Case Report

E Journal of Cardiovascular Medicine

EJCM 2022;10(3):144-147

DOI: 10.32596/ejcm.galenos.2022.2022-07-042

Successful Surgical Management of Aortopulmonary Window and Supravalvular and Valvular Aortic Stenosis in Five Years Old Child

Onur Barış Dayanır[1], Kaan Yıldız[2], Şebnem Paytoncu[3], Mustafa Kır[1], Öztekin Oto[1]

[1]Dokuz Eylül University, Faculty of Medicine, Department of Cardiovascular Surgery, İzmir, Turkey

[2]University of Health Sciences Turkey, İzmir Tepecik Training and Research Hospital, Clinic of Child Health and Diseases, Division of Pediatric Cardiology, İzmir, Turkey

[3]Manisa City Hospital, Clinic of Pediatric Cardiology, Manisa, Turkey

Abstract

The aortopulmonary window (APW) is a rare congenital abnormality referring to communication between the main pulmonary artery and ascending aorta. To avoid irreversible pulmonary vascular disease, this type of congenital abnormalities should be repaired whenever it is diagnosed. The recommended timing for the surgery, in common practice, is three months of age. In older patients, the outcome is determined by pulmonary vascular resistance at the time of surgical repair. In this report, we present our five years old patient who underwent a valvular and supravalvular aortic surgery and concomitantly closure of APW successfully.

Keywords: Aortopulmonary window, supravalvular aortic stenosis, aortic valvular stenosis, pulmonary hypertension

Introduction

The aortopulmonary window is a rare cardiac anomaly representing 0.2%-0.6% of all congenital heart diseases[1]. Since these types of congenital abnormalities occur between the ascending aorta and the common pulmonary artery, they may cause pulmonary arterial hypertension in a short while if they are not closed by surgical or transcatheter techniques[2,3]. The pathophysiology and clinical manifestations of APW in general are quite similar to those of patients with patent ductus arteriosus. Even if

Address for Correspondence: Onur Barış Dayanır, Dokuz Eylül University, Faculty of Medicine, Department of Cardiovascular Surgery, İzmir, Turkey

e-mail: onurbarisdayanir95@gmail.com **ORCID:** orcid.org/0000-0003-1293-3898

Received: 26.07.2022 **Accepted:** 20.08.2022

Cite this article as: Dayanır OB, Oto Ö. Successful Surgical Management of Aortopulmonary Window and Supravalvular and Valvular Aortic Stenosis in Five Years Old Child. EJCM 2022;10(3):144-147.

DOI: 10.32596/ejcm.galenos.2022.2022-07-042

arterial pulmonary hypertension is manifests and elevated pulmonary vascular resistance has occurred, the closure of APW has still been recommended with acceptable long-term results[4]. Since the patient had been closely followed up with severe valvular and supravalvular aortic stenosis, the timing of the operation was delayed due to the possible need of mechanical aortic valve replacement (AVR). Although pre-operative computed tomography (CT) angiography showed an APW, it was not confirmed at any pre-op echocardiography. Here we report a case of successfully surgical closure of APW concomitantly with aortic root and supravalvular aortic stenosis enlargement and mechanical AVR in a five-year old boy.

Case Report

A five-year-old male child with echocardiographic diagnosis of supravalvular and valvular aortic stenosis at birth was admitted to the hospital with mild short of breath on exertion. CT angiography revealed enlargement of the pulmonary artery at the conus level and a large APW and increased diameters of pulmonary veins with enlarged left atrium and left ventricle enlarged significantly (Figure 1). Pre-operative echocardiography showed a supravalvular aortic stenosis with a peak gradient of 65 mmHg, first-degree aortic regurgitation, mild mitral regurgitation and 50 mmHg peak gradient of valvular aortic stenosis, with no confirmation of APW.

