## Comment

The advantage of using a stentless bioprosthesis for aortic valve replacement has been well documented [1–3]. Given our patient's reluctance to undergo anticoagulation, a mechanical valve was not an option in this case. It would have been technically easier to implant a stented tissue valve, but only a very small bioprosthesis would have fit. Other reasonable options include root reconstruction with either a Freestyle stentless porcine aortic root or a homograft, but this would be a more complicated operation notwithstanding the mitral valve repair procedure also required in this case.

Ikonomidis and Miller [4] recently reported a successful implantation of a Toronto SPV in a 51-year-old woman with severe aortic regurgitation after a David-I-type valve-sparing aortic root replacement. Their experience and the case we describe here are unique because they involve an implantation of a stentless valve through the confines of a preexisting aortic graft. In our case hand, we have adopted our previously described technique of implantation of the Toronto SPV valve [5] with minor adjustments to account for the different anatomy as outlined above.

The use of aortic homograft facilitates reconstruction in complex aortic valve endocarditis and offers resistance to reinfection [6], hence the choice of a homograft at this patient's first operation. A valid concern in the case at hand is recurrent endocarditis affecting the stentless valve. Furthermore, although reports of the durability of stentless aortic valves have been encouraging [1, 3], it poses a significant concern especially in this patient who has already had a failed homograft reconstruction in a brief time span.

This case is unusual given the absence of the typical findings that are associated with allograft structural degeneration such as calcification and disfiguration. The finding of valvulitis manifested by leaflet pathology with gross sparing of the remaining components of the homograft is intriguing. That could be due to autoimmune factors related to the patient's history of rheumatoid arthritis, with further variables imposed by the history of liver transplantation and immunosupression. Native aortic valvulitis has been described in a variety of autoimmune disorders, and both granulomatous and nonspecific valvulitis have been reported in patients with rheumatoid arthritis [7]. A case of recurrent autoimmune valvulitis affecting an aortic bioprosthesis has been described in a patient with systemic lupus erythematosus [8]. Our case demonstrates the versatility of the stentless aortic bioprosthesis and its successful application in an operation tailored to the patient's specific variables.

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FEATURE ARTICLES

# Isolated Unilateral Absence of Right Proximal Pulmonary Artery: Surgical Repair and Follow-Up

Kristin Welch, MD, Frank Hanley, MD, Troy Johnston, MD, Colleen Cailes, RDCS, and Maully J. Shah, MBBS

Department of Pediatrics, University of Washington School of Medicine, Seattle, Washington, and Department of Cardiothoracic Surgery, Stanford University School of Medicine, Palo Alto, California

**The isolated unilateral absence of a proximal pulmonary artery is a rare congenital lesion with a diverse clinical presentation. If the connecting ductus arteriosus closes after birth, the ipsilateral pulmonary artery will lose its source of blood supply, resulting in hypoplasia or obliteration of intrapulmonary vessels. Despite a seemingly benign early clinical course, a significant number of untreated patients will develop pulmonary hypertension, hemoptysis, and recurrent respiratory infections. Early detection and surgical repair provides restoration of physiologic pulmonary circulation, regression of pulmonary hypertension, and the potential for normal distal pulmonary vascular development.**

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Address reprint requests to Dr Shah, University of Pennsylvania, Childrens Hospital of Philadelphia, 34th and Civic Ctr Blvd, Philadelphia, PA 19104; e-mail: shahm@email.chop.edu.



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The isolated unilateral absence of a proximal pulmonary artery (UAPA) is a rare congenital lesion with a wide spectrum of presenting symptoms [1]. Detection of UAPA in infancy offers the opportunity for early surgical intervention to provide antegrade blood flow to the hilar pulmonary arteries and potentially prevent development of hypoplasia of the ipsilateral pulmonary vascular bed. We report the surgical repair and follow-up of two patients with UAPA in whom surgical interventions were performed early in life.

## Case Reports

### *Patient 1*

A neonate was referred for evaluation of a patent ductus arteriosus at 7 days of age. The chest roentgenogram showed slightly diminished right lung volume and pulmonary vascular markings compared with the left. A right pulmonary artery (RPA) could not be identified by an echocardiogram. Hemodynamic data from cardiac catheterization was significant for systemic level pulmonary hypertension. An angiogram revealed an absent proximal RPA, but a pulmonary vein wedge angiogram revealed a hilar RPA (Fig 1A, 1B). In addition, there was a left aortic arch with a left patent ductus arteriosus and the presence of a right ductus diverticulum from the base of the right innominate artery. The patient was started on a prostaglandin infusion (0.1 $\mu$g/kg/min) in an attempt to reestablish right ductus arteriosus patency but this was unsuccessful.

