CASE REPORTS

Dilated Unroofed Coronary Sinus Mimicking Cor Triatriatum in Cardiac-Type Total Anomalous Pulmonary Venous Connection

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ABSTRACT **We present a rare case of a patient with a dilated unroofed coronary sinus mimicking cor triatriatum that altered mitral inflow in a cardiac-type total anomalous pulmonary venous connection. The membranous tissue in the left atrium, thought to be cor triatriatum tissue preoperatively, was actually a remnant of a dilated coronary sinus tissue. Although this tissue did not create a significant pressure gradient and merely induced a turbulent mitral inflow, we resected it to avoid future problems with mitral leaflet function.** doi: 10.1111/j.1540-8191.2012.01490.x *(J Card Surg 2012;27:621-623)*

We present a rare case of an unroofed dilated coronary sinus altering mitral inflow. This appeared to mimic cor triatriatum in a cardiac-type total anomalous pulmonary venous connection (TAPVC), lacking a persistent left superior vena cava (LSVC). Because of the widely unroofed coronary sinus, there were no unusual findings indicating anomalous connections between the pulmonary veins and the left atrium. The preoperative diagnosis of a TAPVC to the coronary sinus was diagnosed by preoperative three-dimensional (3D) computed tomography (CT), but this could not be confirmed at the time of surgery. The management of this unusual anatomical defect is the subject of this case report.

CASE REPORT

A six-year-old female was referred to our institute for surgical treatment of a coronary sinus defect. The patient was asymptomatic except for frequent upper respiratory infections. On preoperative echocardiography, some membranous tissue was seen in the supramitral area of the left atrium, which was indicative of cor triatriatum (Fig. 1A). However, this tissue did not cause significant mitral stenosis, and just induced turbulent mitral inflow (Fig. 1B). Shunting of blood flow from left to right was observed through the unroofed coronary sinus with 7 to 8 mm in diameter. A preoperative CT scan revealed that all pulmonary veins were connected to an enlarged coronary sinus (cardiac-type TAPVC) without a persistent LSVC (Figs. 1C and D). The membranous tissue in the left atrium was also observed at the supramitral level on the initial CT scan.

Traditional cardiopulmonary bypass was performed using bicaval venous drainage (SVC and inferior vena cava), and a right atriotomy parallel to Waterston's groove was made. The atrial septum was intact and the coronary sinus orifice was enlarged significantly. The atrial septum was incised at the fossa ovalis to examine the membranous tissue in the left atrium, suggestive of cor triatriatum tissue. All the orifices of pulmonary veins, which had been confirmed to drain into coronary sinus by the preoperative CT scan, were easily seen through the incised atrial septum without any interrupted tissues because the coronary sinus was widely unroofed. All pulmonary veins seemed to drain normally into the left atrium. The mitral valve was not easily observed because of the membranous tissue that extended from the dilated unroofed coronary sinus; this is not a typical finding in cases of cor triatriatum. The membranous tissue was mainly located in the posteromedical commissure side of the mitral valve. The membranous tissue had a crescent moon shape and covered the posteromedial side of the mitral valve, from the 1 o'clock to 8 o'clock positions of the mitral annulus. We resected this tissue until the posteromedial side of the mitral annulus was fully exposed without any retraction of the adjacent tissues so that the mitral inflow did not seem to be disturbed (Fig. 2). We then closed the incised atrial septum and coronary sinus orifice with an autologous pericardial patch. As a result, coronary venous return drained into the left atrium. Postoperative echocardiography showed no turbulent mitral inflow. There was no tissue disturbing the mitral inflow in the left atrium and pulmonary venous return was not restricted (Fig. 3). The patient's postoperative course was uneventful.

COMMENTS

There have been some reports about TAPVC to unroofed coronary sinus.[1,2] There is also a report describing a case of cor triatriatum accompanied by a partially unroofed coronary sinus with persistent LSVC; this cor triatriatum tissue was distinct from unroofed coronary sinus tissue.[3] However, there are few

