ESC European Society of Cardiology

European Heart Journal - Case Reports (2025) **9**, ytaf147
https://doi.org/10.1093/ehjcr/ytaf147

CASE REPORT
Adult congenital heart disease

Aorta-right atrium tunnel and mitral valve prolapse complicated with flail leaflet: a case report of a multimodality approach for an accurate diagnosis

Andreea Varvara[1], Ruxandra Oana Jurcuț[1,2], Lucian Predescu[1,2], Ioana Lupescu[1,3], and Daniela Oana Andrei [2]*

[1]Faculty of Medicine, University of Medicine and Pharmacy 'Carol Davila,' Eroii Sanitari Bvd., No. 8, Sector 5, Bucharest 020021, Romania; [2]Department of Cardiology, Emergency Institute for Cardiovascular Diseases 'Prof. Dr. C.C. Iliescu,' Sos. Fundeni, No. 258, Sector 2, Bucharest 022322, Romania; and [3]Department of Radiology, Fundeni Clinical Institute, Sos. Fundeni, No. 258, Sector 2, Bucharest 022328, Romania

Received 28 May 2024; revised 12 November 2024; accepted 19 March 2025; online publish-ahead-of-print 28 March 2025

Background An aorta-right atrial tunnel (ARAT) is a rare congenital cardiac anomaly that connects the aorta at any coronary sinus to the right atrium. Its cause remains unclear and may involve abnormal origins of the coronary arteries. There is no established link between ARAT and mitral valve prolapse (MVP). Diagnosing ARAT can be challenging, often necessitating multiple imaging techniques, while its management is complex and is further complicated by the association with MVP in our patient.

Case summary We report a case of a 46-year-old man admitted for dyspnoea and palpitations. Thirteen years prior, he had been diagnosed with a cardiac murmur. Echocardiography revealed ARAT and MVP complicated by a flail leaflet and severe regurgitation. AngioCT confirmed the right coronary sinus as the origin of the ARAT and demonstrated the right coronary artery (RCA) arising deep within the tunnel. Cardiac catheterisation identified unclassified pulmonary hypertension due to a left-to-right shunt, along with severe mitral regurgitation. Given the unfavourable anatomy for interventional treatment and the patient's decline in surgery despite multiple consultations, our team opted for conservative management. Currently, the patient's clinical status and echocardiographic parameters remain stable.

Discussion ARAT is a complex congenital disorder that presents diagnostic and treatment challenges, requiring multimodal imaging and teamwork. Since the RCA originates deep within the tunnel and is associated with MVP complicated by flail leaflet and severe regurgitation, we recommend surgical intervention as the appropriate treatment approach. Considering the patient's preferences, conservative treatment was chosen.

Keywords Aorta-right atrium tunnel • Coronary artery anomaly • Mitral valve prolapse • Flail mitral leaflet • Multimodality imaging • Case report

ESC curriculum 2.1 Imaging modalities • 2.4 Cardiac computed tomography • 2.2 Echocardiography • 4.3 Mitral regurgitation • 9.7 Adult congenital heart disease

* Corresponding author. Tel: +40721313973, Email: oanaandrei75@gmail.com; oanaandrei2000@yahoo.com
Handling Editor: Flemming Javier Olsen
Peer-reviewers: Rizwan Ahmed; Melonie Johns
Compliance Editor: Abdullah Abdullah

Learning points

- The aorta-right atrium tunnel (ARAT) is a rare congenital condition that may be linked to other cardiac anomalies. While no cases have yet associated it with mitral valve prolapse, such a connection is possible due to shared extracellular matrix abnormalities.
- A collaborative approach using multimodal imaging is essential for understanding clinical challenges and determining the best management strategy.
- The treatment strategy of the ARAT depends on the tunnel's size, sharp U-turns, and the origin of the coronary artery, along with other associated anomalies.

