, 2025

Accepted: April 22, 2025

DOI:
https://doi.org//10.35516jmj.v60i1.4172

Abstract

Background: Coarctation of the aorta (CoA) is a congenital heart defect occurring in approximately 6–8% of live births with congenital heart diseases. It is characterised by narrowing of the aorta, and although often diagnosed during infancy or childhood due to its impact on blood pressure (BP) regulation, some cases may not be identified until adolescence.

Case: We report a case of a 25-year-old male who presented to the ER with non-exertional chest pain, palpitations, and facial plethora. He has a history of undiagnosed hypertension, an average BP of 150/90, a significant BP gradient between upper and lower limbs (>40 mmHg), and radio-femoral delay on physical examination.

Decision-making: Comprehensive tests were conducted; results included a negative troponin assay, an electrocardiogram (ECG) showing normal sinus rhythm with left ventricular hypertrophy (LVH) and a left bundle branch block (LBBB), an echocardiogram revealing a bicuspid aortic valve (BAV) and severe concentric hypertrophy of 1.7 cm with a low-normal ejection fraction, and a normal chest X-ray. Subsequently, a computed tomography angiography confirmed the diagnosis of severe CoA with extensive collaterals, which warranted catheterization. A covered Cheatham Platinum stent was implanted, and the patient was discharged on appropriate medications afterwards.

Conclusion: This rare case of a BAV with CoA presenting in adulthood underscores the significance of timely detection, intervention, and long-term management. The late onset of symptoms is noteworthy, with the extensive collateral circulation accounting for it. Further follow-up is essential to monitor his chest pain and hypertension.

Keywords: Aortic stenosis, bicuspid aortic valve, hypertension.

INTRODUCTION

Coarctation of the aorta (CoA) is a congenital heart defect occurring in approximately 6-8% of live births with congenital heart diseases. It is characterised by narrowing or constriction of the aorta, typically located just distal to the origin of the left subclavian artery[1]. This aortopathy is characterised by abnormal vascular properties, leading to stiffness and abnormal relaxation of the aorta[1]. Although CoA is often diagnosed during infancy or childhood due to its impact on blood pressure (BP) regulation, some cases may not be identified until adolescence[2]. Risk factors for CoA include several other cardiac and vascular anomalies, such as bicuspid aortic valve (BAV) disease, ventricular septal defect, patent ductus arteriosus, and aortic arch hypoplasia. Additional risk factors could be genetic disorders, viral infections during pregnancy, and male gender[3].

BAV is the most prevalent congenital cardiac defect, with an estimated prevalence ranging from 1% to 2%. It demonstrates a male predominance, with a ratio of 1:2 to 1:4. In over 70% of cases, this condition arises from the fusion of the left and right coronary cusps (LCC and RCC). Approximately 7% of patients with BAV also have CoA, while a striking 70–75% of patients with CoA have a concomitant BAV[4]. Presented here is a rare case involving a 25-year-old man initially diagnosed with BAV, who was subsequently found to have CoA.

Patient and Observation

We report the case of a 25-year-old man, a 19-pack-year smoker and heavy alcohol user, with a history of undiagnosed elevated BP with an average of 150/90 mmHg. He presented to the emergency department with sudden-onset, constant, left-sided chest pain described as heavy in nature, radiating to the left jaw. The pain was associated with palpitations, facial plethora, and shortness of breath, reaching a Canadian Cardiovascular Society (CCS) grade of II-III. He also reported blurry vision with floaters and scotomas.

Clinical examination revealed a BP of 150/90 mmHg in the right arm and 145/85 mmHg in the left, with a significant pressure gradient between the upper and lower limbs (>40 mmHg). A hyperdynamic radial pulse, diminished femoral pulses, and radio-femoral delay were also noted. Auscultation revealed normal S1 and S2 heart sounds without murmurs or additional sounds, and an electrocardiogram showed a normal sinus rhythm at 75 beats per minute, left ventricular hypertrophy, and a left bundle branch block. Comprehensive laboratory tests and investigations were conducted; results included a negative troponin assay, and chest radiography did not reveal any significant findings. The echocardiogram (Figure 1) demonstrated a BAV with severe concentric hypertrophy of 1.7 cm and a low-normal ejection fraction of 50%. However, further evaluation with a computed tomography angiography (CTA) (Figure 2) confirmed the diagnosis of severe coarctation and extensive collateral circulation. The coarctation was located distal to the origin of the left subclavian artery, with a length of 1.5 cm between the two least narrowing points.

