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Case Report

Successful Repair of Unusual Complex of Aortic Atresia with Interrupted Aortic Arch: A Case Report

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Abstract

Interrupted aortic arch with aortic atresia is an unusual complex of congenital heart disease. Incompatibility for life is a potential risk unless there is blood flow in the ascending aorta supplying the coronary arteries from another existing malformations. We report a five days old case with interrupted aortic arch type B, aortic atresia, ventricular septal defect and an aortopulmonary window as a source of blood flow to the coronary arteries, ascending aorta and proximal aortic arch. A successful surgical correction with biventricular repair was performed. The significance of this is to expand the candidates for one stage total correction of congenital heart diseases in neonates, which spares patients multiple staged surgeries and spares health service providers the burden of multiple treatment strategies.

Keywords

interrupted aortic arch; aortic atresia; aortopulmonary window; case report

Introduction

The coexistence of aortic atresia (AA) with interrupted aortic arch (IAA) is a rare anomalous complex of the aortic valve and aortic arch. In this congenital heart disease, the survival of the baby during the intrauterine and neonatal periods depends primarily on the presence of the coronary circulation which is supplied by blood coming from the ascending aorta (Asc.Ao) that usually comes from other anomalies, like ductus arteriosus or aberrant right subclavian artery (RSA) [1]. In addition to the above, these cases often have a well-developed apex-forming left ventricle, due to the presence of the ventricular septal defect (VSD), so total biventricular surgical correction is a possible therapeutic option [2]. We present successful biventricular surgical correction for a case with the AA, IAA type B, VSD and an aortopulmonary window (APW) as a direct source of blood to the coronary arteries, Asc.Ao and proximal aortic arch.

Case Presentation

After an uncomplicated gestation of 38 weeks, a male infant weighing 2.9 kg delivered by spontaneous vaginal delivery, and became cyanotic immediately after delivery, in the first hour, then turned blue with a drop in oxygen saturation to 75%. The patient was given nasal prongs for oxygenation, but the oxygen saturation did not improve. An echocardiogram was ordered immediately. The cardiac examination was normal, except for a drop in oxygen saturation. The patient had no dysmorphic features. The chest X-ray was unremarkable. The transthoracic echocardiography (TTE) confirmed the presence of AA, type B IAA, and malalignment VSD. Both ventricles were evaluated and the result was that they were of normal size. It was also revealed that there was an abnormal right subclavian artery (RSA) from the descending aorta (DAO) and suspicion of APW (Fig. 1). The patient was started on prostin 0.05 mcg/kg/min and was immediately intubated and mechanically ventilated. Multidetector computed tomography (MDCT) revealed an aberrant right subclavian artery of the DAO and suspected APW (Fig. 2A–C). The decision was made to perform surgical correction on the second or third day. The Norwood-Rastelli biventricular repair was the approved therapeutic option by the discussion meeting of cardiology and cardiac surgery teams. Informed consent obtained from the parents and all steps and risks were explained. Surgery performed when the baby was 6 days old. During surgical repair and careful inspection, it was found that Asc.Ao (4 mm) supplies both carotid arteries and the anomalous RSV from the proximal DAO (Fig. 3). The conventional cardiopulmonary bypass (CPB) was commenced using the arterial cannula of Asc.Ao and venous cannula of the right atrial appendage. The arterial