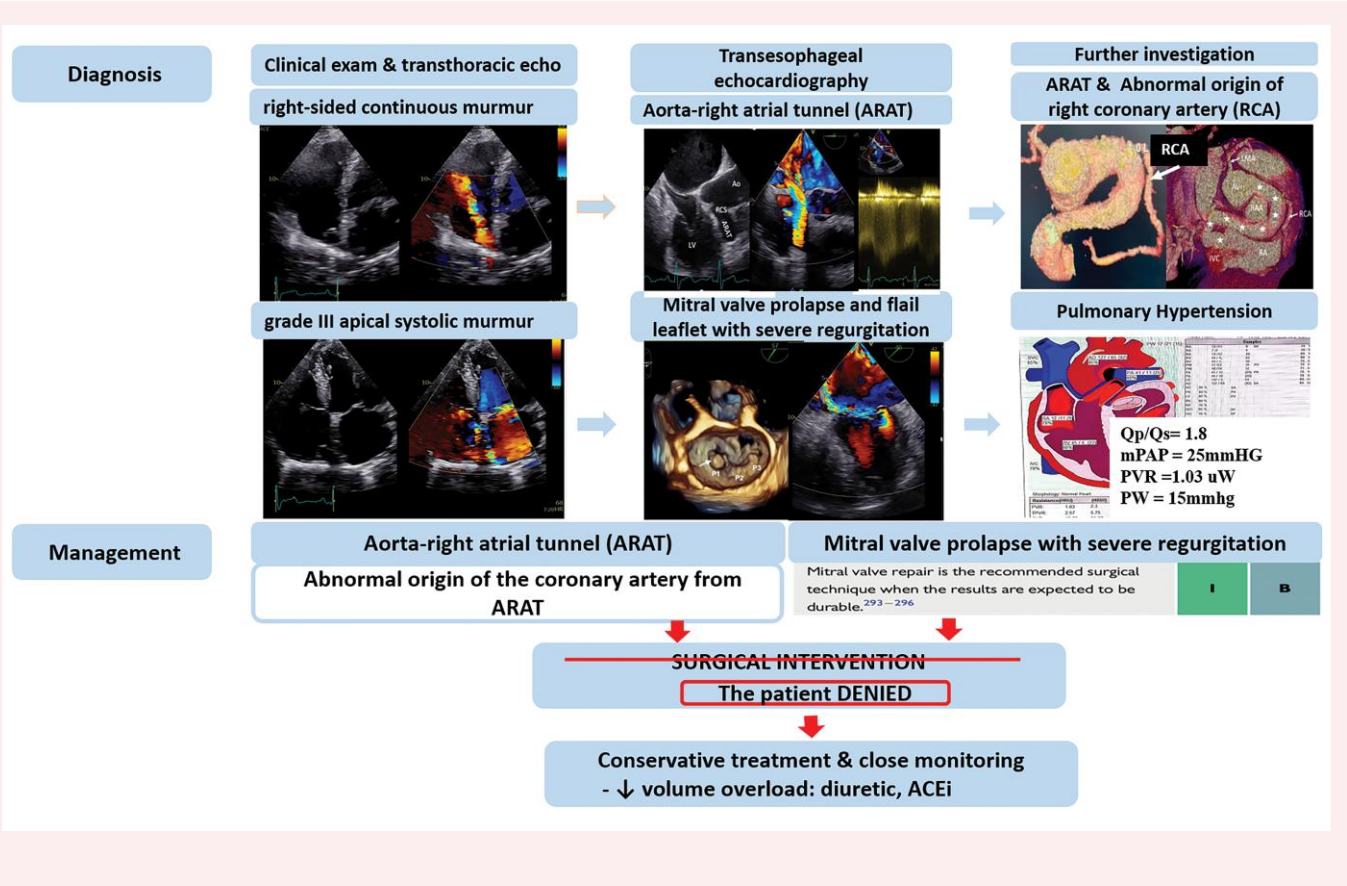
Introduction

The aorta-right atrial tunnel (ARAT) is a rare congenital anomaly that creates communication between the aorta and the right atrium (RA).[1–4] It features a tunnel-like extracardiac structure that acts as a left-to-right shunt and should be considered in the differential diagnosis of persistent cardiac murmurs.[1–4] Although Coto *et al.*[1] first described this condition over forty years ago, the exact causes remain unclear.[1] This anomaly's complexity is further heightened by its association with abnormal coronary artery origin and mitral valve prolapse (MVP). Therefore, a multimodal imaging approach is crucial for an accurate diagnosis and the best treatment strategy choice.