The patient underwent aortic open heart surgery with standard technique. When vertical standard pericardiotomy was performed, a large APW was observed when (Figure 2). At this very moment, inoperability was discussed due to the large APW and pulmonary artery. For a scientific decision making, pulmonary artery pressure was measured directly with a diagnostic line. Pulmonary arterial peak pressure was measured as 55 mgHg while simultaneous arterial pressure was 105 mmHg. An aortotomy was performed and selective antegrade cold blood cardioplegia was applied to the right and left coronary ostium. On these measurements, the patient was accepted as being operable, and the operation was carried out. After the cross-clamping of the aorta, bicaval cannulation and dual venting was applied (one to left atrium via patent foramen ovale, a second one to pulmonary artery) to provide a completely clear, bloodless exposure. Selective coronary antegrade cold blood cardioplegia was applied for myocardial protection. The APW, which measured approximately 2.0-2.5 cm, was observed between the ascending aorta and the pulmonary artery. An oblique aortotomy extended into the non-coronary sinus of the valsalva to the aortic annulus to perform Nick's aortic root enlargement with a bovine pericardial patch. A 19 size Medtronic mechanical bileaflet valve was replaced after the root enlargement. Before the enlargement of the supravalvular segment of the aorta, a separate round pericardial patch was tailored to

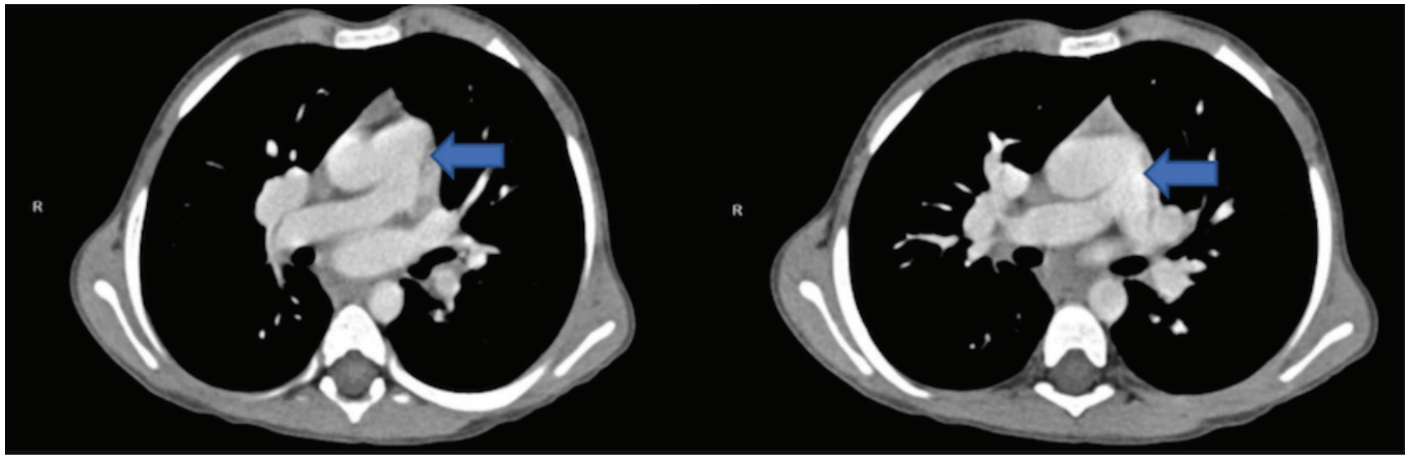

Figure 1. Aortopulmonary window CT angiography image from two different sections marked with a blue arrow
CT: Computed tomography

2 cm diameter and the APW was closed with 5-0 prolene and the continuous running suture technique.

The patient was separated from the extracorporeal circulation without positive inotropic support, and was followed an uneventful post-operative course with extubation at the 4th hour postoperatively.

Post-operative echocardiography confirmed no residual shunt and no supravalvular gradient with normal mechanical aortic valve functions and surprisingly normal pulmonary artery pressure. The patient was discharged from the hospital on the fourth post-operative day with no symptoms.

Discussion

Aortopulmonary window

The aortopulmonary window is a developmental anomaly of the conotruncal septum and has an incidence of 0.2%-0.6% among all congenital heart diseases. It is an extremely rare cardiac[1,5]. Should be repaired when diagnosed, preferably before three months of age. Survival in APW depends on the defect size and the pulmonary arterial hypertension. The ideal timing for repair of APW is before the development of irreversible pulmonary arterial hypertension[1]. Cases where APW is not treated at the ideal time may result in heart failure and death at an advanced age. Early closure is often the best treatment to ensure patient survival[6]. In this case, APW was not noticed and could not be treated at the ideal time, although the patient was able to survive until late childhood. Postoperative management of pulmonary hypertension and its long-term course are the most important factors affecting the decision to surgical treatment in patients past infancy[1]. Other literature has demonstrated favorable early and long-term outcomes after surgical correction, regardless of age or pulmonary vascular resistance[2,4]. We decided to repair the defect based on preoperative pulmonary artery pressure measurement. The surgery was uneventful and the post-operative recovery was successful. In the surgical treatment of multiple anomalies, APW closure is important because it reduces pulmonary artery pressure. The aortic stenosis and APW observed in our patient are examples of this situation.

