

# Non Confluent Pulmonary Arteries with Bilateral Patent Ductus Arteriosus: Unifocalization and Concomitant Bilateral Bidirectional Glenn Procedure

Sachin Talwar\*, Ashwani Bansal, Dinesh Chandra and Balram Airan

Department of Cardiothoracic and Vascular Surgery, All India Institute of Medical Sciences, New Delhi, India

## Abstract

Bilateral patent ductus arteriosus (PDA) is a rare congenital anomaly and is vital for survival in complex congenital anomaly like non-confluent pulmonary artery (PA), severe coarctation of aorta, interrupted aortic arch and hypoplastic left arch. We report a patient with non-confluent pulmonary arteries associated with pulmonary atresia, double outlet right ventricle (DORV), ventricular septal defect (VSD), bilateral patent ductus arteriosus and bilateral superior vena cavae (SVC). This patient required different surgical strategy due to presence of univentricular physiology and non-confluent PA, for which unifocalization of PA's and bilateral bidirectional Glenn procedure was performed. To best of our knowledge, we have not encountered such a reported patient earlier.

**Keywords:** Unifocalization; Non confluent pulmonary artery; Univentricular heart; Bidirectional glenn

## Introduction

Patent ductus arteriosus (PDA) is a common congenital cardiac lesion usually encountered one side. Rarely both left and right aortic arches may persist giving rise to bilateral PDA [1,2].

Embryologically the distal pulmonary arteries arise from respective lung buds, which join the proximal portion of the sixth aortic arch while the main pulmonary artery is derived from pulmonary arterial portion of truncocoarctic sac. Occasionally the distal and proximal portions of pulmonary artery do not fuse leading to non-confluent pulmonary arteries [2,3].

We recently encountered a patient with bilateral PDA along with non-confluent pulmonary arteries. This patient also had a double outlet right ventricle (DORV), pulmonary atresia, right aortic arch and bilateral superior vena cavae (SVC), further adding to its rarity. The patient underwent a successful unifocalization along with a bilateral bidirectional Glenn anastomosis as a single stage procedure.

## Case Report

A 2 years old male child weighing 8.3 Kg presented with cyanosis since birth. Pertinent clinical findings included presence of central cyanosis and a loud pansystolic murmur on right side of lower sternum. Chest X-ray showed cardiomegaly. Echocardiography revealed DORV, subaortic VSD, pulmonary atresia, non-confluent pulmonary arteries and PDA. The atrioventricular and ventriculararterial connections were concordant. However, the right ventricle was hypoplastic as determined by tricuspid valve Z score of -4.5, thus effectively ruling out a biventricular repair. CT angiography (CTA) was performed to clarify the anatomy further and to estimate the PA size accurately. Echocardiographically estimated pulmonary artery pressure was mean of 10 mm Hg. Besides confirming the echocardiography findings, the CT angiogram revealed discontinuous pulmonary arteries with both the individual right and left pulmonary arteries being supplied by separate right and left PDA respectively. The right PDA was arising from the base of the innominate artery and the left PDA was arising from the base of the left subclavian artery. Both the pulmonary arteries were attached to each other by a 2.5 cm long fibrous band without any luminal continuity. In addition, there were bilateral SVC and right aortic arch. RPA, LPA and descending thoracic aorta measured 6 mm, 6 mm and 10 mm respectively.

Surgical approach was via a standard median sternotomy. The

innominate vein was absent. Right aortic arch and dextrocardia was noted. Intraoperatively measured PA pressure was a mean of 10 mm Hg. Due to malposition of heart; a different cannulation strategy was adopted. Aorta and left-sided IVC were cannulated and cardiopulmonary bypass was instituted. RA appendage was then cannulated. Both right and left PDAs were divided. Right and left non-confluent pulmonary arteries were mobilized completely till the hilum of the lungs and the intervening atretic fibrous band connecting the two was excised taking care to excise completely the ductal tissue. This resulted in a nearly 3 -3.5 cm gap between the two ends of the RPA and the LPA. Now an end-to-end tension-free anastomosis of LPA and RPA was performed. Both LSVC and RSVC were dissected out and looped and the azygous and hemiazygous veins were divided. End to side anastomosis was then fashioned between the RSVC - RPA and between LSVC LPA in a standard fashion after dividing both the SVC one by one and closing their respective cardiac ends. The patient was uneventfully weaned off cardiopulmonary bypass. Total cardiopulmonary bypass time was 94 min. The mean pressure in the Glenn circuit was 6 mm Hg. The systemic saturation at the completion of the procedure was 84% and increased to 88% after removal from mechanical ventilatory support. The duration of latter was 7 hours. Child responded well to procedure and in post operative period, satisfactory oxygen saturation (80-85%) was attained. Child remained hemodynamically stable and was discharged on post-operative day 5. At six months of follow-up, both Glenn circuits are patent and there is equal blood flow into both the lungs. There is no gradient at the site of PA reconstruction.