Case presentation

We present the case of a 46-year-old male who was admitted to our hospital with palpitations and shortness of breath during mild exertion. His medical history notes an asymptomatic systolic murmur indicated at the age of 17, with no other reported cardiovascular abnormalities at that time. After that, he was lost to follow-up. Upon admission, the physical examination revealed dyspnoea with minimal exertion and mild ankle swelling. Cardiac auscultation indicated an apical systolic murmur graded at 4/6 and a continuous murmur at 3/6 at the right parasternal border. His vital signs were stable: a regular heart rate of 85 beats per minute, a blood pressure of 125/80 mmHg, and an oxygen saturation of 97% on room air. Upon auscultation, lung fields were clear bilaterally, and no significant abnormalities were noted on the chest X-ray. Routine blood tests showed normal results, except for an elevated level of NT-proBNP at 700 pg/mL (normal range: <125 pg/mL).

Summary figure



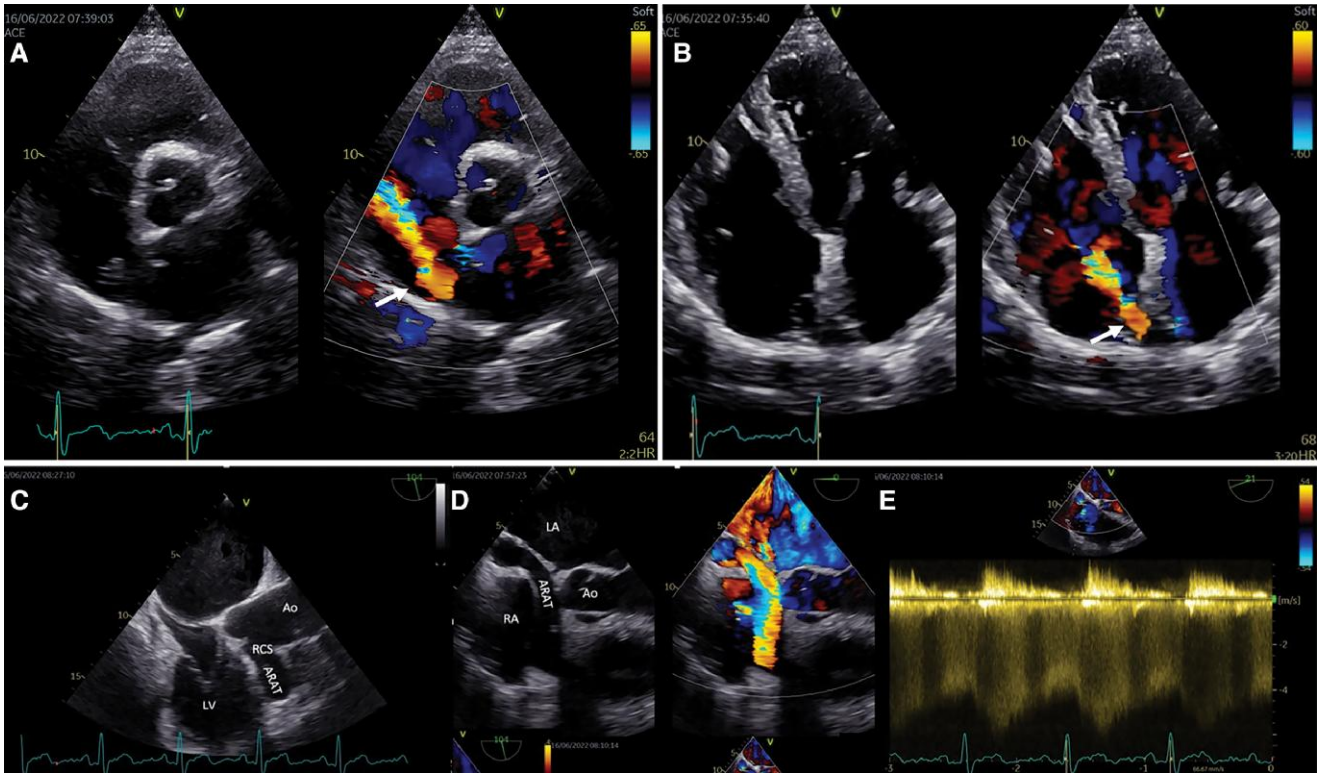


Figure 1 2D and colour Doppler transthoracic echocardiography: parasternal short-axis (*A*) and modified four-chamber view (*B*) both show continuous turbulent flux on colour Doppler examination (white arrows), probably from the aortic root to the RA roof towards the tricuspid valve. 2D modified mid-esophageal long axis and short-axis view: aortic root (Ao) to the RA tunnel (ARAT), originating from the right coronary (RCS) sinus and turning towards the RA (*C*). The atrial outlet segment of the U-turn ARAT on colour Doppler (*D*). Continuous systolic-diastolic flow with a systolic velocity of 5.2 m/s in the ARAT (*E*). Ao, aorta; ARAT, aortic root to the right atrial tunnel; LA, left atrium; LV, left ventricle; RA, right atrium.

