One-stage total repair of anomalous origin of right pulmonary artery from aorta by the double-flap technique, followed by coarctation repair using extended end-to-end arch reconstruction

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ABSTRACT

The anomalous origin of the right pulmonary artery from the ascending aorta combined with coarctation of aorta is a rare congenital malformation. The method chosen for performing a prompt surgery to correct the multiple disease lesions is important. Here we report one-stage surgical strategy which involved a double-flap technique alongside an extended end-to-end arch reconstruction in a newborn baby.

Keywords: Anomalous origin of the pulmonary artery, anomalous origin of the right pulmonary artery, AORPA, arch hypoplasia, double flap technique

INTRODUCTION

The anomalous origin of one pulmonary artery branch from the ascending aorta is a rare congenital heart disease. Its pathology can be associated with other cardiac malformations or aortic arch abnormalities, and is caused by partial or complete developmental failure of the left sixth arch.[1] Owing to this abnormality, a large left-to-right shunt occurs with the other one lung receiving the whole output from the right ventricle, and as such the pulmonary vascular bed of both lungs may be vulnerable to develop pulmonary vascular obstructive disease. The corrective surgery is essential and urgent and must be performed immediately following diagnosis, because mortality rates reach 70% before 6 months of age without surgical intervention.[2]

Here we describe the case of a premature baby with right pulmonary artery arising from ascending aorta (AORPA), aortic arch hypoplasia (coarctation of the aorta, CoA), and patent ductus arteriosus (PDA), and report a new treatment modality which involves using a double-flap technique for AORPA repair first, which then facilitates the subsequent extended end-to-end arch reconstruction.

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CASE REPORT

A newborn male baby with a birth weight of 2100g was sent to the emergency department because of cyanosis and hypoxemia (SpO_2: ~70%), with no obvious heart murmur. After prompt medical treatment with O_2 via nasal continuous positive airway pressure, the respiratory condition of the patient gradually stabilized. Echocardiography showed the AORPA, aortic arch hypoplasia, and one vessel connecting the main pulmonary artery to the descending aorta. Thoracic computerised tomography (CT) angiography was performed, and clearly revealed the right pulmonary artery arising from the posterior wall of the ascending aorta, along with tubular hypoplasia of the aortic arch to the isthmus portion, and large PDA [Figure 1].

Surgical technique

The corrective procedure conducted was a median sternotomy supported by hypothermic cardiopulmonary bypass. The ascending aorta, proximal aortic vessels, main pulmonary artery (MPA) and left pulmonary artery (LPA) were extensively mobilized, and the PDA, LPA, and AORPA were looped before cannulation. The arterial cannula was inserted into the brachiocephalic artery using a 4mm graft connection (W. L. Gore and Assoc, Flagstaff, U.S.). Venous return cannula was then inserted into the right atrium. After the initiation of cardiopulmonary

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bypass, both PAs were snared, PDA was ligated and transacted, and cardioplegia solution was delivered by needle puncture when the aorta was cross-clamped. The double-flap technique for AOPRA repair was then followed [Figure 2]. The ascending aorta was transacted both above and below the RPA origin, which provided a symmetrical large aortic ring almost 1.5 times the RPA diameter. The ascending aorta was reconstructed using end-to-end anastomosis, employing a 6 - 0 prolene suture. Then, a posterior pulmonary flap was created at the MPA anterio-right-lateral surface. According to the length of pulmonary flap, the aortic ring was cut transversely, leaving a small aortic flap to the posterior and an anterior aortic flap was created in order to balance the posterior pulmonary flap. The double-flaps were anatomosed using 6 - 0 prolene to create a new communication between the anomalous RPA and the MPA, which was anterior to the ascending aorta. When the rectal temperature reached

