Successful surgical repair of pulmonary dominant common arterial trunk without aortic arch obstruction in a neonate

Harikrishnan Nair[1], John Valliattu[1], Raghavannair Suresh Kumar[1], Robert H. Anderson[2], Sajith Sulaiman[1], Ashwin Varghese Alexander[3]

[1]Believers International Heart Centre, Believers Church Medical College Hospital, Thiruvalla, Kerala, India, [2]Biosciences Institute, Newcastle University, Newcastle-upon-Tyne, United Kingdom, [3]Department of Radiology, Believers Church Medical College Hospital, Thiruvalla, Kerala, India

ABSTRACT

The pulmonary dominant variant of the common arterial trunk has always been reported to be associated with aortic coarctation, or interruption of the aortic arch, along with a duct-dependent systemic circulation. This mandates a complex surgical repair with attendant high surgical mortality. We report a 23-day-old baby with a pulmonary dominant trunk with mild aortic hypoplasia but with an arch free of coarctation or interruption, who underwent successful surgical repair. In the preoperative evaluation of a common arterial trunk, pulmonary dominance may not necessarily denote an adverse risk factor when the aorta is only mildly hypoplastic.

Keywords: Aortic arch, congenital heart surgery, genetics, truncus arteriosus communis (common arterial trunk)

INTRODUCTION

Common arterial trunk is a solitary trunk exiting the heart through a common ventriculoarterial junction and directly supplying the systemic, pulmonary, and coronary arterial pathways.[1] When proposing that the lesion was best classified on the nature of the systemic pathways as aortic or pulmonary dominant, Russell *et al.* noted that the feature of the pulmonary dominant variant was hypoplasia of the ascending aorta, with either interruption of the aortic arch or aortic coarctation, the combination providing a formidable surgical challenge. We report the successful surgical repair of an unusual case of a pulmonary dominant trunk in the absence of coarctation or interruption, despite some degree of hypoplasia of the ascending aorta.

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CASE REPORT

A prenatally diagnosed baby (length 46 cm, weight 3.4 kg) with common arterial trunk was admitted to the neonatal intensive care unit with respiratory distress on day 15. The fluorescence *in situ* hybridization test was negative for 22q11 deletion. Echocardiography revealed a common arterial trunk striding a large subtruncal interventricular communication. The trunk arose predominantly from the right ventricle, continuing as the pulmonary arterial segment, which branched to supply the right and left pulmonary arteries at the margins of the pericardial cavity. The tricuspid truncal valve was mildly regurgitant. Computed tomographic angiography revealed the intrapericardial ascending aorta to be a small side branch

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Address for correspondence: Dr. Raghavannair Suresh Kumar, Believers International Heart Centre, Believers Church Medical College Hospital, Thiruvalla - 689 103, Kerala, India.
E-mail: r.sureshkumar.mmm@gmail.com

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of the common trunk (diameter of 6 mm). The arch was unobstructed (diameter 3.4 mm), and a small nonobligate duct was found communicating with the left pulmonary artery [Figure 1 a-c and Supplementary Material 1].

Surgical procedure

The baby underwent corrective surgery through a median sternotomy on the 23rd day of life. On opening the pericardium, there was a large pulmonary dominant common trunk, with a small ascending aorta arising as a side branch, with no evidence of coarctation or interruption. The right and left pulmonary arteries originated from the left and posterior aspect of the trunk, away from truncal valvar sinuses. The small arterial duct was suture closed. Both pulmonary arteries were mobilized. With aortic and bicaval cannulation, cardiopulmonary bypass was established, cooling the patient to 28°C. Both pulmonary arteries were snared, the aorta was cross-clamped, and del Nido cold blood cardioplegia solution was infused antegrade. The caval veins were snared and the right atrium opened obliquely, venting the left atrium through the atrial septal defect. The trunk was opened obliquely, and the origins of both pulmonary arteries excised with a supporting cuff. The aorta was closed primarily with Prolene 5-0 sutures. Through a right ventriculotomy, hypertrophied septoparietal trabeculations were resected and the large subtruncal interventricular communication was closed with a 0.4-mm Gore-Tex patch using continuous Prolene sutures. A size 12 Contegra pulmonary valved conduit (Medtronic, MN, USA) was anastomosed distally to the cuff supporting the pulmonary arteries. The atrial septal defect was closed primarily with a continuous suture, leaving a tiny defect superiorly. After de-airing the left heart and removing the cross-clamp, the proximal end of the conduit was anastomosed to the right ventriculotomy [Figure 2a-c]. The right atriotomy was closed with continuous Prolene sutures, and the patient was weaned from bypass. Standard chest closure was performed. The baby was extubated after 48 h and discharged 14 days later.

