Surgical repair for common arterial trunk with pulmonary dominance, hypoplasia of ascending aorta, and interrupted aortic arch

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ABSTRACT

The arrangement of aortic and pulmonary pathways is extremely variable in the hearts with a common arterial trunk. Almost always, interruption of the aortic arch is seen in the setting of hypoplasia of the ascending aorta and dominance of the pulmonary circulation. This subset poses substantial challenges in surgical repair and portends poor outcomes. In this report, we briefly describe the technique of ascending aorta reconstruction and other aspects of the surgical repair of this rare malformation.

Keywords: Congenital heart disease, interrupted aortic arch, truncus arteriosus

INTRODUCTION

A common arterial trunk is a solitary trunk that exits the heart through a common ventriculoarterial junction and supplies directly the systemic, pulmonary, and coronary arterial pathways. It is a rare cardiac malformation and accounts for approximately 0.7% of congenital heart disease.[1] The relative size of the aortic and the pulmonary component relates to the status of the pulmonary blood flow. In patients with an obstructed aortic outflow, as in the setting of coarctation of aorta or arch interruption, the aortic component is hypoplastic while the pulmonary component is enlarged. This unique arrangement is categorized as Type 4 in classification by Van Praagh.[2,3] In a more recent simplified classification, this entity is referred to as having pulmonary dominance.[4]

The coexistence of hypoplasia of the ascending aorta and aortic arch interruption in patients with a common arterial trunk mandates complex surgical repair and predicts high surgical mortality. We hereby describe surgical repair of one such case for brief demonstration of the surgical technique.

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DESCRIPTION

A 1-month-old male infant was admitted with respiratory distress. On evaluation by echocardiography, he was found to have a common arterial trunk which was predominantly committed to the right ventricle (RV). The subtruncal interventricular communication was restrictive while there was an additional small interventricular communication in the midmuscular septum. The aortic arch was interrupted, and the patent arterial duct continued as the descending aorta. Subsequently performed computed tomography confirmed the diagnosis of the common arterial trunk with an interrupted arch [Figure 1]. The truncal valve was trisinusal. The pulmonary component of the common arterial trunk was enlarged while the ascending aorta was hypoplastic. Interestingly, the ascending aorta was arising from the rightward and posterior aspect of the trunk. The pulmonary arteries arose separately from the posterior aspect of the trunk. The orifices of the pulmonary arteries, however, were not widely separated.

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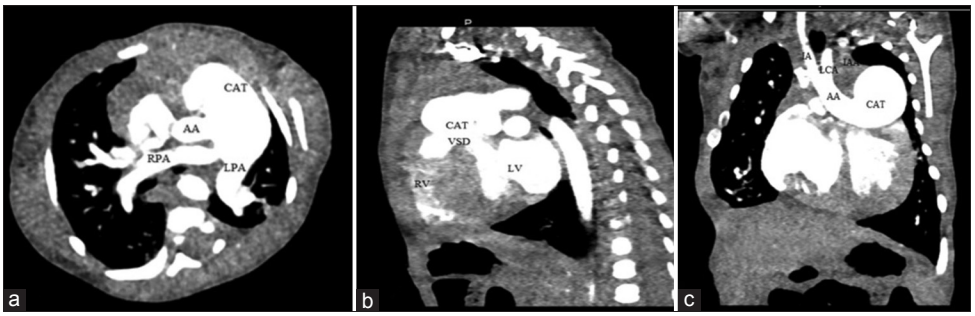

Figure 1: (a) Axial section of computed tomography showing CAT with origin to ascending aorta as a side branch from the right side and also origin of both pulmonary arteries from the posterior aspect of CAT. (b) Sagittal view showing overriding of CAT with RV predominance. (c) Coronal view showing origin of hypoplastic ascending aorta from CAT with Type B IAA. RV: Right ventricle, LV: Left ventricle, CAT: Common arterial trunk, IAA: Interrupted aortic arch, IA: Brachiocephalic artery, LCA: Left common carotid artery, RPA: Right pulmonary artery, LPA: Left pulmonary artery

The morphoanatomical details were well illustrated using virtual dissection of the computerized tomographic datasets [Figure 2]. There was a single coronary artery arising from the aortic component of the trunk. The aortic arch was left sided and was interrupted proximal to the left subclavian artery. The child was positive for 22q11 deletion.