As catheter treatment is indicated for hypertensive patients with pressure gradients exceeding 20 mmHg (Class I, Level C)[5], a left heart catheterisation (LHC) (Figure 3) was performed via the right femoral artery using a 12 Fr long sheath. A 3 cm x 1.5 cm covered Cheatham Platinum stent, mounted on a 16 mm x 4 cm balloon, was deployed at 5 atmospheres of pressure, successfully crossing the coarcted segment of the aorta

and resolving the BP gradient. The LHC revealed favourable angiographic results, showing normal coronary arteries and eliminating the need for an additional echocardiogram.

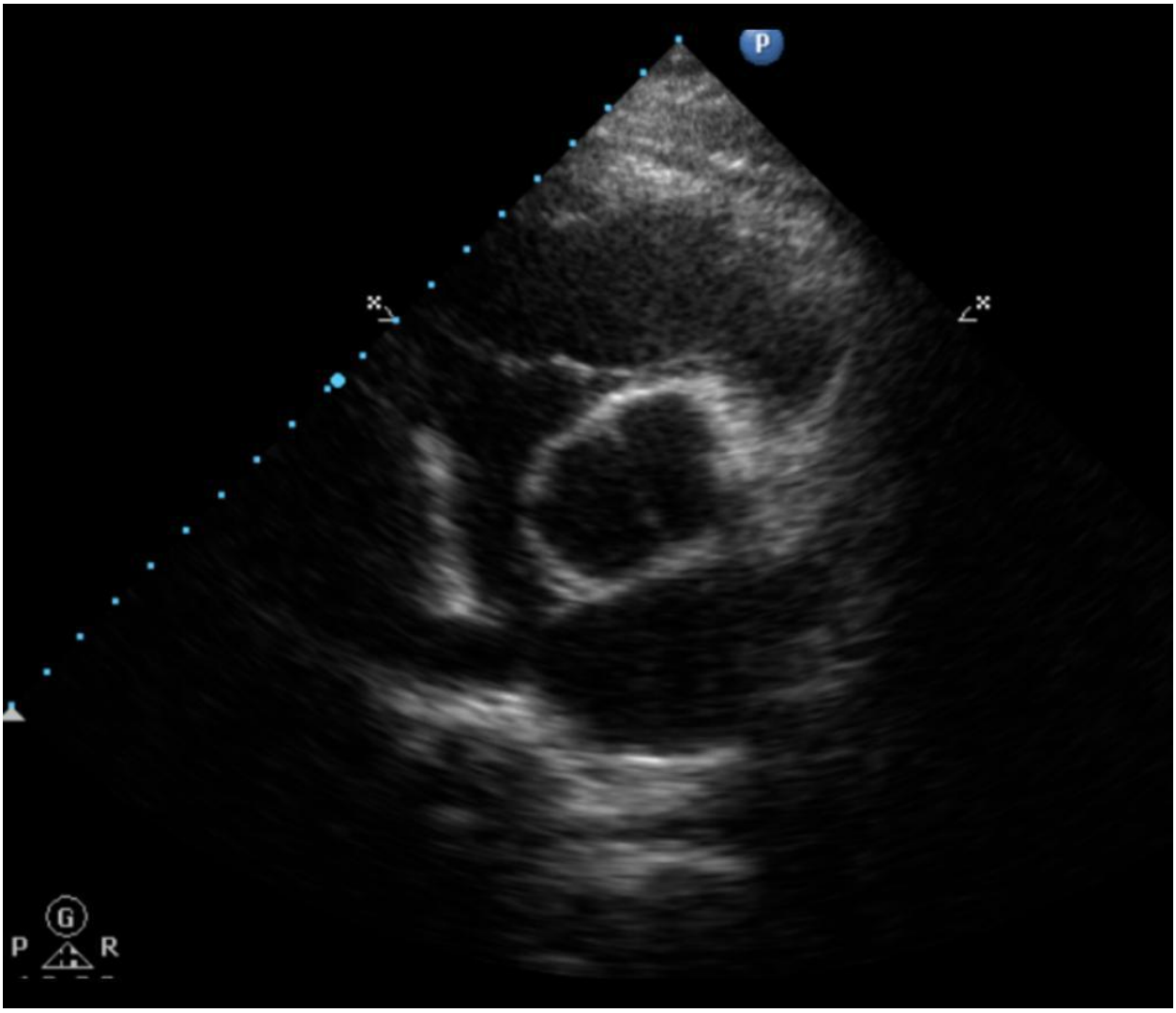

Figure 1: Echocardiogram showing BAV and severe concentric hypertrophy.