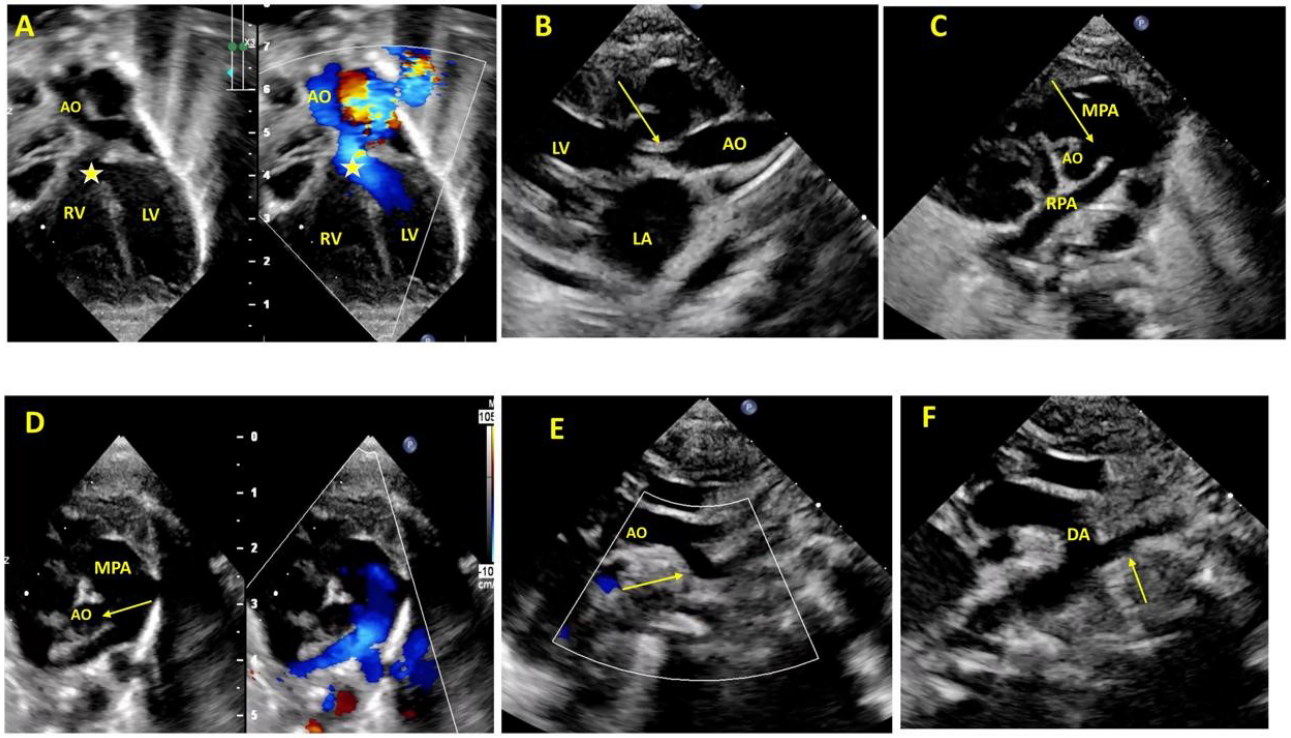

Fig. 1. Preoperative echocardiography. (A) Apical 5 chamber view showing large malaligned subpulmonic VSD (asterisk). (B) parasternal long axis view showing severe LVOTO with the absence of the aortic valve (arrow). (C,D) Parasternal short-axis view showing AP window (arrow). (D) Suprasternal view showing aortic arch interruption (arrow). (E) Suprasternal view showing aortic arch interruption (arrow). (F) Modified suprasternal view showing ductal arch with left subclavian artery arising from the ductal arch (arrow). Abbreviations: AO, aorta; DA, ductal arch; LV, left ventricle; LVOTO, left ventricle outflow tract obstruction; MPA, main pulmonary artery; RV, right ventricle; AP, aortopulmonary; VSD, ventricular septal defect; ARSA, aberrant right subclavian artery; RPA, right pulmonary artery.

cannula was extended to the right carotid artery to maintain antegrade cerebral perfusion. The left and right pulmonary arteries were snared during cooling, aortic cross clamp was applied and antegrade cold blood-based Del-Nido cardioplegia was infused into Asc.Ao. Good coronary perfusion was observed during cardioplegia. We initiated deep hypothermic circulatory arrest (DHCA) to repair the IAA. The ductus arteriosus was treated by ligation and division with removal of ductal tissue. We cut the main pulmonary artery (MPA) where it bifurcates. At the sinotubular junction, Asc.Ao was opened longitudinally and extended into the left carotid artery. We clearly observed a hole about 20 mm in diameter between the posterior wall of the MPA and Asc.Ao and the connection was identified as APW (Fig. 3). The posterior wall of the proximal aortic arch and the distal aortic arch were directly anastomosed. An equine pericardial patch (Matrix patch™) manufactured by Auto Tissue, Berlin GmbH and certified by TÜV Rheinland land grid array (LGA) Products GmbH was used to enhance the entire aortic arch and Asc.Ao. The proximal end of the resected MPA was anastomosed to the repaired aortic arch. The VSD was closed using equine pericardium and through right ventriculotomy (RV). A 12 mm valved xenograft conduit was our choice to connect the RV to the distal MPA. Postoperative echocardiographic assessment revealed good function of both ventricles, non-significant pressure gradients across repaired arch and RV to MPA conduit, outflow obstruction and competent valves (Fig. 2D–G).

