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Journal of Cardiac Critical Care TSS

Surgical Technique *Cardiac Critical Care*

Technical Details of Anatomic Restoration of an Isolated Anomalous Origin of the Right Pulmonary Artery from the Ascending Aorta into the Pulmonary Trunk: A Video Presentation

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Received: 31 August 2024
Accepted: 09 September 2024
Published: 24 October 2024

DOI
10.25259/JCCC_42_2024

Video available online at
https://dx.doi.org/10.25259/JCCC_42_2024

Quick Response Code:

ABSTRACT

We present here-in a 3-month-old male child with anomalous origin of the right pulmonary artery from the ascending aorta with a left-sided persistent arterial duct who underwent division of the anomalous right pulmonary artery, re-implantation of the same in the pulmonary trunk with concomitant ligation of the arterial duct Post-operative recovery was uneventful.

Keywords: Anomalous origin of the right pulmonary artery from the ascending aorta, Hemitruncus, Aortopulmonary septal defect

INTRODUCTION

Described first in 1868, the anomalous origin of a pulmonary artery from the ascending aorta commands substantial interest due to the rarity of this conotruncal malformation and expectation of a nearly normal life expectancy for survivors of repair in infancy.[1-6] The malformation, also known as hemitruncus and aortopulmonary septal defect type III, accounts for 0.12% of all congenital heart diseases.[7] Nearly, all cases involve the right pulmonary artery.[5,8]

It occurs as an isolated lesion in 20% of cases.[1-6,9] The most common co-existing lesion is persistent arterial duct, present in about 50% of cases.[5] Origin of the left pulmonary artery from the ascending aorta is rare. The anomalous origin of the left pulmonary artery occurs as an isolated lesion in about 40% of cases, usually co-existing with right aortic arch.[7,10-14] Other less common associated cardiac anomalies include ventricular septal defect, aortopulmonary window, coarctation of aorta, interrupted aortic arch, tetralogy of Fallot, and atrial septal defect.[8,15-20] Severe contralateral left pulmonary vein stenosis may co-exist.[8,15-20]

The right pulmonary artery origin is rarely stenosed and vessel is usually as large or larger than the normally connected left pulmonary artery.[21,22] Despite differences in the origin of the pulmonary arteries, pulmonary hypertension in both the normally connected and anomalously connected pulmonary arteries are uniformly present.[10,11,23-26]

Anomalous origin of the right pulmonary artery from the ascending aorta in the presence of separate aortic and pulmonary valves without interposition of ductal tissue is the result of severe unequal partitioning of the aortopulmonary trunk by conotruncal ridges.[9] Till 2010, 171 cases of anomalous origin of the right pulmonary artery and 77 cases of anomalous origin of the left pulmonary artery have been reported.[6,27,28]

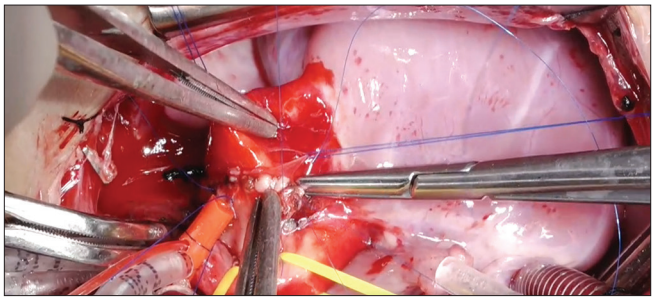
Diagnosis of this disease entity in infancy is an indication of an urgent operation. The operation is contraindicated in late presenters with excessive elevation of pulmonary vascular resistance in the normally connected lung.[5] The difficulty lies in assessing resistance in the anomalously connected lung before surgery. A nuclear lung perfusion scan in combination with data from cardiac catheterization may be helpful to calculate independent pulmonary vascular resistance in each lung.

About 70% of untreated patients are dead by 6 months of age and 80% by 1 year of age. Intractable cardiac failure is the usual cause of death.[3,29-38] Survivors of repair in infancy have normal pulmonary artery pressure late postoperatively and presumably a normal life expectancy.[3,4] Literature documents two isolated reports of satisfactory reduction of the right and left pulmonary artery pressure after surgery, operated at an age of 20 years and 23 years, respectively.[37,38]

SURGICAL TECHNIQUES

Position and surgical approach

Following median sternotomy, the thymus was subtotally excised taking care not to expose the brachiocephalic vein. The pericardium was incised to the right of the midline with scissors and not cautery to avoid inadvertent cautery-induced ventricular fibrillation. Note the tense distended right pulmonary artery arising anomalously from the left posterolateral side of the mid-ascending aorta [Video 1].