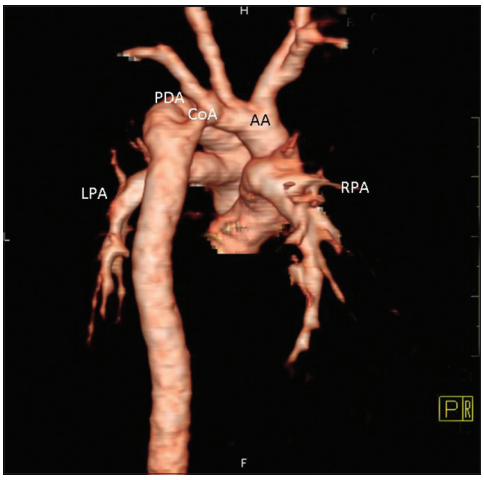

Figure 1: Three-dimensional volume rendering image showing the anomalous right pulmonary artery arising from the right posterior aspect of the ascending aorta, the left pulmonary artery arising normaly from the main pulmonary artery, large patent ductusarteriosus, and arch hypoplasia with coarctation of aorta

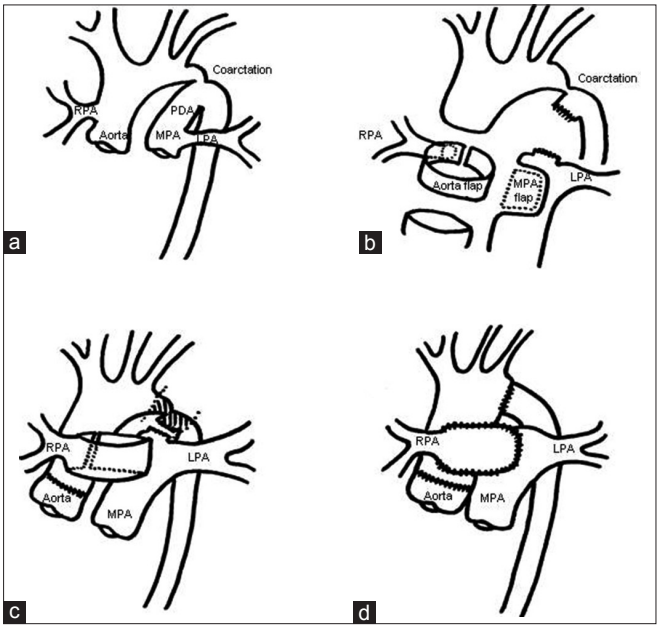

Figure 2: One stage pulmonary double-flap and arch extended end-to-end reconstruction surgery. (a) The diagrammatic section of diseased abnormality. (b) Aortic and pulmonary flap preparation with PDA ligated. (c) The neo right pulmonary artery was created by aortic flap and MPA flap suturing with end-to-end anastomosed ascending aorta caused aorta retract more posteriorly. (d) Under systemic circulatory arrest with selective antegrade cerebral perfusion, extended end-to-end surgical correction for coarctation of aorta