Discussion

The association of IAA with AA is a rare congenital heart complex. Survival in these patients is conditional on the presence of blood supply to the coronary circulation [2]. The bilateral patent PDAs [3], the aortopulmonary communications [4], or the collateral of the DAO are direct pathways for coronary artery perfusion [5]. As for patients who do not have direct paths to the coronary circulation, in this case there may be other indirect paths, such as aberrant right subclavian artery (ARSA) arising from the DAO and from it to the coronary circulation, or along the vertebrobasilar circulation and then to the circle of Willis and the right carotid artery, to AscA, and finally to Coronary arteries [6]. In our case, there was a direct connection in the form of APW and ARSA. Single-stage total repair of the

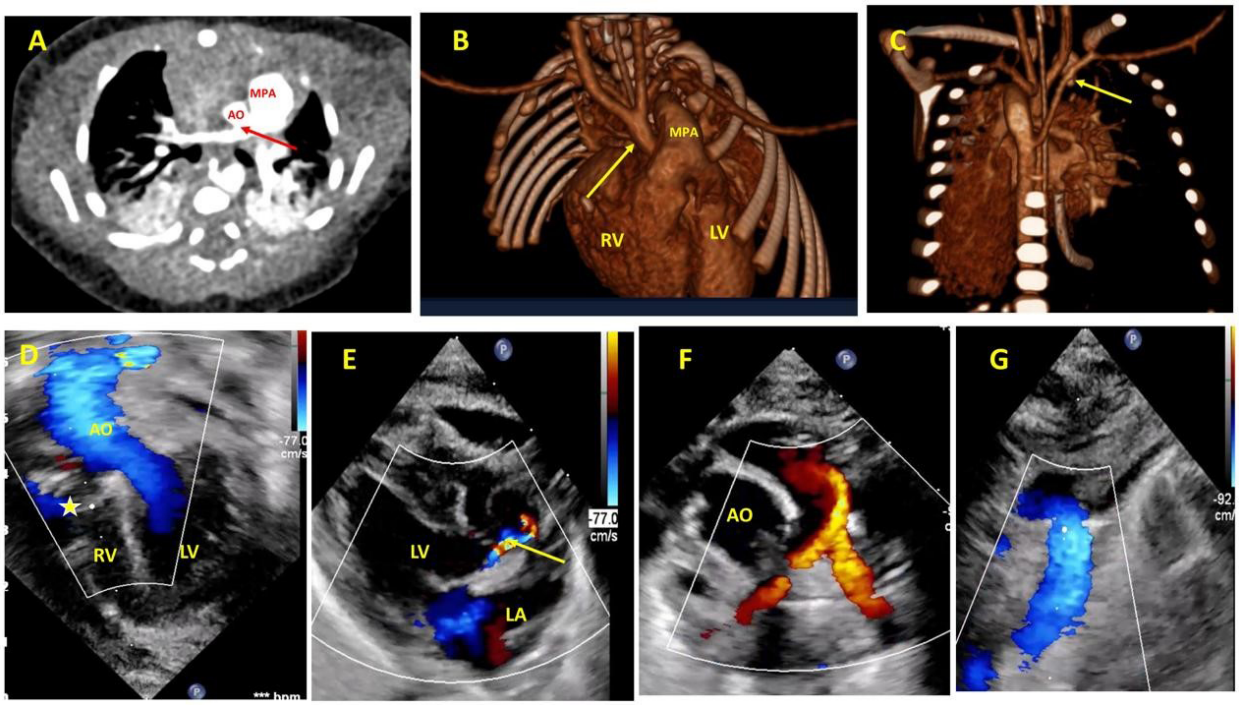

Fig. 2. A preoperative MDCT and postoperative echocardiography. (A) MDCT with an axial cut showing the AP window (arrow). (B) MDCT with three dimensional (3D) reconstruction anterior projection showing dilated MPA with type B interruption of the aortic arch (arrow). (C) MDCT with 3D reconstruction posterior projection showing aberrant right subclavian artery arises distally from the ductal arch (arrow). (D) Postoperative TTE with an apical 4-chamber view showing no residual VSD across the VSD patch (asterisk) with no LVOTO. (E) Postoperative TTE with a parasternal long-axis view showing mild aortic insufficiency (arrow). (F) Postoperative TTE with a parasternal short-axis view showing a patent RV-MPA conduit with moderate to severe pulmonary insufficiency (red flow). (G) Postoperative TTE suprasternal view showing patent aortic arch. Abbreviations: MDCT, multidetector computed tomography; TTE, transthoracic echocardiography.