The electrocardiogram revealed sinus rhythm, premature supraventricular beats, left atrial (LA) enlargement, and right ventricular (RV) hypertrophy.

Transthoracic echocardiography revealed a continuous turbulent flow on colour Doppler examination, likely directed from the right coronary sinus to the roof of the RA (*Figure 1A and B*). Additionally, MVP with severe regurgitation was observed. The LV was dilated with LVEDV of 172 mL and LVESV of 73 mL but maintained a preserved ejection fraction (LVEF of 58%) and severe dilated LA with an LAVi of 85.9 mL/m^2. The right heart chambers (RV and RA) were dilated, showing mild systolic dysfunction (RV fractional area change of 35%), moderate tricuspid regurgitation, and pulmonary hypertension (PH), indicated by a systolic pulmonary artery pressure (PAP) of 51 mmHg. Transesophageal echocardiography showed that the turbulent flow likely resulted from a connection between the right coronary sinus and the RA (*Figure 1C and D*; Supplementary material online, *Videos S1* and *S2*), with a flow velocity of 5.2 m/s at the drainage orifice of the RA (*Figure 1E*). The mechanism of severe mitral regurgitation (MR) due to the flail of the P1 scallop and prolapse of the P2 and P3 scallops was also identified (*Figure 2*).

Cardiac catheterisation conducted for haemodynamic assessment revealed a left-to-right shunt with a flow ratio (Qp: Qs) of 1.80 and unclassified PH [mean PAP was 25 mmHg, the pulmonary capillary wedge pressure was 15 mmHg, and the pulmonary vascular resistance (PVR) was 1.03 Wood units]. Additionally, coronary angiography showed a normal origin of the left main coronary artery, while a large tunnel originating from the right coronary sinus flowed into the RA without identifying the origin of the right coronary artery (RCA).

We performed a computed tomography angiography that showed an extracardiac anterior-type tunnel originating from the anterior aspect of the ascending aorta at the level of the right coronary sinus. After running in an anteroinferior direction, the tunnel makes a sharp U-turn, redirecting the passage towards the roof of the RA. It travels along the posterior side of the interatrial septum and is oriented towards the RA just above the entrance of the inferior vena cava. The distal part of the tunnel is narrower than the initial segment and drains into the RA through a restrictive opening (see *Figures 3* and *4* and Supplementary material online, *Video S1*). This design protects the pulmonary circulation from high pressure, maintaining normal PVR. The RCA originates from the tunnel near the U-turn (*Figure 3E*). Due to the RCA's abnormal origin, surgical closure of the tunnel was necessary, along with reimplantation of the coronary artery into the respective sinus of Valsalva. Even if the atrial end had the proper dimensions, potential rupture and retrograde RCA thrombosis could complicate percutaneous closure. Furthermore, due to the associated severe MR, surgical treatment was the recommendation for our patient.

After reviewing the clinical, anatomical, and haemodynamic data, our multidisciplinary team concluded that surgical treatment would