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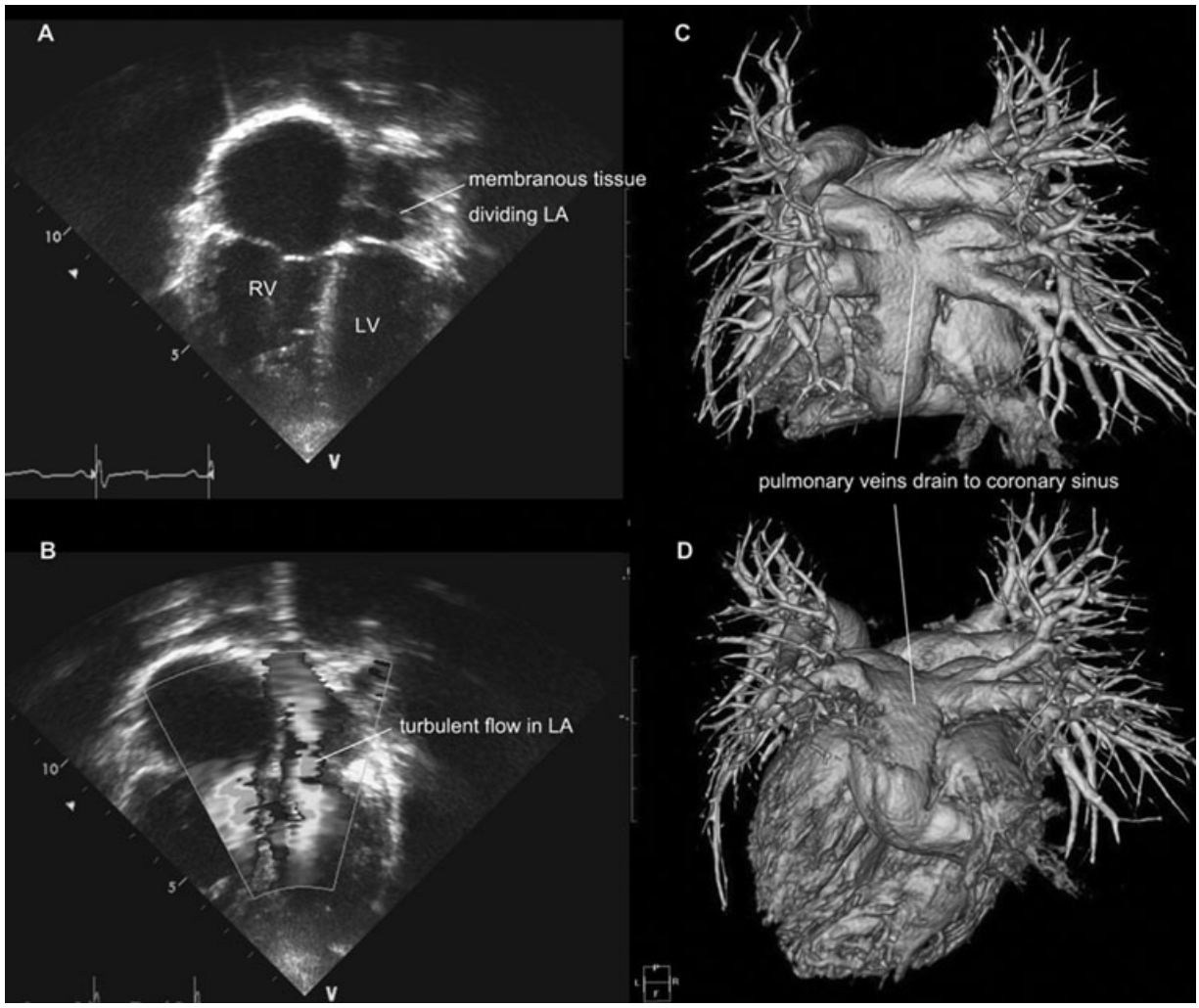

Figure 1. *Preoperative diagnostic findings. Panels A and B are echocardiography images showing membranous tissue in the left atrium mimicking cor triatriatum. Panels C and D are CT 3D images showing all pulmonary veins connected to the enlarged coronary sinus. LA = left atrium; LV = left ventricle; RV = right ventricle.*

Figure 2. *Operative finding in the left atrium through incised atrial septum after resection of membranous tissue disturbing mitral inflow. The black solid line designates the resected tissue area that originally covering the mitral valve.*

reports regarding a TAPVC with an unroofed dilated coronary sinus disturbing mitral inflow and mimicking cor triatriatum in the left atrium, especially without a persistent LSVC. From our experience of this rare case, we would like to make two clinical observations: This case illustrates the necessity of resecting the dilated coronary sinus tissue inducing a turbulent mitral inflow without a significant pressure gradient, and the role of 3D images from CT scans as a preoperative diagnostic tool.