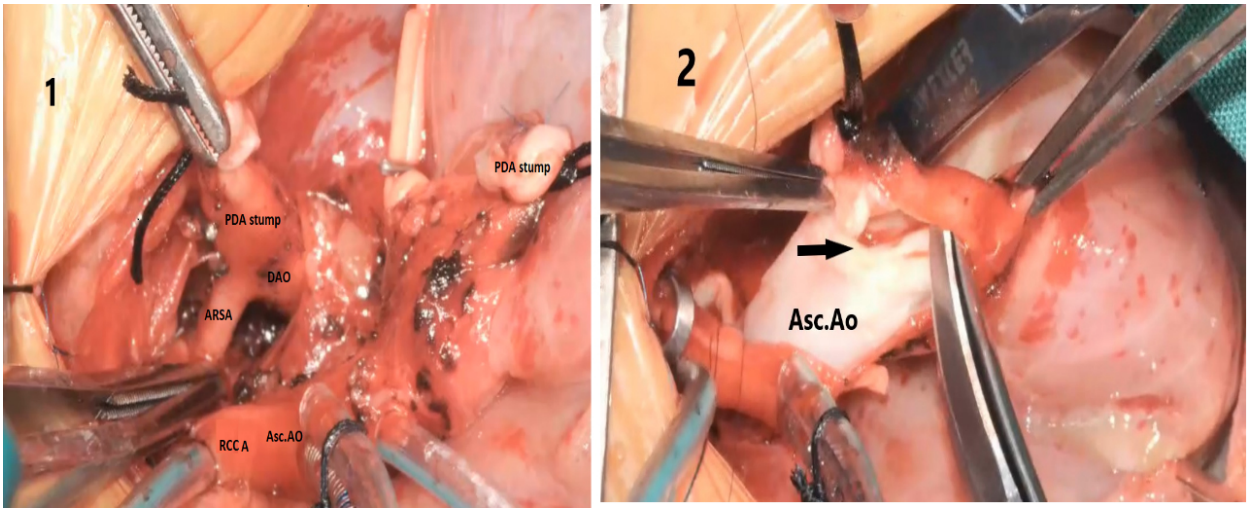

Fig. 3. Intraoperative image. (1) Revealed ARSA coming from the DAO and image. (2) Revealed the communication (black arrow) between the PA and the Asc.Ao (APW). Asc.Ao, ascending aorta; RCCA, right common carotid artery; DAO, descending aorta; ARSA, aberrant right subclavian artery; PDA , patent ductus arteriosus; APW, aortopulmonary window.

IAA in neonates has become a widely available procedure with good results. Many and varied cases of this uncommon defect of IAA with AA has been reported and different procedures for its management have been registered, which include univentricular type, biventricular type, single-stage and multi-stage repairs. Among these recorded operations,

Norwood and Stelin [7] reported successful surgical repair using a conduit from the apex of the left ventricle to the DOA, and repair of the IAA by graft with pulmonary artery ligation. VSD was later closed. Tannous and colleagues [8] in 2006 performed a total surgical correction in a patient with IAA type B and AA. As for the Yasui operation technique, it allowed a single-stage operation to perform total biventricular repair of this rare complex congenital defect [9]. Yang EL *et al*. [10] stated that one stage repair is currently increasing in neonates with good outcomes and they selected Yasui operation technique with arch repair. Also, Soynov *et al*. [11] reported the same concept that the Yasui procedure in patients with aortic atresia and interrupted aortic arch can be performed with minimal complications, even in low-weight patients. In our case, we did the same, which is aortic arch repair for IAA, and adopting Yasui procedure to repair the AA, but we used another terminology, which is Norwood-Rastelli, believing that Damus-Kaye-Stansel technique to bypass AA with aortic arch repair is like Norwood principle.

Conclusion

Aortic atresia with interrupted aortic arch, the presence of direct or indirect connections with the Asc.Ao is crucial to maintain the coronary circulation. Complete surgical correction of one stage in neonates or low-birth-weight children is now possible and reproducible.

Abbreviations

AA, Aortic atresia; Asc.Ao, Ascending aorta; APW, Aorto-pulmonary window; CPB, Cardiopulmonary bypass; DAO, Descending aorta; DHCA, Deep hypothermic circulatory arrest; MDCT, Multidetector computerized tomography; MPA, Main pulmonary artery; TTE, Transthoracic echocardiography; ARSA, Aberrant right subclavian artery; RV, Right ventricle; VSD, Ventricular septal defect.

Availability of Data and Materials

The data that supports the findings of this study are available in the supporting information of this article.

Author Contributions

Conceptualization and data feedback by ZZ, case report designing and editing by AE, the acquisition of data by NA and AA, analysis by MF and FF, and interpretation of data and revision for the work was done by OAR. All authors have participated sufficiently in the work and agreed to be accountable for all aspects of the work. All authors contributed to editorial changes in the manuscript. All authors read and approved the final manuscript. All authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Ethics Approval and Consent to Participate

The study was carried out in accordance with the guidelines of the Declaration of Helsinki. The study was waived by Unit of Biomedical Ethics, faculty of medicine, King Abdulaziz University, because from a research point of view it is considered a simplified study work and formed consent was granted by the patient for publication.

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Conflict of Interest

The authors declare no conflict of interest.

Supplementary Material

Supplementary material associated with this article can be found, in the online version, at https://doi.org/10.59958/hsf.8067.

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