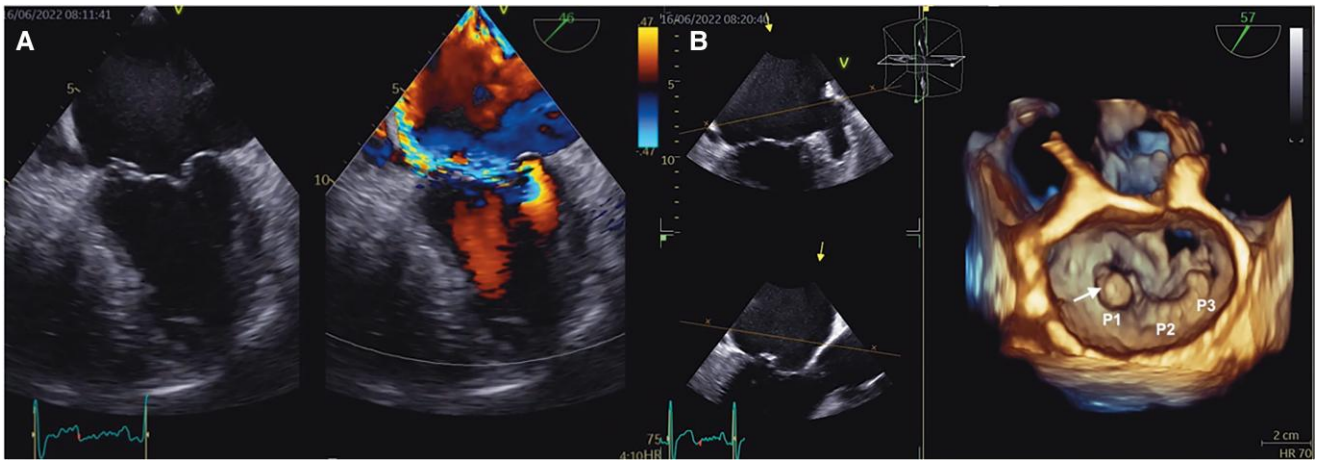


Figure 2 Transesophageal echocardiography evaluation of the mitral valve. (*A*) 2D and colour Doppler transesophageal echocardiography at mid-esophageal level—severe MR with eccentric jet due to posterior leaflet prolapse. (*B*) 3D atrial view of the MV showing flail of the P1 scallop (arrow) and prolapse of the P2 and P3 MV scallops.

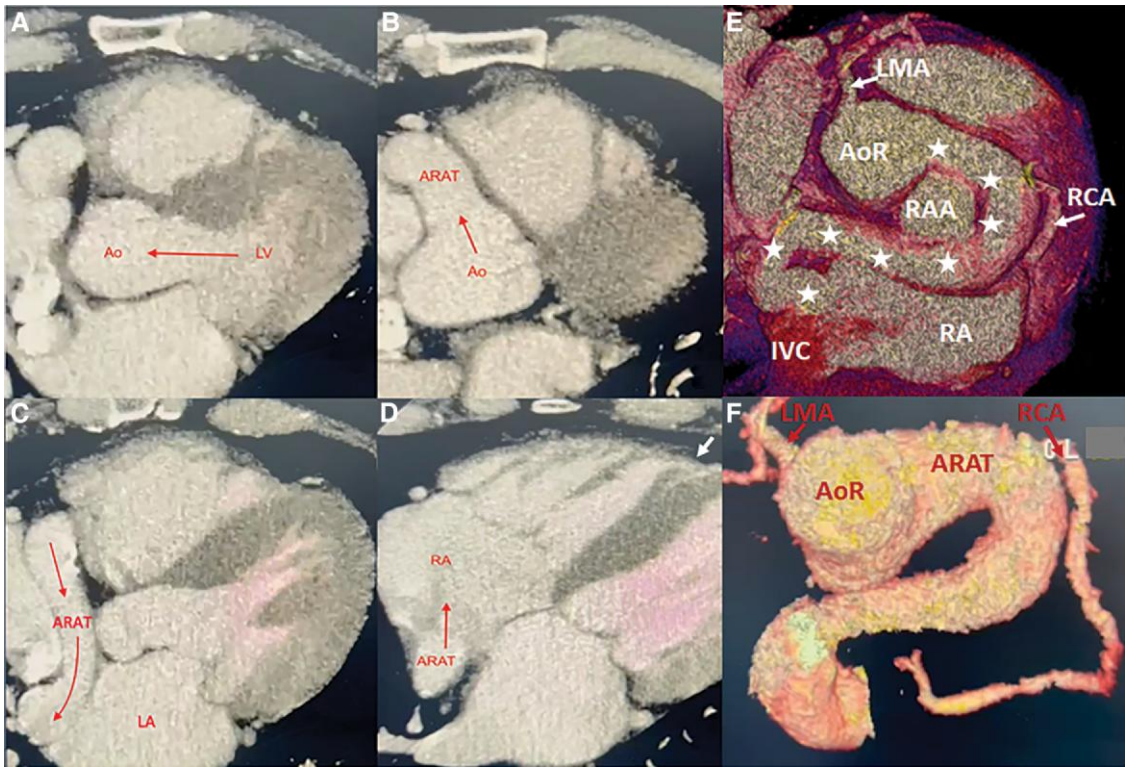


Figure 3 Computed tomographic angiography illustrates a large ARAT originating from the right coronary sinus (*A*, *B*). It takes an anterior initial course, followed by a first U-turn with anterior convexity, changing direction towards the RA. It then proceeds along the posterior side of the interatrial septum (*C*), turning off to the superolateral side of the RA (*D*). (*E*, *F*) 3D CT reconstruction of the ARAT (stars show its entire course) also shows the coronary arteries' origins. Ao, aorta; AoR, aortic root; ARAT, aorta right atrial tunnel; IVC, inferior vena cava; LV, left ventricle; LMA, left main artery; RA, right atrium; RAA, right atrial appendage; RCA, right coronary artery.

