The Heart Surgery Forum #2008-1010
11 (4), 2008 [Epub August 2008]
doi: 10.1532/HSF98.20081010

Online address: http://cardenjennings.metapress.com

A Complex Case of Congenital Cardiac Anomaly: Pulmonary Atresia and Ventricular Septal Defect Associated with Major Aortopulmonary Collaterals

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ABSTRACT

Pulmonary atresia and ventricular septal defect associated with major aortopulmonary collaterals (MAPCAs) is a rare, complex, and heterogeneous congenital cardiac anomaly. The majority of untreated patients present with severe congestive heart failure and respiratory distress in the first decade of life. We describe a 15-year-old cyanotic boy, both of whose pulmonary arteries arise from the arcus aorta via patent ductus arteriosus. In addition to this anomaly, the patient has MAPCAs originating from the descending aorta that perfuse the right upper lobe of the lung, a persistent superior vena cava, an aberrant right subclavian artery, pulmonary atresia, and ventricular septal defect.

INTRODUCTION

Pulmonary atresia (PA) and ventricular septal defect (VSD) associated with major aortopulmonary collaterals (MAPCAs) is a rare, complex, heterogeneous congenital cardiac anomaly. The majority of untreated patients present with severe congestive heart failure and respiratory distress in the first decade of life. We present a 15-year-old cyanotic boy, both of whose pulmonary arteries arise from arcus aorta via patent ductus arteriosus (PDA). In addition to this anomaly, the patient has MAPCAs originating from the descending aorta that perfuse the right upper lobe of the lung, a persistent superior vena cava, an aberrant right subclavian artery, PA, and VSD.

CASE REPORT

A 15-year-old boy presented with dyspnea, weakness, and palpitation that were aggravated in the previous year. He has had a history of cold, bluish extremities for 14 years and had not been seen by any physician. A physical examination revealed cyanosis of the lips and tips of the extremities, and clubbing. There was a moderate degree systolic murmur in the mesocardiac area, and the patient's functional capacity was severely restricted. The electrocardiogram revealed right ventricular hypertrophy, right axis deviation, and a normal sinus rhythm. Chest radiography revealed cardiomegaly. The patient had high hemoglobin levels (19.2 mg/dL). His arterial oxygen saturation was 82%, and the PO_2 and PCO_2 in room air were 78 mm Hg and 36 mm Hg, respectively. Transthoracic echocardiogram revealed a large VSD, dextropositioned aorta, biventricular hypertrophy, and pulmonary atresia. Cardiac catheterization (Figure 1) revealed a single truncus that drained blood from both ventricles and insufficiency of the truncal valve. A PDA connected the right and left pulmonary arteries to the arcus aorta. The left pulmonary artery supplied the left lung, and the right pulmonary artery supplied the middle and lower lobe of the right lung. In addition, both arteries were taken off from the PDA. Additionally, MAPCAs originated from the descending aorta and supplied the upper lobe of the right lung. There were stenotic segments on the MAPCAs and on the right and left pulmonary arteries. The right subclavian artery was formed from an aberrant artery from the aorta, which branched off distally from the left subclavian artery. The coronary sinus was enlarged because of the persistent left superior vena cava. Cardiac catheterization revealed systolic pressures of 150, 138, and 142 mm Hg in the left ventricle, right ventricle, and aorta, respectively. Selective pressure measurements in the left pulmonary artery revealed a gradient of 36 mm Hg. A cardiac computed tomography scan was performed to further assess the boy's anatomy (Figure 2). The patient refused to have corrective surgery and was discharged from the hospital against medical advice.

DISCUSSION

The combination of PA, VSD, and MAPCAs is an uncommon complex congenital cardiac anomaly with a variable clinical presentation and morphologic findings depending on the source of the pulmonary blood flow. It is defined as a severe form of Fallot tetralogy. This combination of anomalies occurs in 1.5% of patients with congenital cardiac anomalies, and constitutes 20% of tetralogy of Fallot cases. They are difficult to treat by surgery [DeReuiter 1993].

Received January 23, 2008; accepted May 29, 2008.