Preoperatively, because there were no other specific anatomical causes of a dilatation of the coronary sinus, such as a persistent LSVC, we considered that the patient's tissue in left atrium was composed of cor triatriatum rather than remnant tissue of a dilated unroofed coronary sinus. At the time of surgery, this membranous tissue appeared to have extended from the coronary sinus, so we believed that this tissue was remnant roof tissue from the dilated unroofed coronary

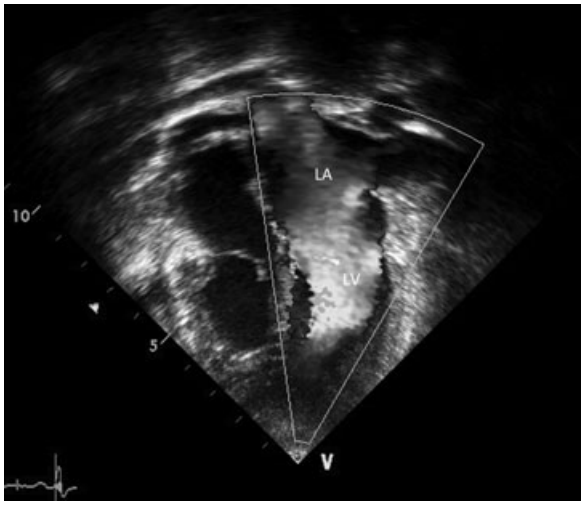

Figure 3. *Postoperative echocardiographic findings. No turbulent flow which was found in the preoperative echocardiographic findings (Fig. 1B). LA = left atrium; LV = left ventricle.*

sinus. Because it did not create a significant pressure gradient or mitral stenosis, we were uncertain whether resection of this membranous tissue would be necessary. However, considering the possible negative effects of turbulent flow on the mitral valve leaflet tissue over time, we decided to remove this tissue. In our patient, all orifices of the pulmonary veins were clearly observed in the left atrium through an atrial septal incision because of the widely unroofed coronary sinus and the locations of the pulmonary veins seemed to be normal. Because there were no definite intraatrial structures connecting the coronary sinus and pulmonary veins, such as a coronary sinus roof, we could not confirm the preoperative diagnosis of a cardiac-type TAPVC to coronary sinus from the operative findings. The only evidence of this was preoperative CT findings (Figs. 1C and D). The 3D images from CT scans were essential in defining the anatomy for this patient.

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Isolated Right Superior Vena Cava Draining into the Left Atrium

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ABSTRACT A 10-year-old male was admitted to our institution with complaints of mild cyanosis and dyspnea. Transthoracic echocardiography and angiography revealed a right superior vena cava (SVC) draining into the left atrium. At the time of surgery, the right SVC was connected to the right atrium. doi: 10.1111/j.1540-8191.2012.01509.x *(J Card Surg 2012;27:623-625)*

A right-sided superior vena cava (SVC) draining into the left atrium is seen in approximately 0.5% of congenital heart anomalies.[1] It is generally associated with other cardiac abnormalities, but rarely occurs as an isolated defect. Anatomically, the right SVC passes medially and dorsally to the aortic root and drains into the left atrium. The right SVC may drain one or more pulmonary veins. Clinically, this anomaly may cause a right to left shunt of approximately 30%.[2] Patients present with mild cyanosis and exercise dyspnea. A brain abscess, due to right to left shunt, may be the first manifestation. We present a case of isolated right SVC draining into the left atrium in a 10-year-old symptomatic patient.

CASE REPORT

A 10-year-old male was admitted with complaints of palpitation, dyspnea, and reduced exercise capacity. Physical examination revealed mild cyanosis and clubbing. Cardiac sounds were normal. No murmur was audible. The oxygen saturation was 87% at room air. Chest X-ray was normal. Transthoracic echocardiography showed abnormal drainage of the right SVC into the left atrium (Fig. 1) with an enlarged left atrium

Conflict of interest: none

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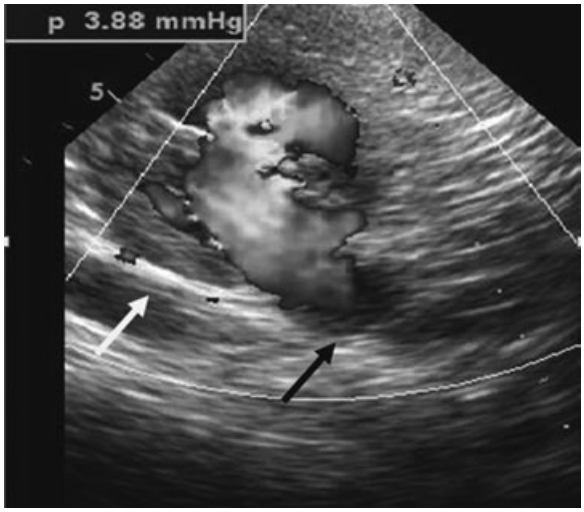