## Discussion

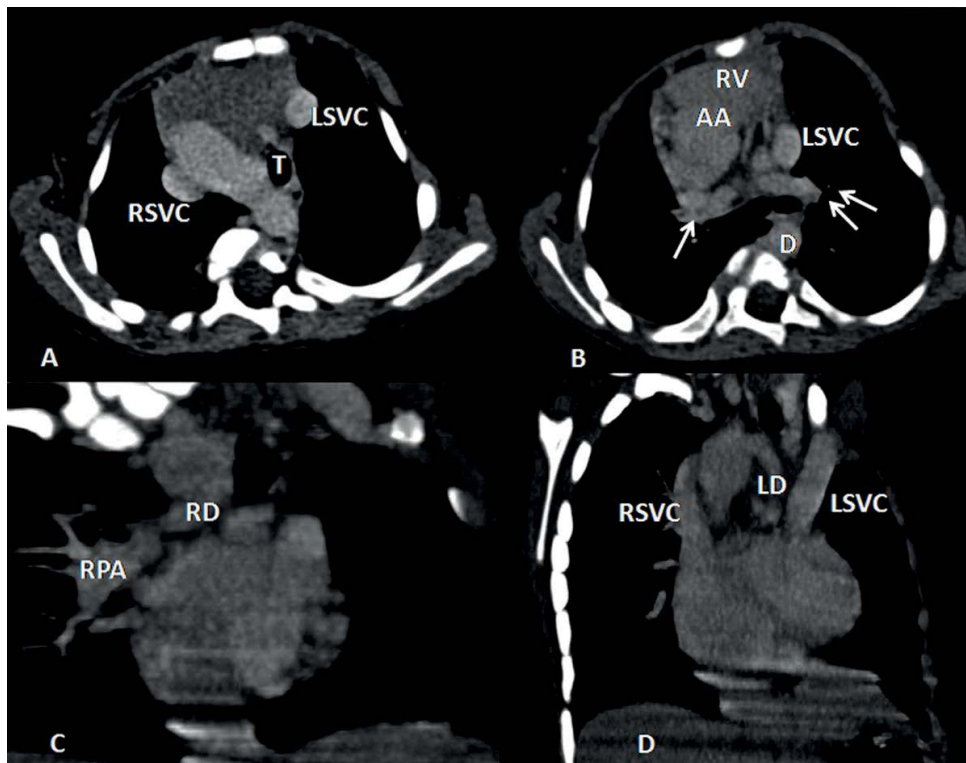
Except for the review by Freedom [4], there have been only sporadic reports mentioning bilateral PDA in association with right arch and/or pulmonary atresia and non-confluent pulmonary arteries [5-8]. In

\*Corresponding author: Talwar S, Mch, Department of Cardiothoracic and Vascular Surgery, All India Institute of Medical Sciences, New Delhi 110029, India, Tel: 91-11-26594835; Fax: 91-11-26588663; E-mail: sachintalwar@hotmail.com

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**Figure 1:** A: Image showing bilateral SVC and left ductus (LD) Axial CT image showing bilateral SVC, T – trachea. B: Axial CT image showing aorta (AA) arising from right ventricle (RV) with non confluent pulmonary arteries [single arrow head showing RPA, double arrow head showing LPA], D – descending thoracic aorta. C: Coronal reformatted CT image showing right ductus (RD) feeding right pulmonary artery (RPA). D: Coronal reformatted CT.

1984 Freedom et al published a study of 27 patients with bilateral PDA and non confluent pulmonary arteries [4], the same group updated their results in 2005 with description of 45 patients over a period of 30 years [9]. Even among the 45 children with ductal origin of the distal pulmonary artery, none of them had complex anatomy like double outlet right ventricle, pulmonary atresia and ventricular septal defect associated with bilateral SVC, non confluent pulmonary arteries and right aortic arch. All these findings in present case report not only increased the surgical complexity but gave us insight into approach of such patients.

An alternative strategy in this patient could have been creation of separate Glenn anastomoses on the RPA and the LPA (unidirectional Glenn) because of fear of narrowing of the reconstructed segment. We targeted to create a palliative cavopulmonary shunt in view of univentricular morphology but a bilateral bidirectional Glenn with pulmonary confluence reconstruction was thought desirable in view of the need for a future completion Fontan operation which would be difficult in the absence of distribution of blood flow to both the lungs. However, successful creation of confluent pulmonary artery was a challenge.

A variety of conduits have been described to recreate continuity between discontinuous pulmonary arteries such as autologous pericardium, equine or bovine heterogeneous pericardium used as tube grafts, homograft aorta or pulmonary artery, and Dacron or polytetrafluoroethylene tube grafts [10]. These conduits have no growth potential. But provision of growth potential to the newly created pulmonary artery was also important for the small child. By completing

a native tissue to tissue reconstruction, we avoided use of any foreign material and performed direct end to end anastomosis after adequate mobilization. Tension free anastomosis was achieved with satisfactory cavopulmonary flow with desired  $PO_2$  (45 mmHg) and saturation (84%).

## Compliance with Ethical Standard

**Funding:** The study did not receive any funding.

**Ethical Approval:** All procedures performed in this study were in accordance with the ethical standards of the institutional and national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

**Informed Consent:** Informed consent was obtained from all individual participants included in the study.

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