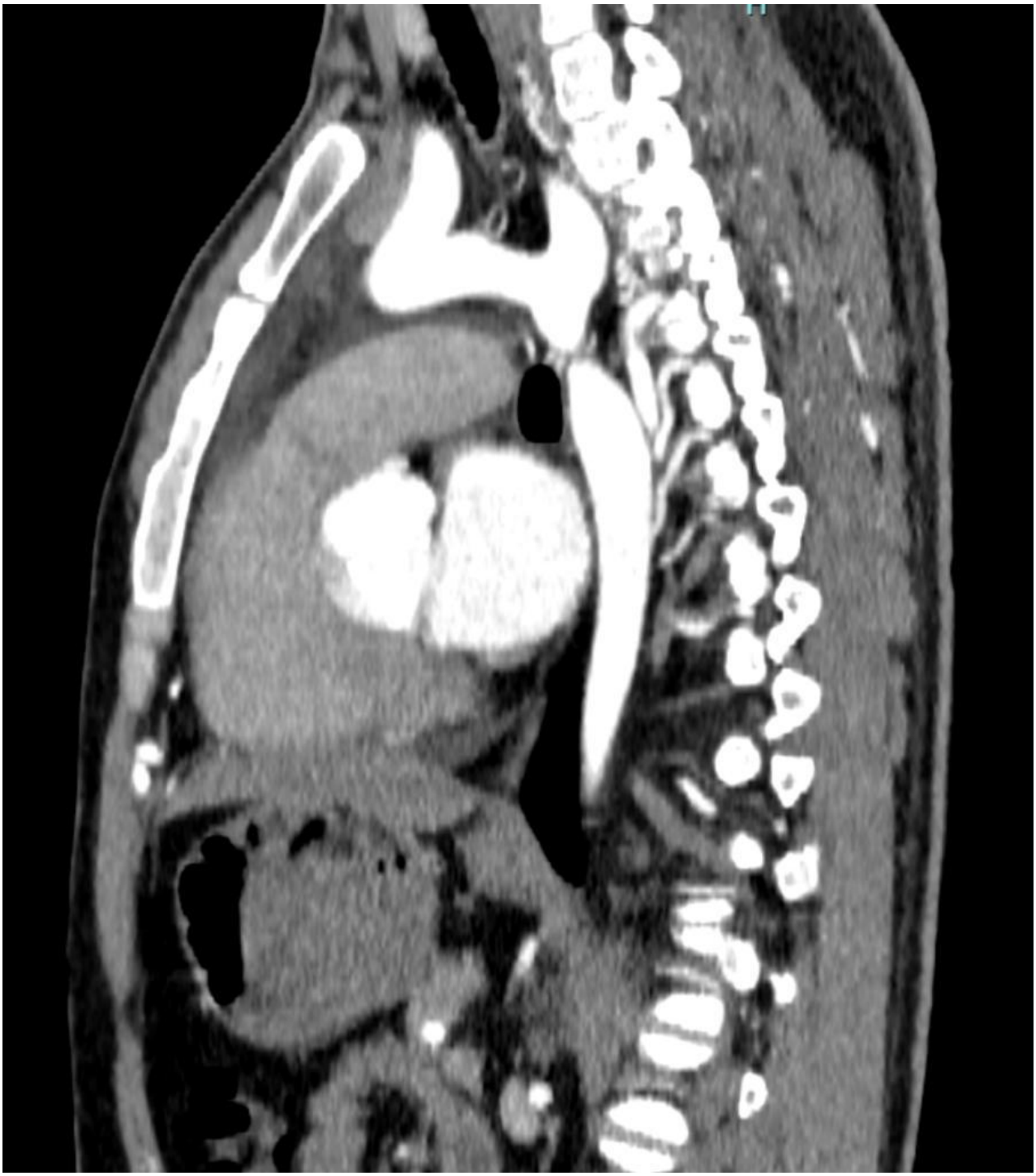

Figure 2: Lateral view CT aortogram showing severe coarctation of the aorta.