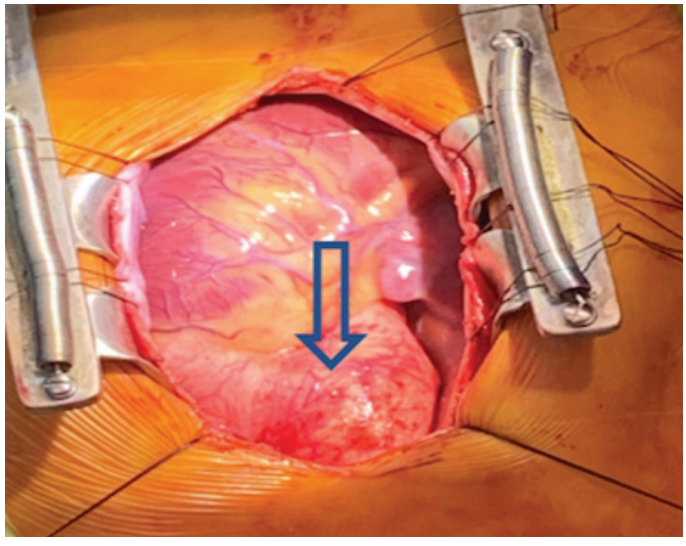

Figure 2. Blue arrow points to the connection between the aorta and the pulmonary artery

Our report strongly supports the need for surgical repair as soon as the diagnosis is made, regardless of the patient's age. Additionally, APW repair contributed positively to the postoperative course in the patient who required surgery due to aortic stenosis.

The reason for lack of confirmation of APW at pre-operative echocardiography seems to be high turbulence due to supravalvular aortic stenosis, which possibly shadowed the large APW flow.

Conclusion

The aortopulmonary window can be treated surgically in early years of childhood. Surgical risk and indications of operation need to be carefully considered regardless of the patient's age. Pulmonary hypertension is the most important parameter affecting the operation decision, postoperative course, and long-term results. Accompanying aortic stenosis with APW may be misleading on echocardiographic examination. Therefore, CT angiography is a useful method in the diagnosis phase. Additionally, APW repair contributed positively to the

postoperative course in the patient who required surgery due to aortic stenosis.

Ethics

Informed Consent: The informed consent was obtained for the article.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions

Concept: O.B.D., K.Y., Ş.P., M.K., Ö.O., Design: O.B.D., K.Y., Ş.P., M.K., Ö.O., Data Collection and/or Processing: O.B.D., K.Y., Ş.P., M.K., Ö.O., Analysis and/or Interpretation: O.B.D., K.Y., Ş.P., M.K., Ö.O., Literature Search: O.B.D., K.Y., Ş.P., M.K., Ö.O., Writing: O.B.D., K.Y., Ş.P., M.K., Ö.O.

Conflict of Interest: The authors have no conflicts of interest to declare.

Financial Disclosure: The authors declared that this study received no financial support.

References

1. Talwar S, Siddharth B, Gupta SK, et al. Aortopulmonary window: results of repair beyond infancy. Interact Cardiovasc Thorac Surg 2017;25:740-4.
2. Gowda D, Gajjar T, Rao JN, et al. Surgical management of the aortopulmonary window: 24 years of experience and lessons learned. Interact Cardiovasc Thorac Surg 2017;25:302-9.
3. Uçar T, Karagözlü S, Ramoğlu MG, Tutar E. Transcatheter closure of aortopulmonary window with Amplatzer duct occluder II: additional size. Cardiol Young 2020;30:424-6.
4. Talwar S, Agarwal P, Choudhary SK, et al. Aortopulmonary window: morphology, diagnosis, and long-term results. J Card Surg 2017;32:138-44.
5. Alsoufi B, Schlosser B, McCracken C, et al. Current outcomes of the surgical management of aortopulmonary window and associated cardiac lesions. Ann Thorac Surg 2016;102:608-14.
6. Zani A, Becchetti E, Chimenti T, C et al. Use of an Amplatz Canine Ductal Occluder (ACDO) device to close an acquired aortopulmonary fistula with a hybrid approach in a dog. J Vet Cardiol 2016;18:377-84.