ECRONICON OPEN ACCESS

EC CARDIOLOGY
Case Report

Aorta to the Right Atrial Tunnel (ARAT): A Case Report

Shanika Wijayalath[1]*, K Tillakaratne[1], Raghunathan[2] and S Perera[3]

[1]*Senior Registrar at Lady Ridgeway Hospital for Children, Sri Lanka*
[2]*Consultant Paediatric Cardiologist at Karapitiya Teaching Hospital, Sri Lanka*
[3]*Consultant Paediatric Cardiologist at Lady Ridgeway Hospital for Children, Sri Lanka*

***Corresponding Author:** Shanika Wijayalath, Senior Registrar at Lady Ridgeway Hospital for Children, Sri Lanka.

Received: June 22, 2021; **Published:** July 31, 2021

Abstract

Aorta to the Right atrial Tunnel is a rare abnormality. We report a case of the aorta- right atrial tunnel (ARAT), a tunnel, which arises from the right Coronary sinus to the right atrium, while RCA arises independently from the fistulous tract closer to the aortic end of the tunnel.

Those patients are relatively asymptomatic apart from a continuous murmur.

Echocardiogram and angiogram will provide definitive diagnostic aid for optimum management. The indications for intervention are debated. The management options include the surgical closure of the tunnel or and the percutaneous transcatheter closure of ARAT with coils, vascular plugs, and duct occluders.

We present a case of 4-year old girl who underwent percutaneous transcatheter closure of ARAT successfully with a duct occluder and vascular plug.

Keywords: *Coronary Vessel Anomalies; Fistula; Aneurysm*

Introduction

A tunnel arising from the aorta to the right atrium is an uncommon congenital anomaly. This anomaly was first described by Coto., *et al*. [1] as the aorta atrial communication. In this congenital anomaly, a large vascular link, establishes a shunt originating from the aortic root and terminating in the right atrium. Moreover, this congenital anomaly can be associated with coronary artery anomalies as well. We present a case of the aorta- right atrial tunnel (ARAT), a tunnel, which arises from the right Coronary sinus to the right atrium, while RCA arises independently from the fistulous tract closer to the aortic end of the tunnel.

Case Report

A 4-year-old girl was evaluated for a continuous murmur best heard at the right upper sternal edge, although her chest x-ray did not reveal a cardiomegaly, her Echocardiograph revealed evidence that suggested A right coronary artery to the right atrial fistula. She was followed up at the cardiology clinic and she remained asymptomatic without syncope, chest pain, or respiratory distress.

The recent follow up echocardiogram showed aneurismal dilatation and she underwent an angiogram (A selective right coronary angiogram and aortic root angiogram) which revealed a large tunnel beginning from the right coronary sinus while the right coronary artery is seen arising independently from the fistulous tract closer to the aortic end of the tunnel. Left main coronary artery (LMCA), left coronary artery (LAD), left circumflex arty (LCx) were normal.

The tunnel was engaged with a 4F RCA catheter and 0.032 angle tip Terumo guidewire was advanced across the tunnel into Right atrium-inferior vena cava (RA-IVC) junction which was then snared from IVC via venous end using 5 mm gooseneck snare and an arterio-venous loop created.

A 5F Amplatzer device occluder (ADOII) delivery sheath was passed from the venous side across the tunnel into the aorta. An 8 mm Amplatzer vascular plug4 (13.5mm in length) was then deployed across the tunnel close to the aortic end, well away from the RCA origin.

RCA flow and position of the vascular plug4 were confirmed by angiograms via a catheter passed into the aorta from the aortic end.

As there was significant flow across the tunnel following deployment, a 6 x 4 ADO II duct occluder was deployed within the tunnel closer to the RA end.

Finally, the aortic end of the tunnel showed unobstructed flow in RCA with complete occlusion of the fistulous tract.

Device closure of the aorta to right atrial tunnel was successful without complications.

She was prescribed antibiotic prophylaxis for 6 months following the procedure as prophylaxis against infective endocarditis and was also advised to maintain good oral hygiene at all times. Moreover, she has to be on lifelong Aspirin and Clopidogrel was advised for 3 months post-procedure.