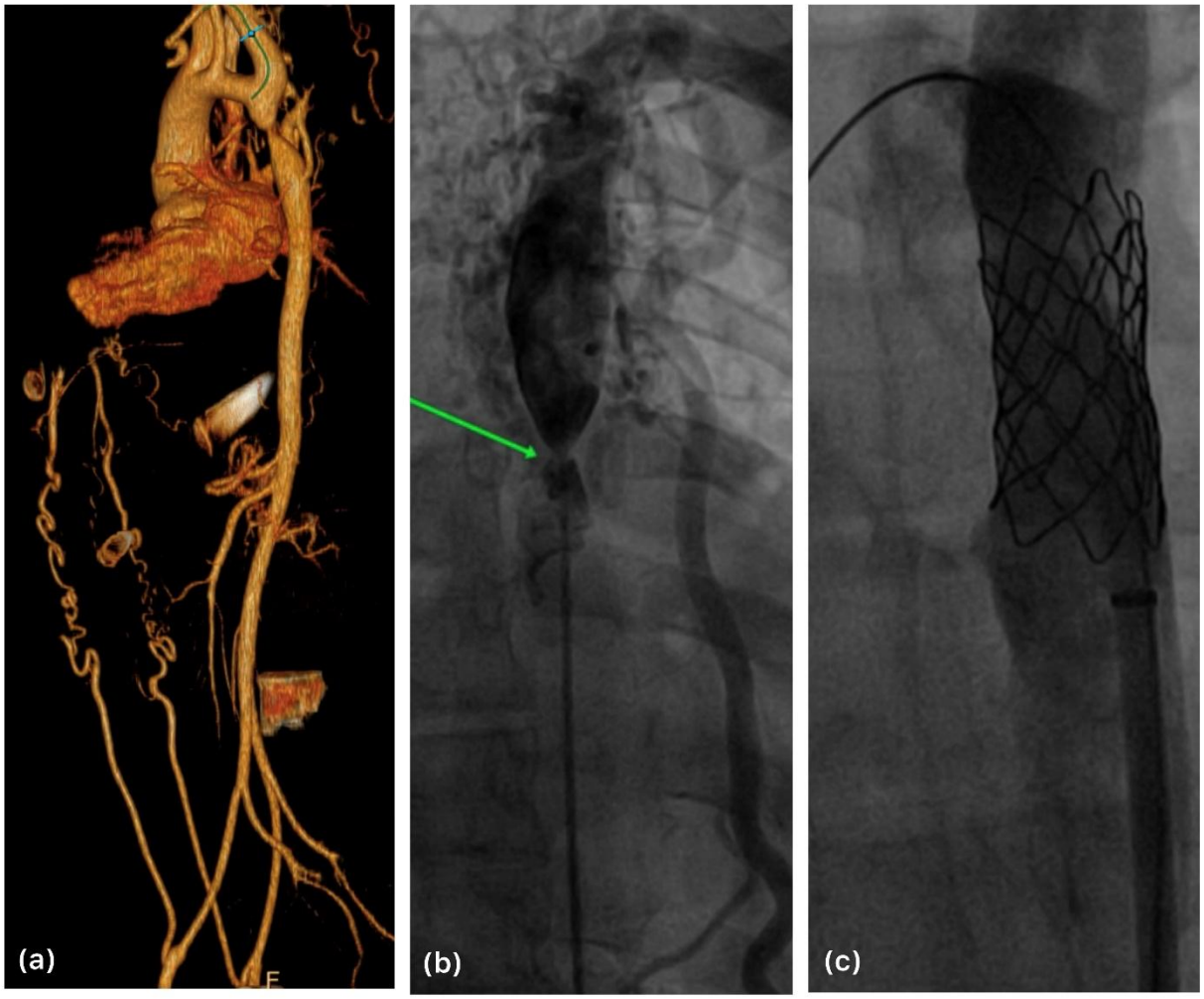

Figure 3: Left Heart Catheterization: (a) Thoracic CT aortogram: severe aortic coarctation with massive collaterals (b) Severe coarctation by aortogram with pressure gradient > 20 mmHg (c) Stenting of coarctation using a 3 cm x 1.5 cm covered CP stent implanted on a 16 mm x 4 cm balloon and deployed with 5 atmospheric pressures.

The postoperative course was uneventful, and the patient was discharged on day 3 with a medication regimen that included metoprolol (100 mg*1), valsartan (160 mg*1) / hydrochlorothiazide (12.5 mg*1), aspirin (100 mg*1), clopidogrel (75 mg*1), and atorvastatin (40 mg*1).

One month later, the patient reported significant clinical improvement, with a BP of 130/85 mmHg and normal, strong femoral pulses with no delay.

DISCUSSION

CoA is a potentially treatable cause of secondary hypertension[6], with a prevalence ranging from 6% to 8% among all congenital heart defects[1]. Hypertension is the most common sign in patients who remain undiagnosed until adulthood. Such patients may report episodes of headaches or claudication of the lower limbs. Clinical findings may include diminished lower extremity pulses and delayed systolic

pressure gradients between the upper and lower extremities[7], with higher readings in the arms[6]. However, patients with well-established collateral blood flow may only present with a slight decline in femoral pulses and lower extremity blood pressures[7]. On the contrary, this patient had diminished femoral pulses regardless of his extensive collateral blood flow.

This condition is diagnosed by clinical examination and further confirmed via two-dimensional echocardiography, which shows increased Doppler flow velocities at the coarctation site. In addition to cardiac catheterisation, which may reveal a significant systolic pressure gradient (>20 mmHg) across the coarctation, while angiography can determine the degree and type of aortic