During the hospital stay before the surgical repair, the arterial duct started constricting which was managed successfully using prostaglandin infusion.

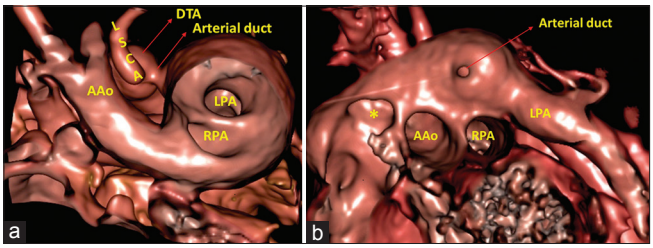

Figure 2: Virtual dissection images obtained from computed tomographic angiography in anteroposterior (a) and lateral projection (b) showing common arterial trunk with pulmonary dominance. The ascending aorta is hypoplastic and arises from the right side of the trunk, and the aortic arch is interrupted proximal to the left subclavian artery. A moderate size patent arterial duct joins the pulmonary component of the trunk with descending thoracic aorta. Separate origin of the left pulmonary artery and the right pulmonary artery is also well seen along with an outpouching (*) proximal to the origin of ascending aorta

SURGICAL PROCEDURE

The surgical approach was through a standard median sternotomy. The thymus was completely excised. The brachiocephalic vein was dissected and looped. The pericardium was harvested and fixed in 0.6% glutaraldehyde. On opening the pericardium, an arterial trunk was seen arising from RV. Aorta was arising from the right and posterior aspect of this single arterial trunk. Just after the origin, the aorta pursued a horizontal course for approximately 1 cm, after which it ascended up and divided into right brachiocephalic and left common carotid artery. The ascending aorta was hypoplastic, and the aortic arch was interrupted beyond the left common carotid artery [Figure 2a and b]. The right and left pulmonary arteries were seen to originate from the arterial trunk close to the origin of the aorta.

A 5-cm long piece of homograft saphenous vein was anastomosed to the brachiocephalic artery. This vein was then cannulated using an 8-Fr Biomedicus cannula for arterial inflow. Bicaval cannulation was achieved by cannulation of the superior caval vein through the right atrial appendage with separate cannulation of the inferior caval vein. Cardiopulmonary bypass was commenced with simultaneous snaring of both pulmonary arteries. Another aortic cannula was inserted through the common arterial into the patent arterial duct for distal body perfusion. Core cooling was accomplished till 18°C. A right superior pulmonary vein vent was placed. After the nasopharyngeal temperature reached 18°C, the aortic cross-clamp was applied and a single dose of del Nido cardioplegia (20 ml/kg) was delivered into the aortic root. Total circulatory arrest was established. As a matter of individual preference, total circulatory without antegrade cerebral perfusion was used. The aortic cross-clamp was removed. Following this, the aorta was incised at the base of brachiocephalic artery, continued to the origin of ascending aorta and also into the dilated structure stopping just short of the sinotubular junction. A common valve was encountered between the ventricle and the aorta with three normal cusps. The ductal tissue was excised completely, and after complete mobilization, the descending aorta was anastomosed end to side to the ascending aorta close to take off of the brachiocephalic artery. The pulmonary arteries were mobilized, and their origin was excised as a single button. This aortic suture line was interrupted at both the ends, and posterior layer of the anastomosis was completed. Anterior part of ascending aorta that had been incised onto the dilated structure was then reconstructed by suturing the fixed pericardial patch. The resulting effect was that the ascending aorta was