Video 1: Position and surgical approach of the right hemitruncus.

Aortic and venous cannulation, dissection of the ascending aorta, and main pulmonary trunk

The operation was performed with mild hypothermic cardiopulmonary bypass at 32°C. Angled venous cannulas were inserted into the superior and inferior caval veins and distal ascending aortic cannulation. The ascending aorta and main pulmonary trunk were separated from each other as much as feasible without compromising hemodynamics.

Ligation of the persistent arterial duct

The persistent arterial duct was dissected on the superior surface of the pulmonary artery and ligated in continuity using No. 2 ductus silk.

Mobilization of the right pulmonary artery and isolation of the aorta and right pulmonary artery

The ascending aorta and pulmonary trunk were dissected using low voltage cautery (15–20 MV) avoiding injury during dissection. The right pulmonary artery was subsequently mobilized along its entire intrapericardial course. The aorta and right pulmonary artery were looped using elastomer vessel loops.

Cross clamping of the aorta and right pulmonary artery; administration of antigrade root cardioplegia

The aorta and right pulmonary artery were individually cross-clamped and antegrade St. Thomas-based (1:4) cold blood cardioplegia was administered into the aortic root. Topical cooling was additionally used for myocardial preservation.

Venting of the cardiac chambers

A small right atriotomy was done after snugging the inferior caval vein for decompression of the right heart chambers. The left heart was vented through a small atrioseptostomy and insertion of a No.13 DLP suction vent (Medtronic Inc., Medtronic Parkway N.E., Minneapolis, MN, USA).

Transection of the right pulmonary artery at its origin from the ascending aorta

The right pulmonary artery was transected at its aortic origin 5-6 mm away from its origin to facilitate later repair without causing aortic narrowing and distortion.

Repair of the aortic window and mobilization of the proximal end of the right pulmonary artery

The aortic opening was subsequently sutured in two layers, using 6-0 polypropylene suture (Johnson and Johnson Ltd., Ethicon,

LLC, San Lorenzo, USA), avoiding iatrogenic narrowing and distortion. The proximal one-third of the right pulmonary artery was mobilized to facilitate tension-free suturing.

Creation of a new window on the side of the main pulmonary trunk

The right lateral border of the proximal pulmonary trunk was opened in between stay sutures at an appropriate location, creating a similar-sized opening as that of the transected right pulmonary artery.

Reimplantation of the transected right pulmonary artery into the neopulmonary trunk

The transected end of the right pulmonary artery was spatulated and matched to the opening of the pulmonary trunk to prevent anastomotic narrowing. Subsequently, the transected end of the right pulmonary artery was anastomosed to the opening of the neopulmonary trunk using 6-0 polypropylene suture.

Deairing of the cardiac chambers and removal of the left atrial vent

After removing the left atrial vent, the left atrium was passively filled with saline for deairing and the atrioseptostomy was suture closed after resuming ventilation.

Closure of the right atriotomy

The right atriotomy was closed in two layers using 5-0 polypropylene suture.

The child was subsequently weaned off cardiopulmonary bypass. After decannulating the cardiac chambers, the chest was subsequently closed in layers.

Short-and long-term results

The child was ventilated overnight and extubated after 12 hours (h). The child required perioperative dobutamine 5 mcg/kg/min and nitro-glycerine 0.5 mcg/kg/min for 72 h. Subsequently, the inotropes were weaned off and discharged home on the 12th postoperative day. Postoperatively, at a median follow-up of 98 months, the child is asymptomatic, on nil medications and not in cardiac failure, with Ross clinical score of 2. Echocardiographically, the child has normal biventricular function without any residual anastomotic gradient.

CONCLUSION

We conclude that division and direct re-implantation of the anomalous right pulmonary artery into the pulmonary trunk is safe, expedient and obviates the need for prosthetic materials. In addition, direct anastomosis maintains viability, grows with age, and helps to avoid thrombosis.

Ethical approval

The Institutional Review Board approval is not required.

Declaration of patient consent

The author certifies that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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How to cite this article: Chowdhury UK, George N. Technical Details of Anatomic Restoration of an Isolated Anomalous Origin of the Right Pulmonary Artery from the Ascending Aorta into the Pulmonary Trunk: A Video Presentation. J Card Crit Care TSS. 2024;8:242-5. doi: 10.25259/JCCC_42_2024