narrowing[6]. Previously, surgery was the sole option for managing aortic coarctation, but due to its associated morbidity and complications, catheter-based techniques are now increasingly preferred. Initial therapeutic options include angioplasty for children, while stenting is the preferred approach for adolescents and adults[6].

The bare and covered metal CP stents were first approved for the treatment of native and recurrent CoA in 2016, based on clinical data from the COAST (Coarctation of the Aorta Stent Trial) and COAST II studies[8]. The COAST study assessed the efficacy of the bare-metal CP stent in treating CoA. It demonstrated that 99% of patients had a peak gradient of less than 20 mmHg at the one-month follow-up, and 90% maintained this at two years. However, complications included 6% of patients developing new aneurysms and 18% requiring another intervention[9]. Conversely, the COAST II study evaluated the covered CP stent and found that it provided complete coverage of aortic wall injuries in 92% of cases, with no new injuries reported afterward. Unexpectedly, a single randomised trial recorded a higher rate of post-stent aortic wall injury in the covered stent group compared to the bare-metal stent group; nevertheless, this rate was still considered low[10].

According to COAST, 4 out of 17 patients who received a covered stent due to pre-existing aortic wall injury, near atresia of the aorta, or based on physician preference developed small aortic aneurysms after the stent implantation. In contrast, among the 105 patients who received bare-metal stents, one patient experienced a localised dissection, which was resolved on a computed tomography (CT) scan the following day, suggesting the injury had healed. During the 31-month follow-up, including high-resolution CTA at six months, pseudoaneurysms were detected in two patients from the covered stent group, while recoarctation occurred in four patients from the bare-metal stent group[11].