Figure 1. *Echocardiography shows superior vena cava draining into the left atrium (white arrow: left atrium, black arrow: right-sided superior vena cava).*

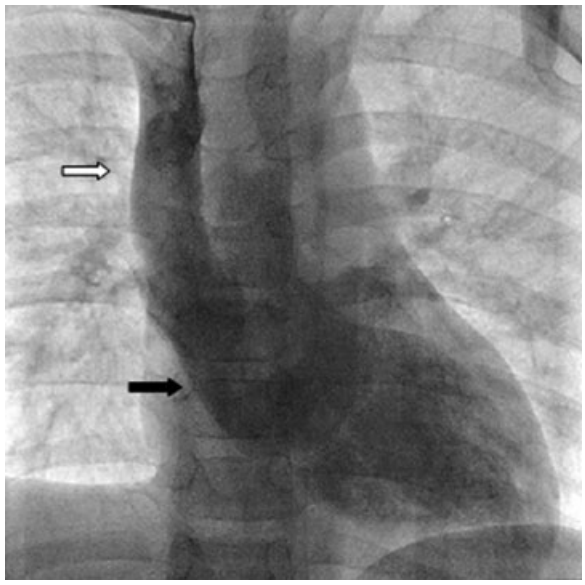

Figure 2. *Superior vena cava injection demonstrates left atrial connection (white arrow: right sided superior vena cava, black arrow: superior vena cava–left atrium connection).*

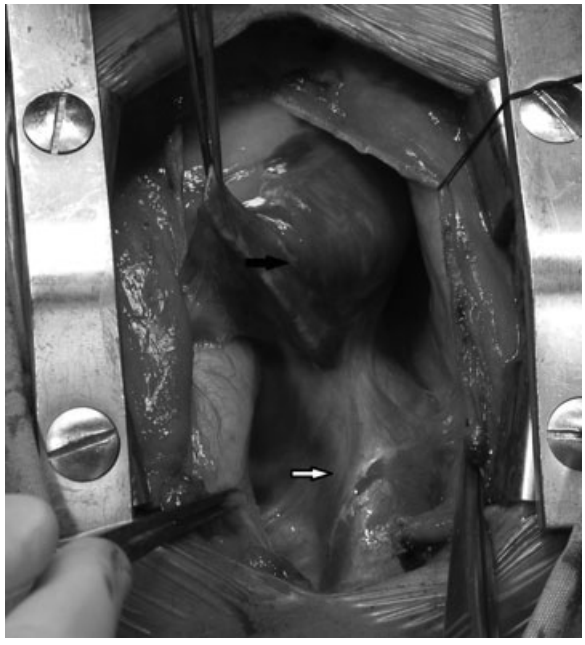

Figure 3. *Operative view shows right SVC draining into the left atrium (white arrow: right sided superior vena cava, black arrow: right atrium).*

(26 mm) and left ventricle (42 mm), and a normal right ventricle. There were no associated defects and the pulmonary venous return was normal. Angiography showed a large superior caval vein draining to the roof of the left atrium via the innominate vein (Fig. 2). Systemic oxygen saturation was 93.8% at the aorta. No accompanying anomaly was diagnosed. The patient underwent cardiac surgery on cardiopulmonary bypass with aortic and bicaval cannulation. The right SVC was seen at the normal position but draining to the left side of the interatrial groove (Fig. 3). A right atriotomy was done following cardioplegic arrest. No atrial septal defect or left SVC was noted. The coronary sinus was not enlarged. The pulmonary veins were also draining to the left artium. The right atrial appendage was removed and significant trabeculations were trimmed. The SVC was directly anastomosed to the right atrium using a continuous suturing technique with 6/0 prolene suture (Fig. 4). The postoperative course was uneventful. The cyanosis was alleviated and his oxygen saturation was 99% at room air. A transthoracic echocardiography showed a normal SVC–right atrium connection without any stenosis or pressure gradient six months following surgery.