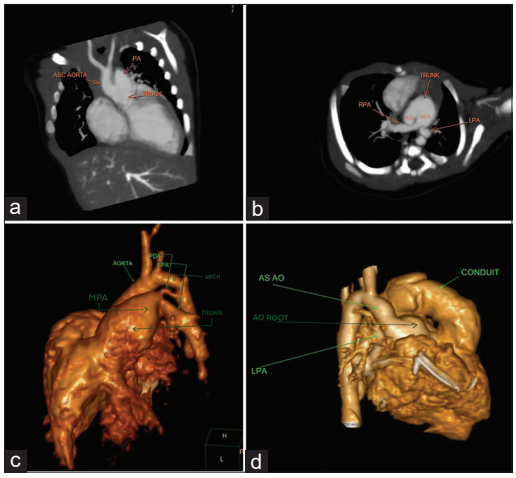

Figure 1: Computed tomography angiogram images showing the anatomic features before (a-c) and after (d) surgical repair of the common arterial trunk. (a) Oblique coronal view showing the common arterial trunk giving off the large main pulmonary artery and the small aorta. (b) Oblique axial view showing the common arterial trunk giving off the large main pulmonary artery and the small aorta. RPA appears to be in direct continuation of the main pulmonary artery, while the LPA origin is in a different plane. (c) Volume-rendered oblique frontal image showing all the anatomic features of this common arterial trunk-small aorta to the right of the huge main pulmonary artery, adequate aortic arch, small nonobligate arterial duct. (d) Volume rendered oblique frontal image after surgical repair. Note the conduit to the left of aorta and the discrepancy between the neo-aortic root and the ascending aorta. Ao: Aorta, MPA: Main pulmonary artery, PDA: Patent ductus arteriosus, RPA: Right pulmonary artery, LPA: Left pulmonary artery

Postoperative echocardiogram showed an intact ventricular septal patch and smooth flow into the conduit and pulmonary arteries. Repeated computed tomographic angiography confirmed the adequacy of the repair [Figure 1d and Supplementary Material 2], albeit with mild stenosis at the origin of the left pulmonary artery.

DISCUSSION

A common arterial trunk is a solitary trunk that exits the heart through a common ventriculoatrial junction and

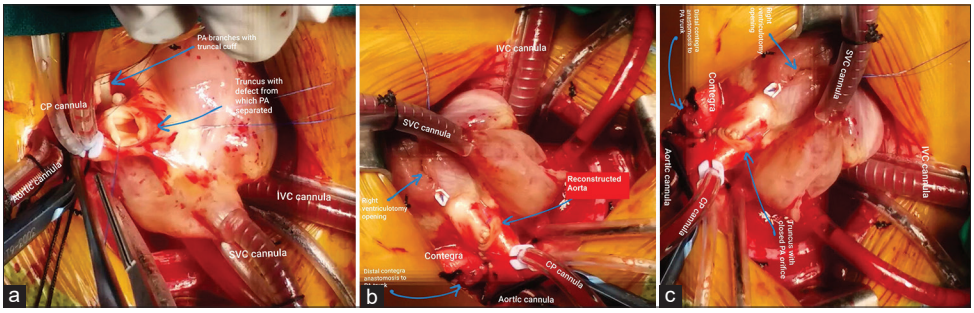

Figure 2: Operative photographs showing (a) truncal root, from which pulmonary artery cuff has been removed. Annotations show anatomic landmarks. Cardiopegia cannula marks the small aorta to the right of the transected truncus. (b) Completed aortic anastomosis. Note right ventriculotomy for ventricular septal defect closure and conduit placement. (c) Completed distal conduit anastomosis. Proximal conduit anastomosis being performed

supplies directly the systemic, pulmonary, and coronary arterial pathways.[1] Surgeons have emphasized the inadequacies of the two popular classification systems, namely those of Collett and Edwards[2] and Van Praagh and Van Praagh,[3] in providing a common language of communication and in identifying meaningful differences in natural history, risk stratification, and surgical approaches.[1] Russell *et al.* proposed classification of the lesion as aortic dominant or pulmonary dominant, depending on which great vessel was dominant in the intrapericardial part of the common arterial trunk, this being a simple means of highlighting the major anatomic features and defining the single most important risk factor for surgical outcome. They found pulmonary dominance only when the aortic component of the trunk was significantly hypoplastic and an arterial duct supplied the majority of flow to the descending aorta. Only in this setting, pulmonary arteries were found to arise from the sides of the major pathway, with the aortic component discrete from the pulmonary component within the pericardial cavity.[1] More recently, Gupta *et al.* also reported arch interruption or coarctation in all of their patients identified with the pulmonary dominant variant.[4] Our case had a pulmonary dominant trunk with a mildly hypoplastic aorta, but no interruption or coarctation. The duct was small, but not part of an obligate circulation. This anatomy was thus an exception to the concept advanced by Russel *et al.*, and it permitted conduit repair without any major problem. In preoperative evaluation of a common arterial trunk, pulmonary dominance may not necessarily denote an adverse risk factor when the aorta is only mildly hypoplastic. 22q11 deletion is noted in around one-third of patients diagnosed with common arterial trunk.[5]

This subset is known to have a higher incidence of other cardiac anomalies, including interruption of the aortic arch.[5] Interestingly, the present case was negative for this genetic abnormality. We speculate whether, in our patient, the absence of the genetic deletion has any causal relation with the absence of obstruction in the aortic arch. Hypoplasia of the aorta could occur in any common trunk with a large pulmonary arterial component, as is the case with any cardiac lesion with increased pulmonary blood flow during fetal life.

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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Nil.

Conflict of interest

There are no conflicts of interest.

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SUPPLEMENTARY FILES DESCRIPTION: (VIDEO-MP4)

Supplementary material 1: Trunk-pre-op-My Movie

Rotational Computed Tomographic angiogram showing the features of CAT as illustrated in Figure 1C

Supplementary material 2: Trunk post-op -My Movie

Rotational Computed Tomographic angiogram showing the post-operative anatomy as illustrated in Figure 1 D.