These studies reinforce the use of bare-metal stents for most patients with CoA, reserving covered stents for high-risk patients or those with pre-existing aneurysms, where a bare-metal stent might require a multi-step treatment approach[11]. Although the definition of high risk is more or less experiment-based, it has evolved from previous reports to include patients above the age of 40, individuals with Turner syndrome, and those with near atresia of the aorta. Accordingly, upcoming studies should direct their focus on ascertaining high-risk groups that may genuinely benefit from the option of implanting a covered stent[11]. As the coarctation was particularly tight, making it a typically high-risk case from the physician's perspective, a covered CP stent was chosen over a bare one.

The manifestation of CoA in a 25-year-old patient is considered atypical, as it deviates from the usual age of diagnosis[2]. Although he ignored mild symptoms like headaches from undiagnosed hypertension during childhood, his lack of typical signs and symptoms of CoA, such as lower extremity claudication, for most of his life is a rare aspect of his case[1,3]. The sudden onset of symptoms mimicking ischaemic cardiac issues initially directed the focus toward the heart. During the primary investigation of this patient, an echocardiogram unexpectedly revealed a BAV, preceding the diagnosis of CoA. BAV is a common associated anomaly typically detected after establishing a CoA diagnosis[4].

The delayed onset of symptoms, which only began in the last two years of his life, suggests a complex adaptation through the development of extensive collateral vessels. These vessels allowed sufficient perfusion to the lower body, concealing the symptoms of lower extremity claudication typically seen in CoA. The collateral vessels played a key role in redirecting blood flow, compensating for the aortic constriction, and masking the traditional manifestations of CoA, leading to the delayed diagnosis in this case. Failure to diagnose CoA in adults can result in serious long-term complications, including high blood pressure, coronary artery disease, and stroke, among others[12].

An important clinical association with CoA worth highlighting is berry aneurysms, or intracranial aneurysms (IAs), especially in adults[13,14,15]. Studies indicate that magnetic resonance angiography (MRA) screening detects IAs in approximately 10% of CoA patients[13,14,15]. The American Heart Association (AHA) and American College of Cardiology (ACC) guidelines suggest that it is reasonable to offer MRA or CTA screening to adults with CoA to detect intracranial aneurysms[15,16]. This recommendation is based on the increased risk of subarachnoid hemorrhage in this population. However, the European Society of Cardiology (ESC) does not recommend routine screening[15,16]. Given the potential risk, especially in patients with additional risk factors like longstanding hypertension or a bicuspid aortic valve, it may be important to consider MRA screening of the cerebral vessels in adults with CoA[13,16].

Life-long follow-up is required following stent implantation for CoA to monitor for possible complications, including restenosis, aneurysm formation, and hypertension[17,18]. Initial post-procedure monitoring is recommended to be done by an experienced cardiologist in adult congenital heart disease, which includes transthoracic echocardiography at one, three, and six months to evaluate stent position, aortic wall integrity, and blood pressure response[17,18]. Following that, CT or MRA is advised every one to two years to detect delayed complications, such as aneurysms or stent fractures[17,18].

CONCLUSION

This rare case of CoA associated with BAV presenting in adulthood highlights the importance of timely detection, intervention, and long-term management. The late onset of symptoms is particularly noteworthy, with the extensive collateral circulation likely responsible for this presentation. In conclusion, physicians should maintain a high degree of suspicion in hypertensive adults, especially if there are discrepancies in blood pressure between the arms and legs or if weak or absent femoral pulses are detected.

Ethical approval was obtained for the case report, and informed consent was secured from the patient included in the study.

Consent for publication

Consent for publication was obtained from the participants involved in the research.

Availability of data and material

All relevant data and materials related to this case reports are included in the manuscript.

Competing interests

On behalf of all authors, the corresponding author states that there is no conflict of interest.