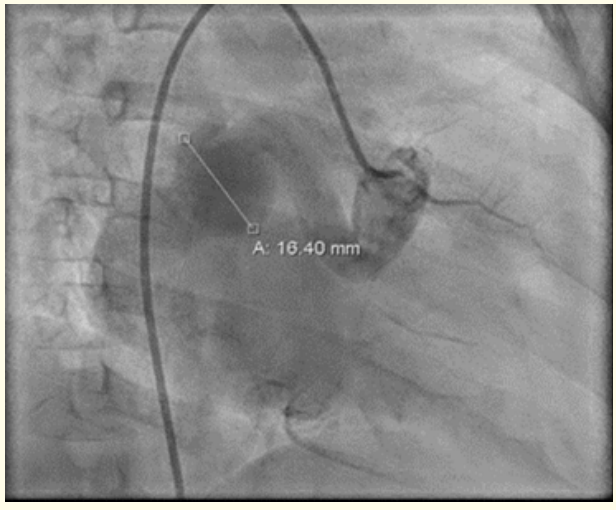

Figure 1: *Selective right coronary angiogram shows Aorta to right atrial fistula with aneurysmal dilatation measured 16.4 mm.*

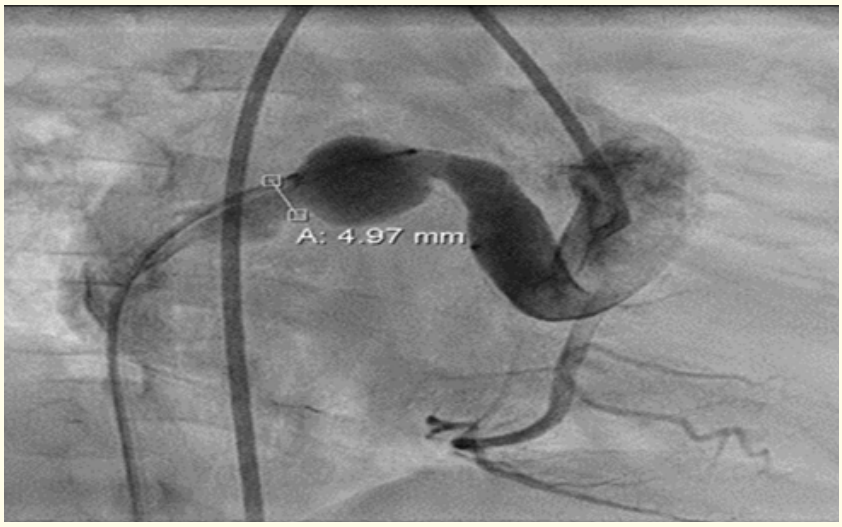

Figure 2: *Selective right coronary angiogram shows restricted opening at right atrial end, measured 4.97 mm.*

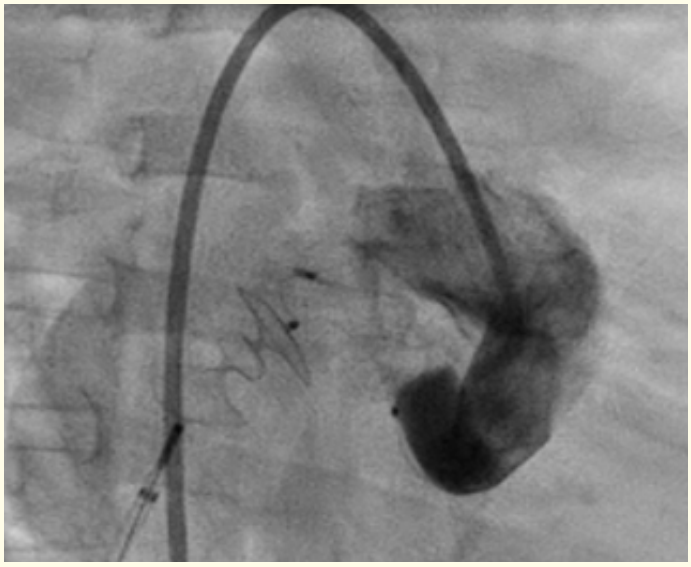

Figure 3: *Angiogram following device closure of the Aorta to Right atrial tunnel.*

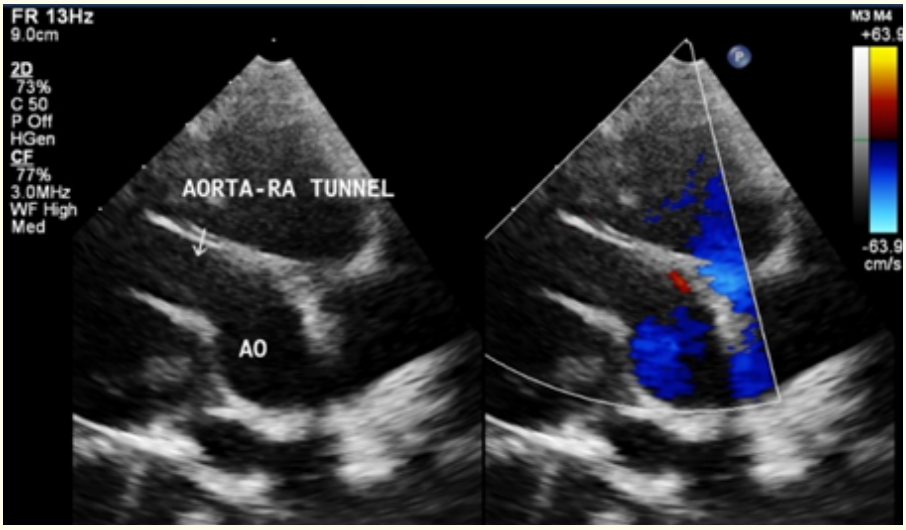

Figure 4: *Following device closure of the aorta-right atrial tunnel, pos t-procedure echocardiogram.*