DISCUSSION

The first case of an isolated right SVC draining into the left atrium was reported in 1956 by Wood.[3] Kirsch et al. published the first surgically corrected patient.[4] In 1975, de Leval et al. reported that 28 of their consecutive 5127 congenital cardiac surgical patients had a SVC draining into the left atrium.[1] All 28 cases were associated with other cardiac anomalies.

The embryological basis of the pathology is unclear. Abnormal development of the sinus venosus is considered to be responsible for this condition. In the human embryo, the sinus venosus has two valves: the left valve, which is smaller and becomes incorporated in the interatrial septum and the right valve, which obscures the foramen ovale. The right valve has two components: the caudal and the cephalic portion. The thebesian and the eustachian valves are remnants of the caudal portion of the right valve. The cephalic portion becomes reabsorbed into the interatrial septum. Abnormal resorption of the cephalic portion of the right valve may form a seal preventing the SVC–right atrium

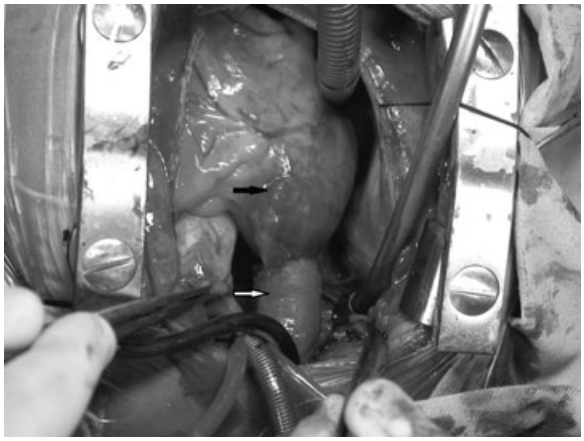

Figure 4. *Operative view shows right atrium superior vena cava anastomosis (white arrow: right sided superior vena cava, black arrow: right atrium).*

connection and may result in a SVC–left atrium communication.[5] Van Praagh et al. postulated that a high venosum atrial septal defect may result from an anomaly of the wall between the right upper pulmonary vein and the right SVC and may cause a left atrium–right SVC shunt.[6]

Several surgical techniques have been used in patients with this anomaly. A preferred technique is an anastomosis of the SVC to the RA in isolated cases.[4,7] Intraatrial rerouting may be considered in patients with atrial septal defect. Kothari et al. used a bidirectional Glenn shunt to repair this anomaly when it was associated with hypoplastic right ventricle.[8] Unfortunately, there are no published long-term follow-up results in the literature to compare different surgical approaches. Thus surgical techniques need to be individualized based on the anatomical features and associated intracardiac anomalies.

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Repair of a Distal Ductal Origin of the Left Pulmonary Artery Associated with a Ventricular Septal Defect

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ABSTRACT Absence of the main branch of the pulmonary artery (PA) with an intracardiac defect is commonly associated with conotruncal cardiac anomaly. It is rarely associated with a ventricular septal defect and PA hypertension. We describe a surgical technique to implant an aberrant left PA to the main PA in a case of a ventricular septal defect, severe pulmonary hypertension, and distal ductal origin of the left PA. doi: 10.1111/j.1540-8191.2012.01506.x *(J Card Surg 2012;27:625-629)*

An absence of the main branch of the pulmonary artery (PA) was first described by Frantzel in 1868.[1,2] The congenital absence of a branch PA is commonly associated with a conotruncal cardiac anomaly[2] and is rarely associated with a ventricular septal defect (VSD) and PA hypertension. We now describe a surgical technique to implant an aberrant left pulmonary artery (LPA) to the main pulmonary artery (MPA) in a 5-year-old female with a VSD, severe pulmonary hypertension, and a distal ductal origin of the LPA. This case report is presented with prior approval of the ethical committee of our institution.

CASE REPORT

A 5-year-old female was admitted with a history of failure to thrive, palpitations, and recurrent respiratory

Conflict of interest: None.