REFERENCES

1. Law MA, Collier SA, Sharma S, Tivakaran VS. Coarctation of the Aorta. In: *StatPearls*. StatPearls Publishing; 2024. Accessed November 6, 2024. http://www.ncbi.nlm.nih.gov/books/NBK430913/
2. Kim YY, Andrade L, Cook SC. Aortic Coarctation. *Cardiology Clinics*. 2020;38(3):337-351. doi:10.1016/j.ccl.2020.04.003
3. Dijkema EJ, Leiner T, Grotenhuis HB. Diagnosis, imaging and clinical management of aortic coarctation. *Heart*. 2017;103(15):1148-1155. doi:10.1136/heartjnl-2017-311173
4. Sinning C, Zengin E, Kozlik-Feldmann R, et al. Bicuspid aortic valve and aortic coarctation in congenital heart disease—important aspects for treatment with focus on aortic vasculopathy. *Cardiovasc Diagn Ther*. 2018;8(6):780-788. doi:10.21037/cdt.2018.09.20
5. Baumgartner H, De Backer J, Babu-Narayan SV, et al. 2020 ESC Guidelines for the management of adult congenital heart disease. *European Heart Journal*. 2021;42(6):563-645. doi:10.1093/eurheartj/ehaa554
6. Rao PS. Coarctation of the aorta. *Curr Cardiol Rep*. 2005;7(6):425-434. doi:10.1007/s11886-005-0060-0
7. Torok RD. Coarctation of the aorta: Management from infancy to adulthood. *WJC*. 2015;7(11):765. doi:10.4330/wjc.v7.i11.765
8. Ringel RE, Gauvreau K, Moses H, Jenkins KJ. Coarctation of the Aorta Stent Trial (COAST): Study design and rationale. *American Heart Journal*. 2012;164(1):7-13. doi:10.1016/j.ahj.2012.04.008
9. Meadows J, Minahan M, McElhinney DB, McEnaney K, Ringel R. Intermediate Outcomes in the Prospective, Multicenter Coarctation of the Aorta Stent Trial (COAST). *Circulation*. 2015; 131(19): 1656-1664. doi:10.1161/CIRCULATIONAHA.114.013937
10. Taggart NW, Minahan M, Cabalka AK, Cetta F, Usmani K, Ringel RE. Immediate Outcomes of Covered Stent Placement for Treatment or Prevention of Aortic Wall Injury Associated With Coarctation of the Aorta (COAST II). *JACC: Cardiovascular Interventions*. 2016; 9(5): 484-493. doi:10.1016/j.jcin.2015.11.038
11. Hijazi ZM, Kenny DP. Covered Stents for Coarctation of the Aorta. *JACC: Cardiovascular Interventions*. 2014;7(4):424-425. doi:10.1016/j.jcin.2013.12.200
12. Tomasino M, Valente F, Rello Sabatè P, Martì Aguasca G, Miranda Barrio B. Stenting of aortic coarctation before coronary artery bypass surgery in an adult with acute myocardial infarction: a case report. Olsen FJ, Tan JH, Abdulhak AB, Abdullah A, eds. *European Heart Journal - Case Reports*. 2024;8(7):ytae330. doi:10.1093/ehjcr/ytae330
13. Connolly HM, Huston J, Brown RD, Warnes CA, Ammash NM, Tajik AJ. Intracranial Aneurysms in Patients With Coarctation of the Aorta: A Prospective Magnetic Resonance Angiographic Study of 100 Patients. *Mayo Clinic Proceedings*. 2003;78(12):1491-1499. doi:10.4065/78.12.1491
14. Pickard SS, Prakash A, Newburger JW, Malek AM, Wong JB. Screening for Intracranial Aneurysms in Coarctation of the Aorta: A Decision and Cost-Effectiveness Analysis. *Circ Cardiovasc Qual Outcomes.* 2020;13(8):e006406. doi:10.1161/CIRCOUTCOMES.119.006406

15. The Polls Are In: Head Imaging in Patients with Coarctation of the Aorta. *American College of Cardiology.* 2020. https://www.acc.org/Latest-in-Cardiology/Articles/2020/11/23/14/54/The-Polls-Are-In-Head-Imaging-in-Patients-with-Coarctation-of-the-Aorta
16. Buckley A, Han Um K, Ganame J. et al. Prevalence of Intracranial Aneurysms in Patients With Coarctation of the Aorta: A Systematic Review and Meta-Analysis. *JACC Adv*. 2023; 2(5). https://doi.org/10.1016/j.jacadv.2023.100394
17. Clinical Practice Algorithm For the Follow-Up of Repaired Coarctation of the Aorta. *American College of Cardiology.* 2023. https://www.acc.org/Latest-in-Cardiology/Articles/2023/02/16/17/09/Clinical-Practice-Algorithm-For-the-Follow-Up-of-Repaired-Coarctation-of-the-Aorta
18. Rao S, Seib P. Coarctation of the Aorta Guidelines: Guidelines Summary, 2014 ESC Recommendations on Coarctation of the Aorta, 2008 Adapted ACC/AHA Guidelines on Adult Coarctation of Aorta. *Medscape*. 2025. https://emedicine.medscape.com/article/895502-guidelines?form=fpf