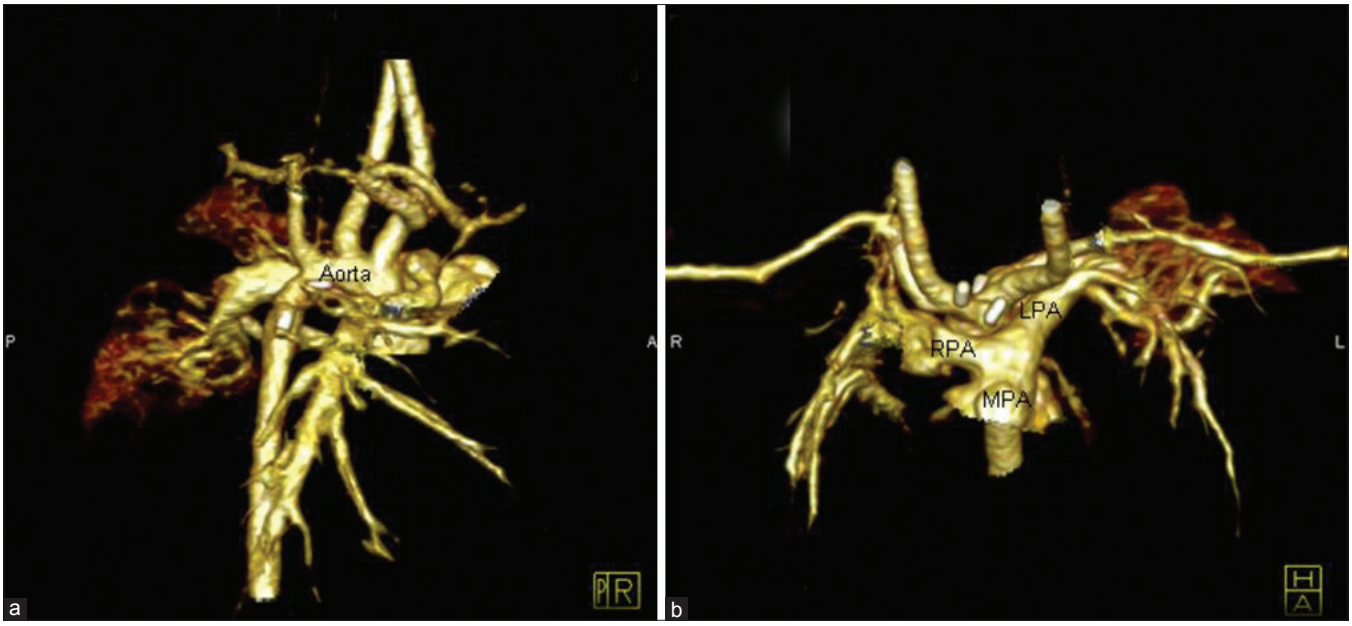

Figure 3: The postoperative three-dimensional volume rendering image illustrated that the reconstructed aortic arch. (a) and neo-right pulmonary artery (b) were smooth and unobstructed

20°C, the descending aorta was also extensively mobilized close to the third spinal artery region. After aggressive ductus tissue resection the extended end-to-end arch reconstruction was performed under circulatory arrest, with selective antegrade cerebral perfusion. The patient was weaned from cardiopulmonary bypass smoothly and was discharged on day 14 post-operation without complications. The follow-up CT angiography six months later illustrated that the reconstructed aortic arch and neo-right pulmonary artery were smooth and unobstructed [Figure 3].

DISCUSSION

The aortic origin of the right or left pulmonary artery is an infrequent cardiac anomaly with an incidence of < 1% of all congenital cardiac malformations. AORPA is 5-6 times more frequent than that of the left pulmonary artery.[1] The presence of AORPA might cause high pulmonary flow, increased circulation of vasoconstrictor substances, neurogenic crossover effects from the unprotected lung, and left ventricular failure, resulting in serious pulmonary vascular disease as early as the third month of life. Therefore, the surgery for the AORPA anomaly needs to be performed earlier in order to avoid pulmonary hypertension and irreversible pulmonary vascular disease.[1,3] This patient was diagnosed even earlier due to the combination of AORPA with arch hypoplasia, resulting in urgent surgery to avoid the further development of pulmonary vascular obstructive disease.

Several surgical strategies can be used for the treatment of AORPA, but the most frequently performed surgery is direct anastomosis of the anomalous pulmonary branch to the MPA.[1,4] However, direct implantation is associated with a higher residual gradient through the anastomotic site, and also with a high reoperation rate. End-to-end anastomosis using a synthetic graft, interposition of a homograft patch, or interposition of an autologous pericardial patch to increase the length of the anomalous branch are all proposed methods when direct implantation is not possible. In this patient, we adapted the double-flap technique[5] to create a new communication line between the MPA and RPA using autologous, viable tissue, which allows for extra tissue lengthening, growth in the future, and a reduced risk of kinking or stenosis of either branch of the pulmonary artery. In addition, the shortened ascending aorta results in movement of the arch both backwards and downwards, which can lessen the anastomotic tension of the reconstructed neo-aortic arch. Nevertheless, long-term regular follow-up is still required for the possible detection of residual anastomosis stenosis.

In conclusion, the double-flap technique for AORPA repair, followed by the extended end-to-end arch anastomosis is a useful strategy for one-stage early repair for this rare disease.

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