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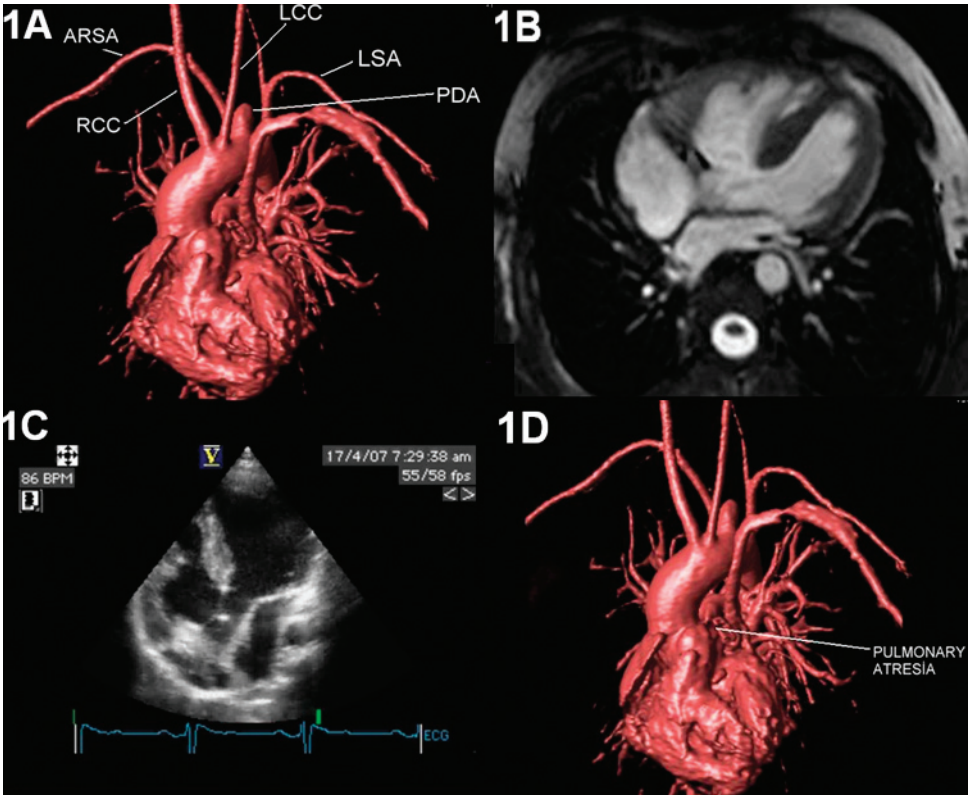

Figure 1. Cardiac computed tomography scan revealing the arcus aorta and its branches (A), ventricular septal defect (VSD) (B), and pulmonary atresia (D). C, Echocardiogram showing the VSD. RCC indicates right common carotid artery; ARSA, aberrant right subclavian artery; LCC, left common carotid artery; LSA, left subclavian artery; PDA, patent ductus arteriosus.

The pulmonary blood flow is provided by the systemic arterial circulation in VSD-PA cases. This anomaly is classified into 3 subgroups according to the origin of the pulmonary blood flow [Liao 1985]. In type A, the most common subgroup, all bronchopulmonary segments are perfused via right and left pulmonary arteries that are connected to each other via the PDA. Type B has many small combined central pulmonary arteries and more distributed MAPCAs, but does not have a PDA. Type C has no native central pulmonary arteries, and the lungs are perfused by multiple MAPCAs. MAPCAs are wide connection arteries that generally originate from the descending aorta and, less commonly, from the arcus aorta or the subclavian artery [Ozden 2007]. Type C anomalies are seen in 20% to 30% of all VSD-PA cases [Puga 1989; Lofland 2004]. Additionally, these patients may also have atrial septal defect or patent foramen ovale. Our patient had the type A form, with MAPCAs originating from the descending aorta and perfusing the right upper lung fields. In addition, he had a persistent left superior vena cava and an aberrant divergence of the right subclavian artery (from the distal left subclavian artery). This condition is an extremely rare variant of this abnormality in the literature.

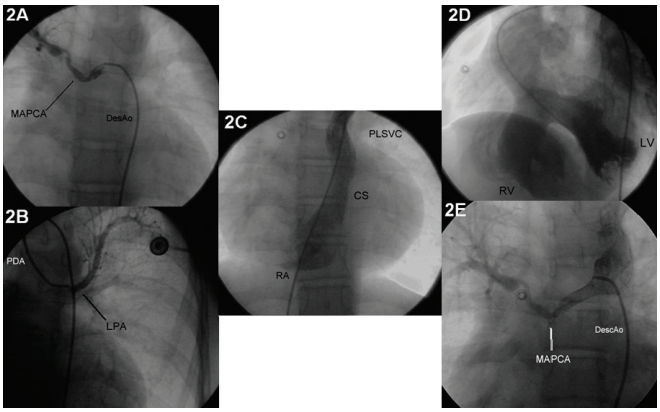

Figure 2. Cardiac catheterization demonstrating major aortopulmonary collaterals (MAPCAs) originating from the descending aorta (DescAo) (A, E). B, Left pulmonary artery (LPA) visualized after it crosses through the patent ductus arteriosus (PDA). C, Persistent left superior vena cava (PLSVC), dilated coronary sinus (CS), and right atrium (RA). D, Left ventriculogram revealing right ventricular (RV) contrast filling secondary to a large ventricular septal defect.

Unifocalization and total-correction surgery are the recommended treatments for patients who have a VSD, PA, and MAPCAs. In such cases, each bronchopulmonary segment of both lungs has a multifocal blood supply. This condition gives rise to an arborization defect. The aim of unifocalization is to rearrange the multifocal perfusion of the lung and make it unifocal again. Single-stage unifocalization and total-correction surgery via a median sternotomy have been recently performed instead of the traditional multistage reconstruction followed by

total-correction surgery with a lateral thoracotomy [Marelli 1994; Belli 2007; Ishibashi 2007]. The former approach has the advantages of fewer surgeries, enabling the pulmonary arteries to develop more effectively. The most controversial issue in surgeries for these cases is the closure of VSDs. The definitive criteria for VSD closure is still debated, although McGoon and Nakata indices are considered in selected cases [Lofland 2004]. Our case was referred for surgery to prevent congestive heart failure; however, the patient refused to give consent for this intervention.

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