كشف التضيّق: حالة ارتفاع ضغط الدم المبكر تكشف تشخيصًا متأخرًا لتشوّه في الشريان الأب

تالا أمجد حداد[1]، هاشم محمد الحموري[1]، زيد عبدالرحيم العبد[1]، نورا أبو أمونة[1]، تولين حداد[1]، محمود قشطة[1]، إياد العموري[1]، حنا كمال مخامرة[1]

[1] كلية الطب، الجامعة الاردنية

Received: January 31, 2025

Accepted: April 22, 2025

DOI:

https://doi.org//10.35516jmj.v60i1.4172

الملخص

الخلفية: تضيق الأبهر (CoA) هو عيب خلقي في القلب يحدث عند حوالي 6–8% من المواليد المصابين بأمراض القلب الخلقية. يتميز بتضيق في الشريان الأبهر، وعلى الرغم من أن تشخيصه يكون غالباً خلال مرحلة الرضاعة أو الطفولة بسبب تأثيره على تنظيم ضغط الدم، إلا أن بعض الحالات قد لا تُكتشف إلا في فترة المراهقة أو البلوغ.

موجز الحالة : نقدم هنا حالة لمريض ذكر يبلغ من العمر 25 عامًا، حضر إلى قسم الطوارئ مصاباً بألم في الصدر غير مرتبط بالجهد، وخفقان، واحمرار في الوجه. تتضمن سيرته المرضية ارتفاع في ضغط الدم غير مُشخص، حيث كان متوسط ضغط دمه 90/150 ملم زئبق، ووجود فرق ملموس في ضغط الدم بين الأطراف العلوية والسفلية (<40 ملم زئبق)، وتأخر نبض الفخذ مقارنة بنبض الكفّ.

التدخل العلاجي : أجُريت مجموعة فحوصات شاملة؛ أظهرت نتائجها: اختبار تروبونين سلبي، وتخطيط كهربائي للقلب (ECG) يظهر إيقاع جيبي طبيعي مع تضخم في البطين الأيسر (LVH) وانسداد في الحزمة الفرعية اليسرى (LBBB)، وتخطيط صدى القلب (Echo) يكشف عن صمام أبهري ثنائي الشرف (BAV) وتضخم مركزي شديد في عضلة القلب بسمك 7.1 سم مع نسبة كسر قذفي متدنية ضمن المعدل الطبيعي السفلي، وصورة أشعة سينية طبيعية للصدر. بعد ذلك، أكدت صورة الأوعية بالتصوير المقطعي المحوسب التشخيص وذلك بوجود تضيق شديد في الشريان الأبهر مع تفرعات أوعية تعويضية واسعة، ما استدعى إجراء قسطرة قلبية. تم زرع دعامة مغطاة من نوع Cheatham Platinum، وأُخرج المريض لاحقاً مع صرف الأدوية المناسبة.

الاستنتاج: تسليط الضوء على هذه الحالة النادرة لصمام أبهري ثنائي الشرف مع تضيق الأبهر الذي ظهر في مرحلة البلوغ يبرز أهمية الكشف المبكر، والتدخل العلاجي المناسب، والمتابعة طويلة الأمد. وتجدر الإشارة إلى أن ظهور الأعراض في وقت متأخر يعود جزئياً إلى وجود دوران تعويضي واسع. كما أن المتابعة المستمرة ضرورية لمراقبة ألم الصدر وارتفاع ضغط الدم.

الكلمات الدالة: تضيق الأبهر، الصمام الأبهري ثنائي الشرف، ارتفاع ضغط الدم.