Surgical exploration confirmed the lack of an intrapericardial RPA. The left ductus arteriosus was ligated and divided. A long segment of occluded ligamentum arteriosis lead from the base of the right innominate artery to a well-developed hilar RPA. The ligamentum arteriosis was ligated and divided at the base of the innominate artery. It was filleted open, revealing a small but present lumen. The vessel was opened onto the right lower lobe artery. The right posterolateral portion of the distal main pulmonary artery was identified as a target site for the implantation of the "neo" RPA. Under cardiopulmonary bypass, the opened right ligamentum and RPA were then placed under the ascending aorta, and a direct tissue-to-tissue anastomosis to an opened orifice in the main pulmonary artery was made with a running 7-0 Maxon (Sherwood-Davis & Geck, St. Louis, MO) suture. A pulmonary arterial homograft patch was used to augment the anterior surface of the pulmonary artery from the right lower lobe artery branch all the way back to the main pulmonary artery, creating a "neo" RPA of approximately 6 to 7 mm in diameter. A postoperative lung perfusion scan showed 47% of flow to the right and 53% of flow to the left. The patient was discharged home on postoperative day 5.

The patient has had serial outpatient follow-up for 31 months after surgery. The patient is asymptomatic and is growing and developing normally. Chest roentgenograms show normal heart size and equal right and left lung volumes. Serial echocardiograms have shown a patent RPA with no increase in flow velocity. There is no evidence of pulmonary hypertension. A lung perfusion scan at 12 months after surgery showed 50.5% flow to the right and 49.5% flow to left lung, and at 24 months after surgery, flow to both the right and left lungs was 50%. In addition, the patient had a cardiac catheterization 30 months after the "neo" RPA construction that revealed normal pressures in the right ventricle and right and left

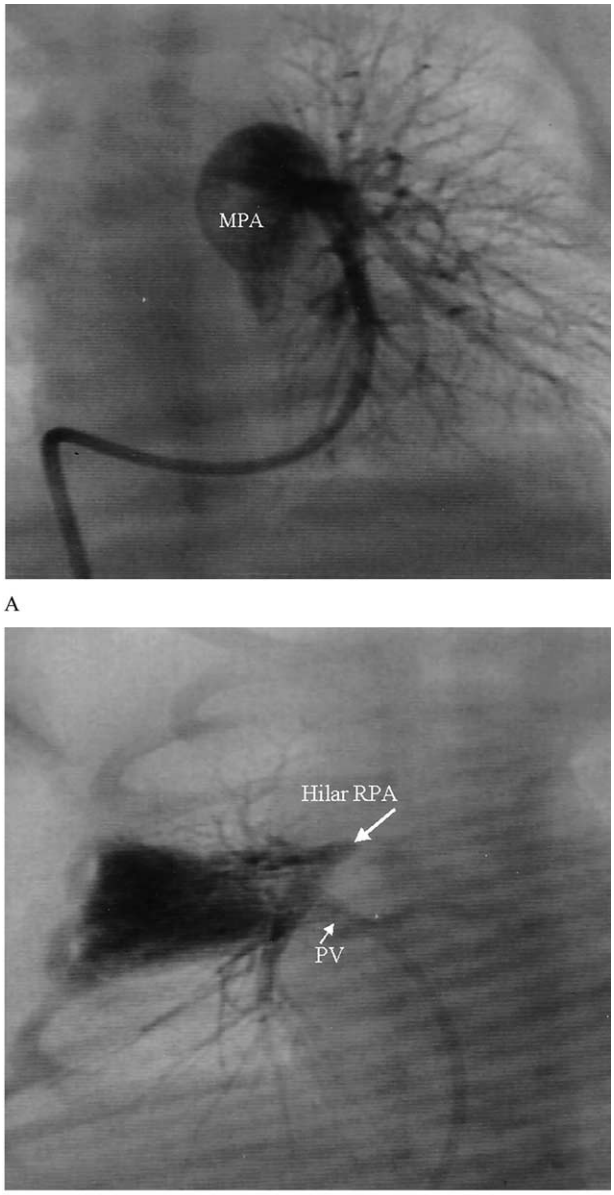


*Fig 1. (A) Angiogram with a catheter positioned in the main pulmonary artery* (MPA) *reveals opacification of the left lung vasculature and the absence of the right pulmonary artery. (B) Right pulmonary vein wedge angiogram obtained in the arterial phase shows a hypoplastic peripheral vessel* (small arrow) *that reaches only to the hilum* (large arrow). (PV = *pulmonary vein;* RPA = *right pulmonary artery.)*

