Surgical Repair of Interrupted Aortic Arch and Interrupted Pulmonary Artery

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Interrupted aortic arch (IAA) is usually associated with ventricular septal defect and patent ductus arteriosus. We report surgical repair in a case of IAA, ventricular septal defect, and interruption of the pulmonary artery with the right pulmonary artery arising from the innominate artery through a separate ductus arteriosus.

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Interrupted aortic arch (IAA) is a rare cardiac malformation [1]. Interrupted and nonconfluent pulmonary arteries have not been reported with IAA. This report focuses on a case of IAA and ventricular septal defect (VSD) with interruption of the pulmonary artery, bilateral ductus arteriosus, and origin of the right pulmonary artery (RPA) from the innominate artery. The surgical technique used to achieve repair in this association of anomalies is described.

A 12-day-old male infant weighing 3.15 kg presented with labored breathing. Di George syndrome was confirmed by fluorescence in situ hybridization test. Echocardiography confirmed type B IAA and perimembranous VSD. The RPA arose through a separate right-sided ductus arteriosus from the innominate artery close to its bifurcation into the subclavian and common carotid arteries. The origin of the RPA was stenotic with a diameter of 2 mm. The distal RPA at the hilum of the lung measured 4 mm. The left pulmonary artery measured approximately 8 mm. The VSD was the perimembranous type, measuring 7 mm.

The infant was promptly scheduled for surgical repair. Through median sternotomy, a patch of pericardium was harvested and treated with gluteraldehyde. Thymic aplasia was noted. The origin of the RPA from the innominate artery was encircled with a vessel loop. After systemic heparinization, an 8F DLP cannula was inserted into a 3.5-mm graft of polytetrafluoroethylene (PTFE) anastomosed to the innominate artery. A second 8F DLP cannula was inserted into the main pulmonary artery and positioned into the ductus arteriosus to perfuse the descending aorta. Venous access was achieved through bicaval cannulation. Cardiopulmonary bypass was initiated, and the branch pulmonary arteries were snared to prevent flooding the lungs. The perfusate was cooled to a nasopharyngeal temperature of 18°C. During the cooling process, the arch vessels and the descending aorta were mobilized extensively. The left pulmonary artery was then mobilized to the hilum of the lung. The origin of the RPA from the innominate artery was interrupted with a vascular clip. It was found that the RPA followed a tortuous course, running laterally posterior to the superior vena cava and entering the hilum of the right lung. The superior vena cava needed to be widely mobilized to retrieve the RPA. The RPA was then divided at its origin, and the mediastinal attachments were freed to get as much length to it as possible. It was found that the vessel was approximately 4 mm at its widest part at the hilum of the lung.

Cardioplegic arrest was achieved, and the snare around the left pulmonary artery was removed. Snares were passed around the arch vessels and tightened, thereby maintaining antegrade cerebral perfusion. The arterial cannula in the ductus arteriosus was removed. A C-shaped vascular clamp was applied to the descending aorta, and the ductus was transected. All ductal tissue from the distal segment was meticulously excised. The undersurface of the proximal segment of the aortic arch was opened longitudinally, extending the incision into the ascending aorta. The descending aorta was brought