augmented to 1-cm size [Figure 3]. Following this, the aortic cross-clamp was reapplied, and perfusion was restarted. A standard right ventriculotomy was made below the origin of the dilated structure taking care not to damage the outlet valve. A small 6-mm restrictive interventricular communication was visualized in the upper part of the outlet septum while another 5-mm interventricular communication was visualized in the upper part of the muscular septum. The subtruncal interventricular communication was then enlarged anteriorly and inferiorly to approximately 2 cm in size to ensure an unobstructed left ventricular outflow tract toward the aorta. The resultant interventricular communication that had been now enlarged was closed with a continuous suture using a Dacron patch to route the left ventricle to the aortic valve. The additional interventricular communication was closed directly using interrupted pledgeted sutures. Now, a no: 12 bovine internal jugular vein conduit (Contegra, Medtronic Inc., Edwardsville, IL, USA) was sutured to establish the RV–pulmonary artery (RV-PA) continuity [Figure 3]. The steps of surgical repair are summarised in Figure 4. After gradual rewarming, the patient was weaned off cardiopulmonary bypass. Intraoperative transesophageal echocardiography demonstrated an unrestricted left ventricle to aorta communication and unobstructed RV-PA – continuity without any residual interventricular communication. There was no blood pressure difference between the upper and lower extremities signifying satisfactory repair of the arch interruption.

Circulatory arrest, aortic cross-clamp, and cardiopulmonary bypass times were 25, 62, and 144 min, respectively. The chest was left open, and a piece of sterile blood bag was used to cover the chest temporarily.[5] Postoperatively, the patient continued to make an uneventful recovery up to 8 h following which sudden bradycardia and hypotension ensued. Immediate cardiopulmonary resuscitation was performed, and the patient was placed on extracorporeal membrane oxygenator support. The patient was weaned off the latter support after 2 days. Postoperative hemodynamics was stable, but the child succumbed to sepsis on the 7th postoperative day.

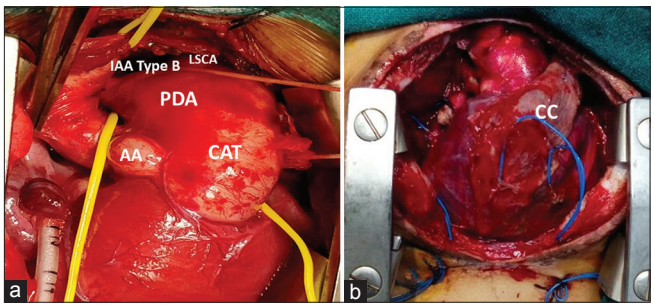

Figure 3: (a) Intraoperative photograph showing CAT with pulmonary dominance and hypoplastic ascending aorta and Type B IAA. CAT continuing as patent arterial duct. (b) Postoperative photograph showing reconstructed ascending aorta and Contegra RV-PA conduit (CC). AA: Ascending aorta; CAT: Common arterial trunk; IAA: Interrupted aortic arch; LSCA: Left subclavian artery; PDA: Patent ductus arteriosus; RV-PA: Right ventricle - pulmonary artery

DISCUSSION

The common arterial trunk has been traditionally classified based on the arrangement of pulmonary arteries. The first classification by Collett and Edwards,[6] however, did not provide a category for patients with coexisting interruption of the aortic arch. Subsequently, Calder *et al.* proposed their classification based on a review of 100 heart specimens with common arterial trunk.[2] They featured this combination in their Type 4. The difficulties in exact categorization as Type 1 or Type 2 remain challenging in many cases, and this has prompted some to propose a simplified classification system.[4]

Once again, based on the study of the heart specimens, Russell *et al.*[4] proposed two broad categories based on relative sizes of the aortic and pulmonary component. Our patient fits in "Type A4" category of Van Praagh and "pulmonary dominance" category of the classification proposed by Russell *et al.*[4] The ascending aorta is hypoplastic in this subset of patients while the pulmonary component dominates.

Common arterial trunk with interruption of the aortic arch is traditionally considered a high-risk surgery with high operative mortality. Even in the modern era, despite some reports suggesting excellent outcomes, operative mortality remains 10%–20%. It is related to long and complex repair with long cardiopulmonary bypass times, often a substantial duration of total circulatory arrest and a prolonged postoperative period with sepsis. The commitment of the trunk to the left ventricle favors surgical repair. The commitment of the trunk to the RV, as was seen in our case, will evidently make the left ventricular outflow tract long and tortuous. In addition, the interventricular communication is shown to be restrictive in patients with commitment of the trunk to the RV.