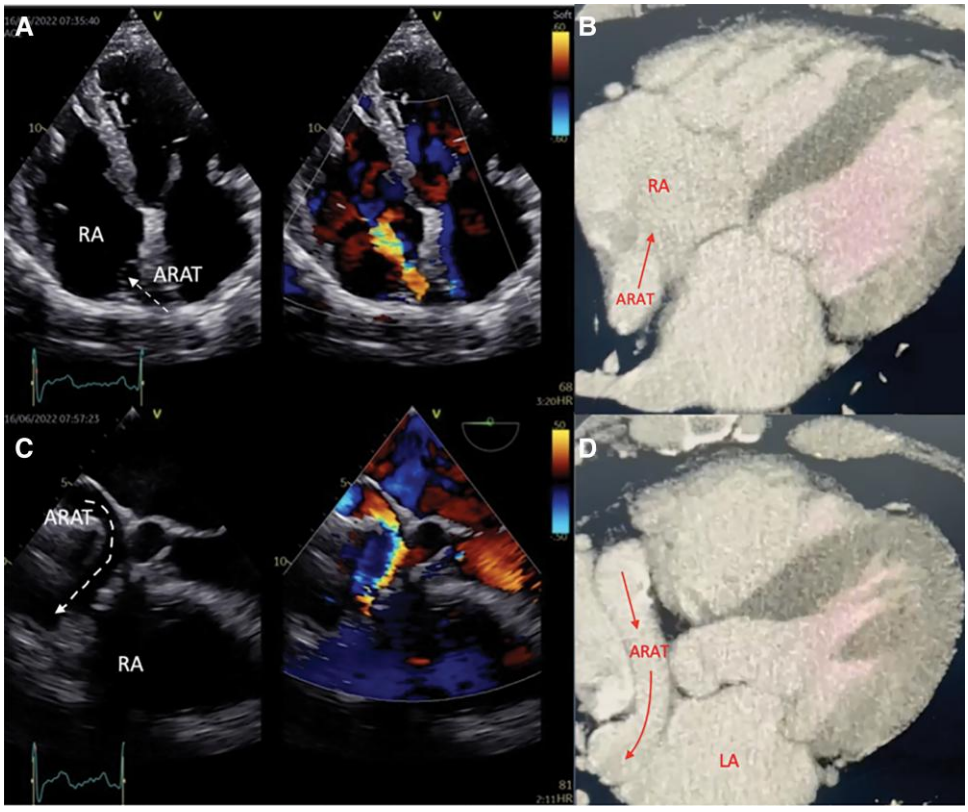


Figure 4 Comparative imaging of the ARAT through echocardiography and computed tomography. 2D and colour Doppler transthoracic echocardiography in a modified four-chamber view reveals turbulent flow on the colour Doppler examination (white arrows), likely originating from the aortic root to the right atrial (RA) roof, toward the tricuspid valve *(A)*. Computed tomographic angiography, modified four-chamber view, reveals a large aorta-right atrial tunnel (ARAT) extending to the superolateral side of the right atrium (RA) *(B)*. 2D and colour Doppler modified mid-esophageal short-axis view and computed tomographic angiography: the atrial outlet segment of the U-turn ARAT on colour Doppler *(C)* proceeds along the posterior side of the interatrial septum *(D)*. ARAT, aorta right atrial tunnel; RA, right atrium.

be optimal. Despite multiple consultations and the risk of future Eisenmenger's syndrome, the patient declined surgery. Consequently, our teamwork considered the interventional approach for the ARAT and severe MR. However, the ARAT anatomy was unfavourable for occlusion by intervention. Furthermore, based on current guidelines and complicated mechanisms of severe MR—including P1 flail and P2 and P3 prolapse—we determined that the interventional approach carries both short—and long-term, high-risk complications in a young patient. Therefore, we decided to proceed with conservative management. Currently, the patient's clinical status remains stable, and the echocardiogram parameters showed no changes during the 3-and 6-month follow-ups. However, we will continue to emphasize the potential benefits of surgical treatment during future follow-up meetings.