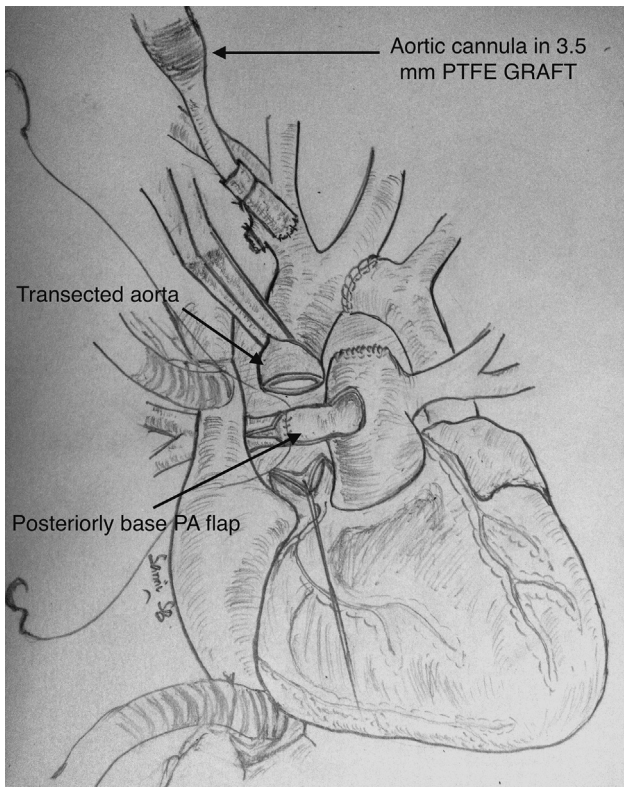

Fig 1. Operative sketch shows the pulmonary artery flap turned medially to the right pulmonary artery (PA). (PTFE = polytetrafluoroethylene.)

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anteriorly, beveled, and anastomosed to the proximal aorta with the use of 7-0 polypropylene suture, thereby achieving a wide and tension-free anastomosis. After this, air was removed from the arch, and the snares around the arch vessels were released. The right atrium was opened, and the VSD was assessed. This was closed with a patch of expanded PTFE and the continuous suture technique.

Having completed the VSD closure, attention was directed to the RPA. The proximal three-quarters of this vessel was of poor quality, resembling ductal tissue, and was required to be excised. It left only the portion immediately proximal to the hilum of the right lung of satisfactory quality to include in the repair. The vessel was found to be of inadequate length to reach the main pulmonary artery. The aorta was transected to gain better exposure. A transverse flap based posteriorly was created from the anterior wall of the main pulmonary artery (Fig 1). This flap of pulmonary artery wall when turned medially and to the right reached the RPA without tension. The flap was sutured to the RPA, thereby creating the posterior wall of the reconstructed RPA. The anterior wall of the distal RPA was then incised longitudinally into the hilar portion to gain the widest area of the RPA. The previously harvested and treated patch of pericardium was then used to reconstruct the anterior wall of the neo-RPA. This created a wide stoma of the RPA from the main pulmonary artery. The transected aorta was re-anastomosed. The child was rewarmed and weaned off cardiopulmonary bypass uneventfully.

Comment

IAA accounts for approximately 1.5% of all congenital cardiac anomalies, and a VSD is the most common associated anomaly seen in approximately 73% of these cases [2]. A PDA is invariably present with IAA, and the descending aorta is a continuation of the ductus.

Besides VSD, IAA may be associated with a variety of anomalies such as truncus arteriosus, transposed great arteries/Taussig-Bing anomaly. There are also modifications to the surgical technique when dealing with these. The results of IAA with these associated anomalies has consistently improved in recent times to the extent that presence of these are not necessarily incremental risk factors [3, 4].

Interruption of the pulmonary artery with bilateral ductus arteriosus has not been reported as an association with IAA. In this case, the RPA arose from the innominate artery through a separate right-sided ductus arteriosus. The proximal three-quarters of the RPA was found to contain ductal tissue, precluding its inclusion in the anastomosis. Wide mobilization of the RPA, the main and left pulmonary arteries facilitate a tension-free anastomosis. Creating a posteriorly and medially based transverse flap from the pulmonary artery ensures a smooth origin for the reconstructed RPA. Being autologous live tissue, it ensures growth in the future. Gluteraldehyde-treated autologous pericardium used to reconstruct the anterior wall of the RPA and the main pulmonary artery where the flap was created from ensures a wide stoma for the RPA. It is of advantage in these cases to transect the ascending aorta to gain adequate exposure. It facilitates anastomosis of the pulmonary artery flap to the RPA and also the reconstruction of the anterior wall with the use of the autologous pericardial patch.

Innominate artery cannulation through a PTFE graft anastomosed to it facilitates the operation by providing adequate space on the ascending aorta and permits continuous cerebral perfusion, thereby avoiding circulatory arrest during the arch reconstruction [5].

In conclusion, IAA, VSD with interruption of the pulmonary artery, and bilateral ductus arteriosus is an extremely rare anomaly. Minor modifications in technique may be required to facilitate a complete repair and is well tolerated.

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