Discussion and Conclusion

Aorta- right atrial tunnel (ARAT), is a very rare congenital cardiac anomaly, characterized by a vascular tunnel between any one of the aortic sinuses and the right atrium. Etiology for this pathology is not yet clearly understood. Gajjar., *et al*. [2] have suggested that congenital deficiency of the elastic lamina in the aortic media may cause an extracardiac tunnel from the aorta to the right atrium, as from a higher aortic pressure to lower right atrium pressure.

In majority of reported cases, the tunnel arose from the left sinus Valsalva [2-9]. The tunnel which originates from right sinus of Valsalva is a very rare phenomenon; therefore only 2 cases have been reported according to our knowledge so far. The other least common type is the tunnel originating from the non-coronary sinus of Valsalva. The tunnel which originates from the left sinus of Valsalva course posteriorly, while the tunnel arising from the right sinus Valsalva travels anteriorly, in relation to ascending aorta.

The origin of the coronary ostia appears to be independent from case to case. The tunnels arising from the left sinus, LAD, and LCX originated separately from the left sinus [3-6] whereas in some cases left coronary or right coronary artery arose from deferent part of the aorta to the right atrial tunnel [4,7].

In our case, the tunnel began from the right coronary sinus while right coronary artery arose independently from fistulous tract closer to aortic end of the tunnel. Left main coronary artery (LMCA), left coronary artery (LAD), left circumflex artery (LCx) was found to be normal in origin. ARAT can also be associated with other congenital anomalies, such as ostium Secundum atrial septal defect and rarely can be associated with absent right superior vena cava and presence of left superior vena cava [1].

Indications for closure remain controversial due to its asymptomatic nature. A few patients complain of dyspnea and palpitation since ARAT is a variant of the arterio- venous shunt. Authors justify the decision of ARAT closure as it minimizes the risk of volume overload to both ventricles as well as aneurysm formation followed by spontaneous rupture and infective Endocarditis.

There are two basic management options, one is surgical closure of the tunnel or and the second is percutaneous transcatheter closure of ARAT with coils, vascular plugs and duct occluders. Percutaneous catheter approach is advised if the coronary ostia originates independently from any of coronary sinus, the opening of the right atrial end is small and if there is no associated cardiac anomaly [2,4,6,8,9] as it is in our case.

In summary, ARAT should be considered as a differential diagnosis of a continuous murmur. Angiography is the basic approach for diagnosis and confirming the relationship with coronary arteries, their course and drainage into the right atrium. Management options are selected considering ARAT anatomy and potential risk factors of patients.

Bibliography

1. Coto EO., *et al*. "Aorta--right atrial communication. Report of an unusual case". *The Journal of Thoracic and Cardiovascular Surgery* 80 (1980): 941-944.

2. Gajjar T., *et al*. "Aorta-right atrial tunnel: clinical presentation, diagnostic criteria, and surgical options". *The Journal of Thoracic and Cardiovascular Surgery* 130 (2005): 1287-1292.

3. Kalangos A., *et al*. "Aorticoright atrial tunnel". *The Annals of Thoracic Surgery* 69 (2000): 635-637.

4. Kursaklioglu H., *et al*. "Aortico-right atrial tunnel in an adult patient". *The International Journal of Cardiovascular Imaging* 21 (2005): 383-385.

5. Rosenberg H., *et al*. "Congenital aortico-right atrial communications. The dilemma of differentiation from coronary-cameral fistula". *The Journal of Thoracic and Cardiovascular Surgery* 91 (1986): 841-847.

6. Sreedharan M., *et al*. "Aorta-right atrial tunnel--a novel therapeutic option". *International Journal of Cardiology* 107 (2006): 410-412.

7. Türkay C., *et al*. "Aorta-right atrial tunnel". *The Journal of Thoracic and Cardiovascular Surgery* 125 (2003): 1058-1060.

8. Baykan A., *et al*. "Aorta-right atrial tunnel closure using the transcatheter technique: a case of a 3-year-old child". *Cardiology in the Young* 23 (2013): 457-459.

9. AtilaIyisoy MD., *et al*. "An interesting type of a congenital Coronary artery anomaly". *Korean Circulation Journal* 44.3 (2014): 193-195.

Volume 8 Issue 8 August 2021