Discussions

To the best of our knowledge, this is the first case in the literature describing the association of the ARAT and the RCA anomaly with the MVP and severe regurgitation.

ARAT is a rare pathological condition characterised by an extracardiac connection between any coronary sinus and the RA. It was first described by Coto *et al.*[1] in 1980. The tunnel can be classified as either anterior or posterior, depending on which side of the aorta it originates from.[4] The tunnel typically curves sharply, making a U-turn to connect to the roof of the RA. Additionally, the coronary artery can arise from any segment of the tunnel, usually either before the U-turn or near the tunnel's origin.[4] Notably, we could distinguish the ARAT from the coronary cameral fistula based on normal calibre, abnormal origin from the tunnel, normal termination of the ipsilateral coronary artery, and the absence of intramyocardial coronary artery branches. We could also differentiate the ARAT from the sinus of the Valsalva aneurysm rupture, as the tunnel had an extracardiac course.[1–4]

Limited information is available regarding the etiopathogenesis of this abnormality.[1–6] Gajjar *et al.*[3] suggested that ARAT may result from a congenital deficiency in the external elastic lamina, which weakens the aortic wall.[3,4] This weakness leads to the gradual enlargement of the tunnel due to high systolic aortic pressure, which drains into the RA, a low-pressure cardiac cavity located nearby. This theory accounts for the tunnel's course, which is directed posteriorly from the left coronary sinus (the most common origin) and anteriorly from the right coronary sinus. Considering this etiopathogenesis, the association between ARAT and MVP may not be coincidental, as similar abnormalities in extracellular matrix components could contribute to both conditions.[3,4,7] Thus, a genetic disorder may underlie both cardiac abnormalities. Genetic testing could provide further insight into the etiopathogenesis of ARAT; however, an association between ARAT and MVP has not yet been documented. Notably, our patient declined genetic testing.

Due to the increased volume overload in both ventricles, the clinical presentation can range from an asymptomatic continuous murmur to more complicated and symptomatic forms of heart failure and PH.[1–3,8–13] Additionally, a flail leaflet may exacerbate MR and its associated symptoms. Without any inflammatory signs, myxomatous degeneration of the MV is likely the cause of the flail leaflet.[14]

We employed various imaging techniques to perform a precise anatomical and haemodynamic evaluation of the ARAT. These techniques included transthoracic echocardiography, transesophageal echocardiography, cardiac catheterisation, and computed tomography angiography. This comprehensive approach enabled us to diagnose the condition accurately, distinguish it from the more common aorta-right atrium communication, and identify the most effective treatment. The management strategy is determined by the tunnel's type, size, and pathway; the location of the origin of the coronary artery in relation to the aortic end, and other associated cardiac anomalies.[3,8–16]

In our patient, congestive heart failure appears to be caused by volume overload resulting from both the left-to-right shunt and severe MR. This condition is likely worsened by the spontaneous rupture of the chordae, leading to a flail P1 leaflet. However, the percutaneous management was unfavourable considering the complex anatomy of the ARAT associated with abnormal coronary artery origin and the P2, P3 MVP complicated by the P1 flail in a young patient.[3,8–16] Therefore, the surgical approach was the choice of our teamwork. Nonetheless, the surgical strategy for our patient is complicated and involves both mitral valvuloplasty and surgical closure of the ARAT with reimplantation of the RCA.

Conclusions

Our case emphasizes the need for a multimodal imaging approach to accurately diagnose complex congenital cardiac diseases. ARAT and coronary artery anomalies are associated with MVP complicated by a flail leaflet. A comprehensive multimodal imaging strategy and a multidisciplinary team aim for optimisation management. Clinical management options were discussed with the patient, and the patient's informed preferences guided subsequent treatment decisions.

Lead author biography

Dr Daniela Oana Andrei is a clinical and imaging cardiologist. Currently, she is a senior consultant at Emergency Institute of Cardiovascular disease CC Iliescu, Bucharest, Romania. She has authored original manuscript and one chapter of book in imaging cardiology.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

Acknowledgements

The authors want to acknowledge the Catheterisation Laboratory staff at the Emergency Institute of Cardiovascular Diseases 'Prof. Dr CC Iliescu' in Bucharest, Romania, and the Radiology Department staff at the Fundeni Clinical Institute in Bucharest, Romania.