Despite growing experience of repairing common arterial trunk, the surgical repair of this variant is challenging. In brief, the surgical technique involves dividing the arterial duct and the common arterial trunk with a pulmonary cuff, aortic arch reconstruction performed under deep hypothermia and circulatory arrest or antegrade cerebral perfusion using direct arch anastomosis and patch augmentation of the hypoplastic ascending aorta. During rewarming phase of the surgery, the RV is incised to close the interventricular communication. Finally, the anatomic continuation is established between the RV and the "new" PA trunk using a direct anastomosis without a conduit or more commonly using a homograft or artificial conduit.

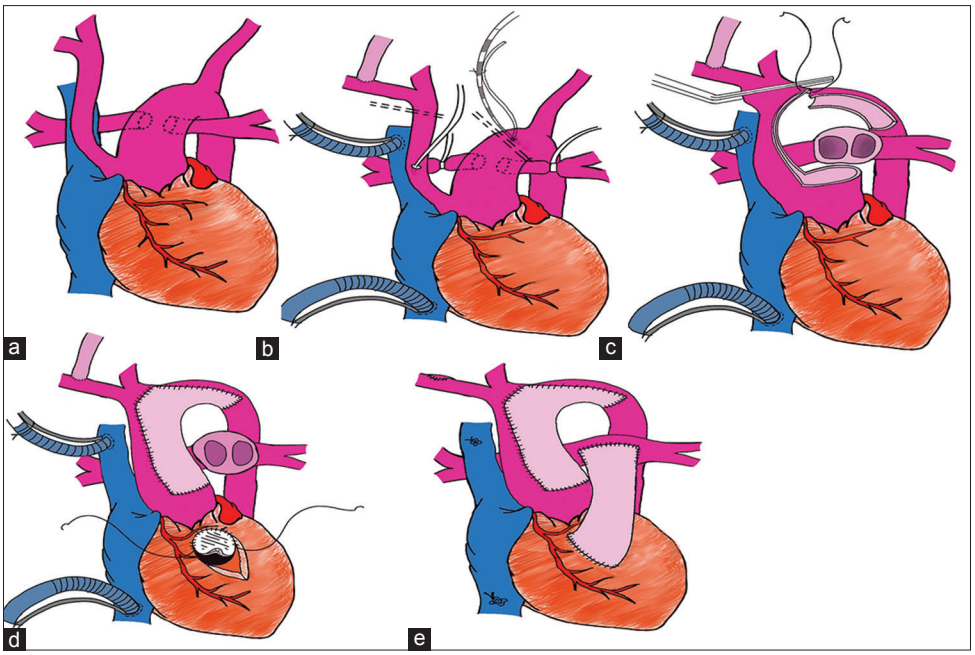

Figure 4: Cartoon showing steps of surgical repair

The poor prognosis for patients with a common arterial trunk and interrupted aortic arch has been confirmed by the recent Congenital Heart Surgeons Society study with an overall survival of 39% at 1 year and 30% at 15 years.[7] Survivors face reinterventions not only to replace the implanted small homografts but also, in a good number, to relieve postoperative obstruction of the aortic arch. Debate still persists as how best to achieve aortic arch reconstruction to prevent reinterventions in future.

The association of common arterial trunk and interrupted aortic arch increases the perioperative risk of death. Despite some reports with a relatively short follow-up showing encouraging results,[8,9] the operative mortality rates remain high (between 10 and 20%)[7-9] compared to the early outcomes of repair of common arterial trunk without arch interruption. When common arterial trunk is associated with an interrupted aortic arch, freedom from reoperation is even worse and is estimated at 69% at 1 month, 54% at 3 years, 30% at 5 years, and 11.1% at 10 years after the first operation.[9] Jahangiri *et al.*[10] reported that the actuarial survival was 96% at 30 days, 1 year, and 3 years. There were no deaths in patients with associated interrupted aortic arch. They report that despite the magnitude of the operation, excellent results can be achieved in complex forms of common arterial trunk and they recommend aggressive application of truncal valvuloplasty methods to neutralize the traditional risk factor of truncal valve regurgitation.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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