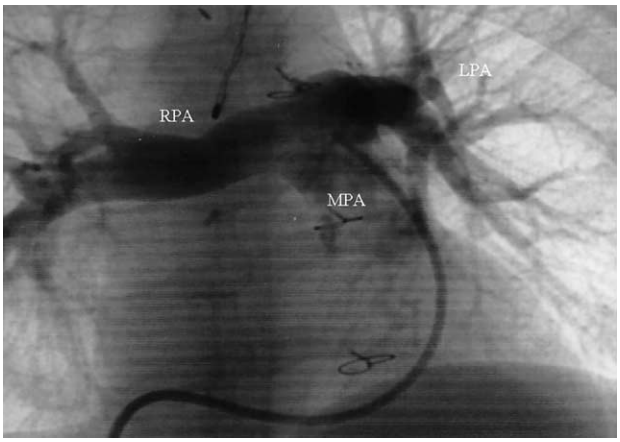


*Fig 2. Angiogram in patient 1 performed 30 months after surgical intervention. Opacification of the "neo" right pulmonary artery (*RPA*) and left pulmonary artery (*LPA*) is seen. The "neo" RPA is unobstructed with a uniform caliber. (*MPA = *main pulmonary artery.)*

pulmonary arteries. The angiogram revealed a patent RPA of normal caliber, with no evidence of stenosis, and a normal arborization pattern (Fig 2).

### *Patient 2*

A 3-month-old infant presented for evaluation of a murmur, increased respiratory effort, and poor weight gain. A chest roentgenogram showed cardiomegaly and mildly diminished right lung volume. An echocardiogram showed the absence of the proximal RPA, moderate tricuspid regurgitation, and right ventricular hypertrophy. Cardiac catheterization demonstrated systemic level right ventricle and left pulmonary artery pressures. An angiogram revealed an absent proximal RPA, but a pulmonary vein wedge angiogram identified a right hilar pulmonary artery. An aortogram showed a left aortic arch and bilateral ductal diverticulum, with the right diverticulum coming off the base of the right innominate artery.

Intraoperative findings confirmed the absence of an intrapericardial RPA. A long segment of occluded ligamentum arteriosis lead from the base of the right innominate artery to a hilar RPA. The "neo" RPA was constructed and anastomosed to the MPA as previously described. A lung perfusion scan on postoperative day 9 showed 63% of flow to the right lung and 37% of flow to the left lung. The patient was discharged home on postoperative day 11.

The patient has had serial outpatient follow-up for 30 months after surgery. The patient is asymptomatic and is growing and developing normally. Serial echocardiograms have shown a patent RPA with no increase in flow velocity. There is trivial tricuspid valve regurgitation with no evidence of pulmonary hypertension. A lung perfusion scan at 12 months after surgery showed 64% flow to the right lung, 36% flow to the left lung, and at 24 months after surgery, flow to the right lung was 65% and flow to the left was 35%.

## Comment

The exact prevalence of isolated UAPA is unknown, but the best estimation is 1:200,000 individuals [2]. The principal embryologic explanation for UAPA is involution of the proximal sixth aortic arch that results in the absence of the proximal pulmonary artery and the persistent connection of the hilar pulmonary artery to the distal sixth aortic arch (ductus arteriosus) [3]. A literature review revealed that all reported cases of UAPA had a ductus arteriosus or ligamentum ipsilateral to the absent pulmonary artery. If the connecting ductus arteriosus closes after birth, the ipsilateral pulmonary artery will lose its source of blood supply, diminish in size, and not be visible with angiographic or echocardiographic imaging. The affected lung is subsequently supplied from the bronchial arterial bed and anomalous systemic collaterals.

The diagnosis of UAPA can be facilitated by chest roentgenogram, echocardiogram, angiogram, contrast-enhanced computed tomographic scan, lung perfusion scans, and above all, a high index of suspicion. Pulmonary venous wedge angiography is particularly useful in delineating the presence of an ipsilateral hilar pulmonary artery and intrapulmonary vessels. The clinical course of unrepaired UAPA is varied and some patients can be asymptomatic for a relatively long time. Symptoms can be unmasked by factors such as pregnancy or high altitude. In the long term, unrepaired UAPA is associated with significant morbidity. A literature review by Ten Harkel and colleagues found that pulmonary hypertension was present in 44%, hemoptysis in 20%, recurrent pulmonary infections in 37%, and limited exercise tolerance was present in 40% of patients with unrepaired isolated UAPA. Overall mortality in this series was 7% [1].

Early surgical intervention may potentially preserve the affected lung vasculature and prevent morbidity and mortality. Regression and hypoplasia of the affected pulmonary artery begin soon after the ductus arteriosus closes. In an experimental animal study with ligation of the LPA, Haworth and colleagues demonstrated diminution and obliteration of the hilar and axial pulmonary arteries in rapidly growing pigs despite a satisfactory bronchial arterial supply to the intraacinar pulmonary arteries [4].