Consent: The authors confirm that written consent for the submission and publication of this case report, including images and associated text, has been obtained from the patient, following COPE guidance.

Conflict of interest. The authors certify that they have NO affiliations with or involvement in any organisation or any entity with any financial interest (such as honoraria, educational grants, participation in speakers' bureaus, membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patent-licensing arrangements), or non-financial interest (such as personal or professional relationships, affiliations, knowledge or beliefs) in the subject matter or materials discussed in this manuscript.

Funding: None declared.

Data availability

The data underlying this article are available in the article and its online Supplementary material.

References

1. Coto EO, Caffarena JM, Such M, Marques JL. Aorta-right atrial communication. Report of an unusual case. *J Thorac Cardiovasc Surg* 1980;**80**:941–944.
2. Rosenberg H, Williams WG, Trusler GA, Smallhorn J, Rowe RD, Moes CA, et al. Congenital aortic-right atrial communications: the dilemma of differentiation from coronary-cameral fistula. *J Thorac Cardiovasc Surg* 1986;**91**:841–847.
3. Gajjar T, Voleti C, Matta R, Iyer R, Dash PK, Desai N. Aorta-right atrial tunnel: clinical presentation, diagnostic criteria, and surgical options. *J Thorac Cardiovasc Surg* 2005 Nov; **130**:1287–1292.
4. Rad EM, Hijazi ZM, Pouraliakbar H, Mirzaaghayan MR, Zamani H. Congenital aorto-cardiac connections (CACC) revisited: introduction of a novel anatomic-therapeutic classification. *Pediatr Cardiol* 2021;**42**:1459–1477.
5. Anderson KR, Ho SY, Anderson RH. Location and vascular supply of sinus node in human heart. *Br Heart J* 1979;**41**:28–32.
6. Bharati S, Lev M, Cassels DE. Aortico right ventricular tunnel. *Chest* 1973;**63**:198–202.
7. Delwarde C, Capoulade R, Mérot J, Le Scouarnec S, Bouatia-Naji N, Yu M, et al. Genetics and pathophysiology of mitral valve prolapse. *Front Cardiovasc Med* 2023;**10**:1077788.
8. Lee S, Kim SW, Im SI, Yong HS, Choi CU, Lim HE, et al. Aorta–right atrial tunnel. *Circulation* 2016;**133**:e454–e457.
9. Sreedharan M, Baruah B, Dash PK. Aorta-right atrial tunnel—a novel therapeutic option [1]. *Int J Cardiol* 2006;**107**:410–412.
10. Kalekar T, Prabhu AS, Dilip D, Dolas A. A rare case of aorta-right atrial tunnel demonstrated on coronary computed tomography angiography. *Afr J Thorac Crit Care Med* 2023;**11**:24–25.
11. Onorato EM, Costante AM, Andreini D, Bartorelli AL. Infective endocarditis of an asymptomatic congenital aorta-right atrial tunnel: a case report. *Eur Heart J Case Rep* 2021;**4**:1–5.
12. Sai Krishna C, Baruah DK, Reddy GV, Panigrahi NK, Suman K, Kumar PVN. Aorta-right atrial tunnel. *Tex Heart Inst J* 2010;**37**:480–482.
13. Jain J, Wani A, Kulkarni A, Yelne P. Aorta-right atrial tunnel presenting with heart failure in an adult case report. 2019; Available from: www.heartviews.org
14. Lancellotti P, Moura L, Pierard LA, Agricola E, Popescu BA, Tribouilloy C, et al. European association of echocardiography recommendations for the assessment of valvular regurgitation. Part 2: mitral and tricuspid regurgitation (native valve disease). *Eur J Echocardiogr* 2010;**11**:307–332.
15. Vahanian A, Beyersdorf F, Praz F, Milojevic M, Baldus S, Bauersachs J, et al. 2021 ESC/EACTS guidelines for the management of valvular heart disease. *Eur Heart J* 2022;**43**: 561–632.
16. Buzzatti N, Van Hemelrijck M, Denti P, Ruggeri S, Schiavi D, Scarfò IS, et al. Transcatheter or surgical repair for degenerative mitral regurgitation in elderly patients: a propensity-weighted analysis. *J Thorac Cardiovasc Surg* 2019;**158**:86–94.e1.