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TABLE 1
Cardiac Catheterization Data

Study	Preoxygenation	Postoxygenation
Pulmonary artery pressure	104/71 (84) mmHg	80/48 (61) mmHg
Pulmonary artery saturation	82.3%	98.3%
Femoral artery saturation	97%	100%
Pulmonary blood flow	4.6 L/min	12.8 L/min
Systemic blood flow	2.8 L/min	3.2 L/min
Left to right shunt	1.7:1 L/min/m^2	3.9:1 L/min/m^2
Pulmonary vascular resistance index (PVRI)	9.5 Woods unit	2.3 Woods unit

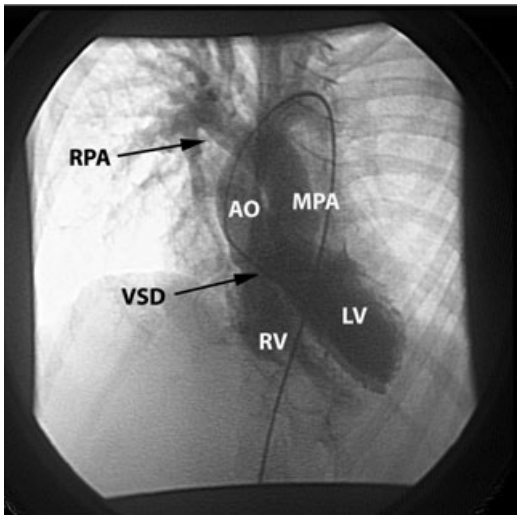

Figure 1. *Left ventriculogram showing large subaortic VSD with normal sized MPA and RPA. The LPA is absent. VSD = ventricular septal defect; MPA = main pulmonary artery; RPA = right pulmonary artery; LPA = left pulmonary artery.*

tract infections. The physical examination showed left ventricular type of apex, Grade II parasternal heave, and Grade 5/6 Pan systolic murmur in the fourth and fifth left intercostal space; the pulmonary second sound was loud with wide and variable split. Chest X-ray showed a cardiothoracic ratio of 60%, reduced pulmonary blood flow on the left side, and pulmonary plethora on the right side. Two-dimensional echocardiography showed situs solitus, levocardia, a large perimembranous VSD, left to right shunt with a gradient of 11 mmHg, and normal biventricular function. The cardiac catheterization data showed preoxygenation PA pressure of 104/71 mmHg with a mean of 84 mmHg, the left to right shunt of 1.7:1 L/min/m^2, and pulmonary vascular resistance index (PVRI) of 3.9:1 L/min/m^2. The postoxygen PA pressure came down to 80/48 mmHg with mean of 61 mmHg, the left to right shunt increased to 3.9:1 L/min/m^2, and PVRI came down to 2.3 Woods unit. The detailed cardiac catheterization data are described in Table 1. The left ventriculogram showed a subaortic VSD and absent LPA (Fig. 1). An aortogram demonstrated a LPA arising from the arch of the aorta (Fig. 2).

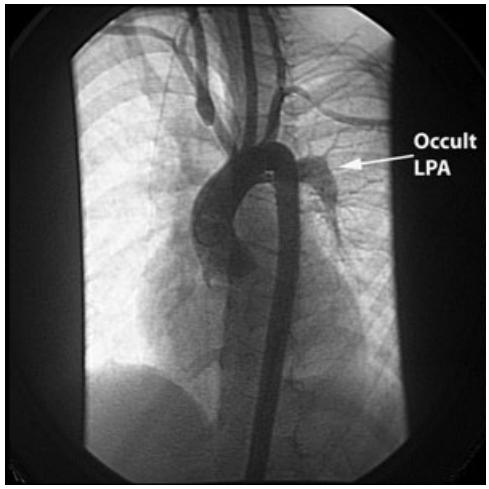

Figure 2. *Aortic root injection showing an occult LPA arising from the undersurface of the aortic arch. LPA = left pulmonary artery.*

SURGICAL TECHNIQUE

A conventional median sternotomy approach was used and cardiopulmonary bypass was established with aortic and bicaval venous cannulae. Moderate hypothermia and antegrade blood cardioplegia was used for myocardial protection. The intraoperative findings revealed normally related great arteries, a left aortic arch, and the MPA was enlarged and tense (Figs. 3 to 6). The LPA was absent at its normal position on the MPA but was arising from the undersurface of the aortic arch. The origin of the aberrant LPA was stenotic but was 1 cm distally. The pulmonary venous drainage was normal with a small patent foramen ovale and a large (3 cm × 2 cm) perimembranous VSD. The VSD was closed with a Dacron patch through the right atrial approach using interrupted 5-0 polypropylene sutures. The LPA was dissected from the aortic end till the hilum and the aortic end was ligated, transfixed, and divided. The proximal 2 mm of the LPA was excised and was slit opened up to the hilum. The divided and slit opened LPA was anastomosed to the MPA in such a way that the native LPA formed the posterior wall of the neo LPA and the anterior wall was prepared from an autologous untreated pericardial patch. The pericardial patch was sized over the hegar dilator for the weight of the patient according to the Kirklin's nomogram.[3] The pump time was 3 hours and 4 minutes and clamp time was 2 hours and 24 minutes. The patient was extubated on the first postoperative day, and was discharged on the sixth postoperative day. The postoperative two-dimensional