The surgical approaches for UAPA include establishing an ipsilateral systemic-to-pulmonary artery shunt, an interposition tube graft with autologous pericardium or prosthetic material, and mobilization with end-to-side anastomosis of the affected artery to the main pulmonary artery [5–8]. The immediate outcomes of most surgical procedures have been gratifying, but follow-up data that includes patency of conduits, growth of the pulmonary artery, and regression or persistence of pulmonary hypertension have been infrequently reported. A review of the literature does indicate that patients who have had surgical interventions for UAPA after infancy have a less favorable outcome [3]. The obvious disadvantages of a synthetic conduit are its "fixed size "and the possible

need for long-term anticoagulation to maintain conduit patency.

The surgical technique in our case series incorporated the filleted ligamentum arteriosus to bridge the gap between the hilar RPA and main pulmonary artery. A pulmonary arterial homograft patch was used to augment the anterior surface of the "neo" RPA from the right lower lobe branch all the way to the main pulmonary artery. Follow-up of both patients indicates an uncomplicated postoperative course. Serial echocardiograms have shown a patent "neo" RPA and no evidence of pulmonary hypertension. Serial lung perfusion scans have demonstrated near normal distribution of blood flow to both lungs. Patient 1 also had a cardiac catheterization 30 months after the surgical repair that confirmed the normalization of pulmonary artery pressures. The angiogram demonstrated a patent RPA, which appears to be growing with the patient.

Our experience with these two patients provides additional support to the hypothesis that the primary repair of an isolated UAPA can be accomplished in the neonatal period and infancy with a relatively low risk and an excellent outcome. Pulmonary hypertension regressed in both of our patients after surgical repair. The surgical technique used to construct the neo "RPA" has allowed for a normal caliber of the RPA as seen by echocardiographic (patient 1 and 2) and angiographic (patient 1) imaging.

Early primary repair of an isolated unilateral absent proximal pulmonary artery provides restoration of physiologic pulmonary circulation, regression of pulmonary hypertension, and the potential for normal distal pulmonary vascular development.

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FEATURE ARTICLES

# Epitheloid Hemangioendothelioma of the Heart in Infancy

Turkan Tansel, MD, Ümrah Aydogan, MD, Dilek Yilmazbayhan, MD, Bilge Bilgiç, MD, Misten Demiryont, MD, and Ertan Onursal, MD

Departments of Cardiovascular Surgery, Pediatric Cardiology, and Pathology, Istanbul University, Istanbul Faculty of Medicine, Istanbul, Turkey

**We report a case of primary cardiac epithelioid hemangioendothelioma arising from the right atrium of a 2-month-old infant. The tumor was found incidentally during exploratory sternotomy for recurrent pericardial effusion. This case represents a very rare situation, because this is the youngest patient found in relevant literature, and because it involves extensive infiltration by the tumor without any development of intracardiac mass appearance.**

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Primary tumors of the heart are rare, with an incidence of 0.0017% to 0.028% in reported series [1]. Hemangioma of the heart constitutes about 2.8% of all primary cardiac tumors [1], whereas cardiac occurrences of epithelioid hemangioendothelioma are exceptionally rare. We believe that only five cases of epithelioid hemangioendothelioma of the heart have been previously reported in the literature. We report a case of epithelioid hemangioendothelioma of the right atrium as the first description of this tumor occurring in infancy.

A 21-year-old primigravida who had no previous medical history and an uncomplicated pregnancy had presented for a routine ultrasound examination. The ultrasound examination at 34 weeks of gestation revealed the presence of a fetal pericardial effusion. A follow up was planned. At 38 weeks of gestation, a male infant weighing 3.050 kg was delivered after spontaneous rupture of the membrane. The infant showed signs of cardiorespiratory distress immediately after delivery with apnea, cyanosis, and signs of poor peripheral circulation, with a heart rate of 90 beats per minute and blood pressure of 45/30 mm Hg. No significant improvement occurred after endotracheal intubation, mechanical ventilation, and inotropic support. No murmurs were heard on auscultation. The liver was palpated 2 cm below the right costal margin. The chest roentgenogram revealed a marked enlargement of the cardiac shadow. Echocardiography demonstrated a large pericardial effusion. A pericardiocentesis

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Address reprint requests to Dr Tansel, Istanbul University, Istanbul Faculty of Medicine, Department of Cardiovascular Surgery, Çapa, Istanbul 34390, Turkey; e-mail: turkant@superonline.com.



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