Figure 3. *Intraoperative picture showing the aberrant LPA arising from the undersurface of the aortic arch being looped. LPA = left pulmonary artery.*

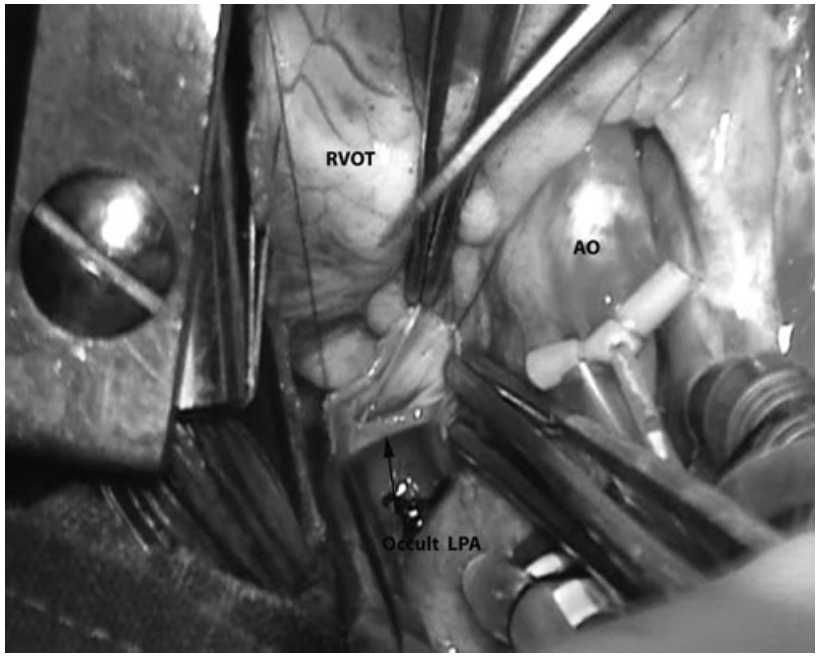

Figure 4. *Intraoperative picture showing vertically cut opened LPA. This portion of LPA will form the posterior wall of the neo LPA. LPA = left pulmonary artery.*

echocardiogram showed no residual VSD, or pericardial effusion, a right ventricular systolic pressure of 44 mmHg, normal PA anatomy, and no gradient across the MPA and LPA. The echocardiography at three-month and two-year follow-up revealed no residual VSD, no gradient at the LPA origin, and normal PA end diastolic pressure. Angiography performed to evaluate the patency of the implanted LPA at follow-up revealed a patent LPA (Fig. 7).

DISCUSSION

In 1954, McKim and Wiglesworth, and Sotomora and Edwards pointed out that the so-called "absent

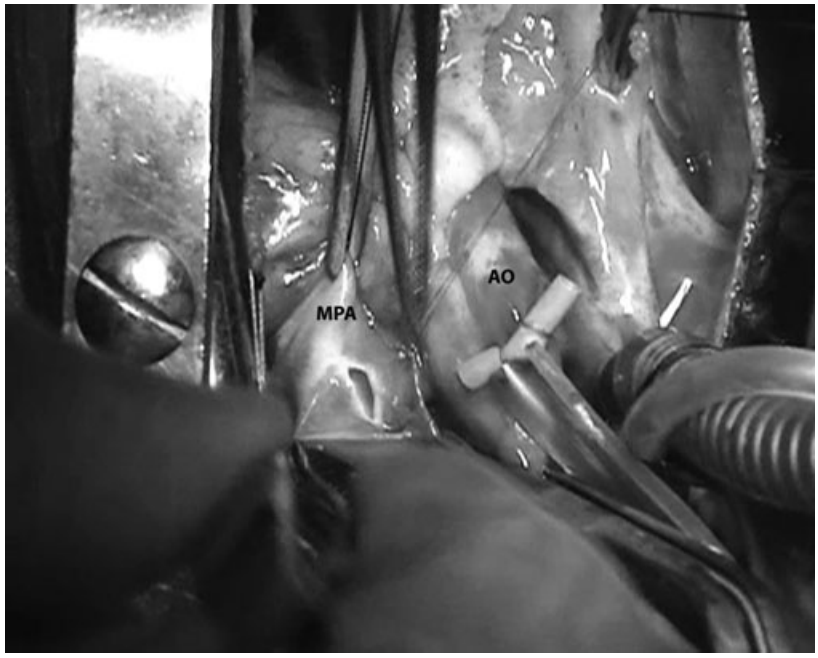

Figure 5. *Intraoperative picture showing a vertical opening made in the MPA to anastomose the occult LPA into its normal position. The posterior wall of the neo LPA will be formed by slit opened LPA and the anterior wall by an autologous pericardial patch. MPA = main pulmonary artery.*

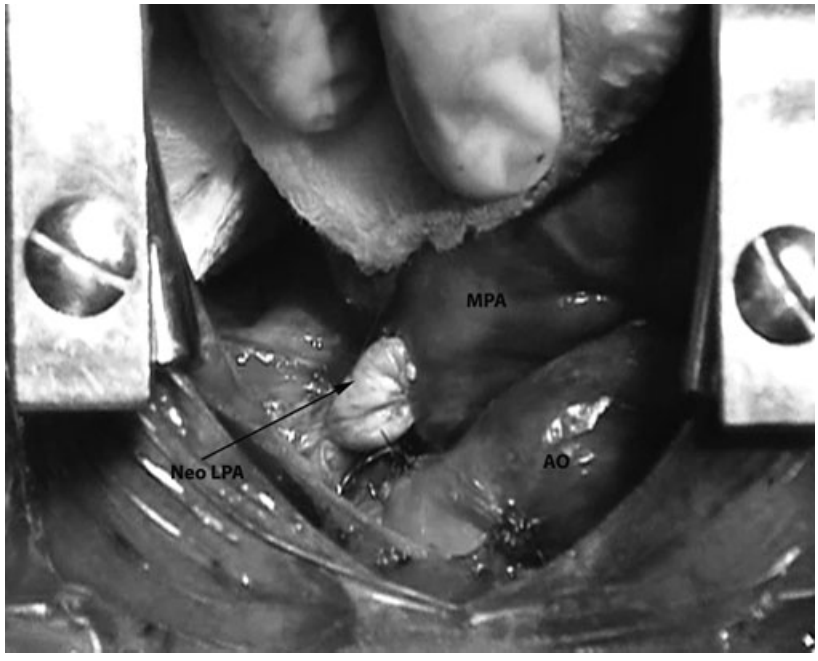

Figure 6. *A completed repair and the final appearance of the neo LPA. LPA = left pulmonary artery.*

PA" is in fact usually present and is associated with persistence of the ductus on the same side as the interruption which was described as "Ductal origin of the distal PA."[1,2,4] The incidence of this condition is approximately 1 in 100,000 newborns.[2] Embryologically the proximal PA branches develop from the proximal sixth pharyngeal arches. Nonconfluence of the pulmonary arteries may be due to regression of the sixth arch segment as part of conotruncal abnormalities.[5] Various surgical methods can be used if the native LPA is not long enough or the anastomosis is under tension. These include a pericardial, homograft, Dacron, or a polytetrafluoroethylene tube. If the LPA is not developed a staged repair can be done using a

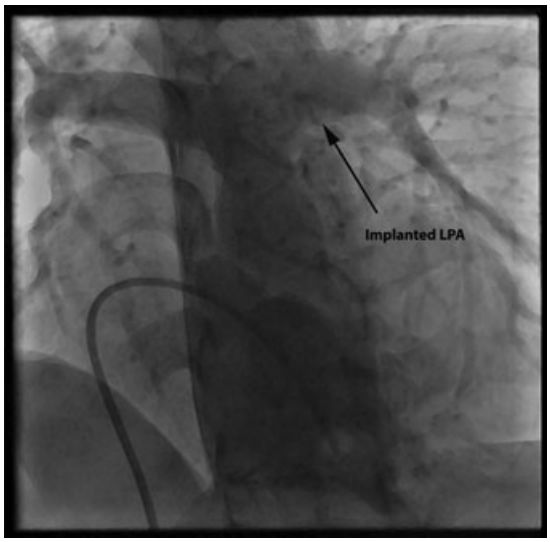

Figure 7. *Postoperative angiogram showing patent implanted LPA. LPA = left pulmonary artery.*

systemic pulmonary shunt to the LPA and closure of the intracardiac defect followed by second-stage implantation of the LPA to the MPA.

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